Dedication

To my wife Rani, who is the love of my life, my soulmate, the best life partner a man can have and who will always be the better part of me. No words are possible to describe how much I owe her or all her wonderful qualities. She has taught me that love can really conquer all. Without her, this book would not be possible, nor my success as a pediatric cardiologist.

To my children Arielle and Benjamin who keep me honest, young and energetic; their love and devotion have a special and permanent place in my heart.

To my brother Bobby, who taught me that being a true seeker of truth and understanding is not just a dream.

To my mother-in-law and father-in-law, Lorraine and Herb Kulik, for all their love and support.

And finally to my mother Sylvia, whose passing occurred during the writing of this book. She not only helped me become who I am today, but her fight to stay alive along with her comfort, warmth, natural insight and sense of humor spurred me on to bigger and better things. She is sorely missed by all who came in contact with her; the light of the world is dimmer without her.
Principles and Practice of Cardiac Magnetic Resonance in Congenital Heart Disease Form, Function, and Flow

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- The world of cardiac magnetic resonance is a small one, but ever growing one. I have been fortunate enough to have interacted with, learned from and become friends with giants who started the field such as Nathaniel Reichek, Leon Axel, Gerry Pohost and Charlie Higgins. It has also been an honor to work with the greatest colleagues in the world such as Victor Ferrari, Edward Martin, Tal Geva, Andrew Powell, Shi Joon Yu, Taylor Chung, Warren Manning, Christopher Kramer … the list is exhaustive and can go on for many pages. Ajit Yoganathan has been a close research collaborator and friend; his keen mind, wit and drive has been goal for me to always strive for. One special thank you needs to be made to Eric Hoffman who gave me my start in CMR research many years ago – his intelligence, kindness and understanding will always be remembered fondly.

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• Finally, no medical student who has gone through the program at Upstate Medical Center in Syracuse, NY in the 80s can ever publish a scholarly textbook without the mention of Robert Rohner who not only was one of the best teachers ever but also instilled the love of medicine in us all and taught us to be proud of who we are.

Ultimately, however, I want to thank you, the reader, whose interest in reading or purchasing this textbook is the spark which gave this project its life in the first place.
In general, I’m a skeptic about medical textbooks and have declined invitations to edit or write chapters for them or to review them whenever possible. Some are out of date on publication while only a minority are lasting major contributions that endure through many editions and have an impact that justifies the immense effort the editor and authors invest. Nonetheless, I recall vividly a few earlier opportunities to read emerging medical classics prior to publication, offered by revered mentors. In each case, it was clear on inspection that something important was at hand. One example was the first edition of Joseph K. Perloff’s *The Clinical Recognition of Congenital Heart Disease*. A second was Harvey Feigenbaum’s first edition of *Echocardiography*. Each marked a milestone in the development of contemporary cardiology and each remains in print to this day in 5th and 6th editions respectively.

As I closed the CD containing the draft chapters of Mark A Fogel’s *Principles and Practice of Cardiac Magnetic Resonance in Congenital Heart Disease: Form, Function and Flow*, it seemed to me that here was another milestone in the development of contemporary cardiology and each remains in print to this day in 5th and 6th editions respectively.

It would be difficult for someone who has never tried to do pediatric CMR to appreciate the host of major technical and clinical challenges that a successful study in a complex patient can present. Rapid heart rates, patient motion, respiratory motion during obligatory free-breathing acquisitions and limited spatial resolution all conspire against success. It takes a great deal of expertise, knowledge, determination and resourcefulness to optimize imaging techniques and strategies and consistently obtain quality diagnostic studies. Nonetheless, the field has flourished, thanks to the efforts of a very talented community of investigators, a great many of whom are represented among the 41 authors of the 25 chapters of this text. The scope of their contributions is truly remarkable, covering the breadth of pediatric cardiology and demonstrating both a deep understanding of cardiac pathology and pathophysiology and a tremendous grasp of the technical issues involved in getting the most from pediatric cardiac CMR. It is clear from their work how powerful a tool CMR has become in the armamentarium of pediatric cardiology and pediatric cardiac radiology. We can expect that this text will endure and evolve as a classic in cardiac imaging for many years to come.

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Mitchell *et al.*, in 1971, defined congenital heart disease (CHD) as a "gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance" [1]. It is not only one of the major causes of morbidity and mortality in pediatrics, it is also very common. Hoffman and Kaplan [2] recently surveyed the literature and found the incidence of moderate to severe forms of CHD to be 6/1000 live births, rising to 19/1000 live births if potentially serious bicuspid aortic valves are included. All forms of CHD represent 75/1000 live births including such lesions as tiny muscular ventricular septal defects. The New England Infant Cardiac Program [3] reported that 3/1000 live births who needed cardiac catheterization died with CHD in early infancy (excluding premature infants with patent ductus arteriosus). This number rises to 5/1000 live births who will need some kind of specialized facilities during their lifetime.

In addition, because of major advances over the past 50 years, more and more of these children are growing into adulthood and require medical attention and follow-up [4,5]. This growing population is being taken care of by an increasing number of adult cardiologists and internists. In 1980, there were an estimated 300 000 adults with CHD while in the year 2000, this rose to approximately 1 million. In 2020, the number is anticipated to be 1.4 million. It is therefore incumbent, not only on the pediatrician, pediatric cardiologist, cardiac surgeon or cardiac anesthesiologist to familiarize themselves with all the nuances of this branch of pediatrics, but the family practitioner, internist, adult cardiologist and intensivist also need to have a working knowledge of this as well.

To understand CHD is to also understand that it is one of the more technical disciplines within pediatrics and medicine in general. This is based on the continued evolution of diagnosis and management grounded on advances in technology. With the advent of specialized imaging techniques, our knowledge of this field is vastly different now than it was just 10 years ago. But CHD is not just technical in that respect; complex anatomy, physiology, hemodynamics, ventricular function and blood flow along with their interactions with each other form an intricate web of connections and associations that need to be deciphered.

It is imperative, therefore, because of their complexity, that the imager performing cardiac magnetic resonance (CMR) understand not only anatomy, surgery, physiology and function in CHD but the technical aspects of imaging as well. The tradeoffs of spatial and temporal resolution cannot be used in small children because their smaller structures demand higher spatial resolution combined with their faster heart rates which demand a higher temporal resolution when compared with adults. Breath-holding in children can also be problematic. “Work-arounds” have been developed for these issues but more needs to be done. In addition, with the ever increasing sophistication of technology, more can be done with CMR in a high quality manner in a shorter period of time without invasiveness than was dreamed of 10 years ago. We owe it to our patients to apply these techniques to improve their care.

There are many textbooks on magnetic resonance imaging but very few on its application to cardiology. Within those, CHD may only have a chapter or two which certainly does not do the field justice. It would be instructive and much more efficient, I thought, to have thorough summaries of all the aspects of CMR in one place which is devoted solely to CHD. The idea for this textbook is to allow the trainee, practitioner and the
researcher to have a repository of information on which to base their use of CMR.

Any text on this subject must be organized in a logical way for the reader to obtain a complete understanding of the issues involved. Towards that end, the textbook is divided into three major sections. The first involves familiarizing the reader with the minimum tools needed to understand the basics of CMR as it is applied to children and CHD such as evaluating morphology, ventricular function and utilizing contrast agents. The next section discusses the broad categories of CHD and the use of CMR in these disease states. Finally, a third section highlights special topics such as CT scanning and interventional CMR.

Because of space and time constraints, this textbook cannot be encyclopedic. Indeed, issues related to other topics such as the details of image generation, research using CMR and new and innovative CMR techniques as well as improved hardware and software have not been explored. I have endeavored to make the format of this textbook a "core curriculum" with references to the high quality publications on the parts of this discipline which are not covered here. My hope is that this textbook will not only be used as a learning tool and reference, but will also act as a springboard to further study of the technology as it relates to CHD. It is also my hope that this textbook will excite the reader about this fascinating field and will spur future advances which may one day impact patients.

Mark A. Fogel
2010

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PART I
The basics of cardiac MR
CHAPTER 1

Physics of cardiac MR and image formation

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Introduction

Magnetic resonance imaging (MRI) of the cardiovascular system continues to expand its application and importance as a diagnostic tool in adult and pediatric patients. In order to successfully apply this technology and interpret cardiac magnetic resonance (CMR) images, it is important to gain some basic understanding of the underlying physics, hardware, and methods used to generate and encode the images. This chapter provides a brief overview of the fundamental physics of MRI, as well as a summary of some techniques of particular importance in cardiac imaging.

The source of the MRI signal

MRI is based on the phenomenon of nuclear magnetic resonance (NMR). This is a “resonance” phenomenon in that a signal is emitted by the sample after radiofrequency (RF) energy is applied to it. The NMR signal is emitted by molecules of the tissue in the body, unlike X-ray imaging methods which rely on the attenuation of externally applied radiation by tissues or injected contrast agent, or nuclear imaging methods based on the detection of radiation from injected radioisotopes.

The atomic nucleus is comprised of protons and neutrons that have magnetic fields associated with their spin and charge distributions. The resonance phenomenon refers to the ability of some nuclei to selectively absorb and later release energy specific to the element and its chemical environment. Not all elements are capable of resonance; it requires an odd number of protons or neutrons for a nucleus to exhibit a magnetic moment associated with its net spin. The hydrogen atom, for example, consists of a single proton with no neutrons, giving it a net spin = \( \frac{1}{2} \). While there are several biologically relevant candidates for MRI, such as \(^{17}\)O, \(^{19}\)F, \(^{23}\)Na, or \(^{31}\)P, it is the hydrogen (\(^{1}\)H) atom’s large magnetic moment as well as its isotopic abundance and biological abundance that make it the primary choice for MR imaging. Hydrogen is found in water molecules and in the methylene (CH\(_2\)) groups of fat; both highly abundant in living tissue. The magnetic moment of each individual hydrogen proton is small, but the additive effect of many magnetic moment vectors makes it detectable in MRI.

The Larmor equation

Under normal conditions the net magnetization of a tissue sample is zero due to the random orientations of the individual protons or “spins” (Figure 1.1a), but this changes when the sample is placed in a strong magnetic field (Figure 1.1b). The magnetic field strength generated by clinical MRI systems ranges from 0.15 Tesla (T) to 7T, with CMR most commonly performed at 1.5 T. In comparison, the Earth’s magnetic field is approximately .05 mT at the surface of the planet. When subjected to any external magnetic field, nuclear spins will align themselves with the applied field.
either in a low energy state parallel to the field, or a high energy state anti-parallel to the field. Moreover, they will precess around the direction of the magnetic field with a frequency proportional to the field strength, as described by the Larmor equation:

\[ \omega = \gamma B_0 \]

where \( \gamma / 2\pi \) is the gyromagnetic ratio and \( B_0 \) the external magnetic field, which in our case is the main magnetic field generated by the MRI system. The gyromagnetic ratio is unique for each atom. For example, the gyromagnetic ratio of hydrogen is \( 42.58 \text{ MHz/T} \), generating a precessional frequency of approximately \( 64 \text{ MHz} \) at \( 1.5 \text{ T} \).

Externally applied RF energy that matches the Larmor precessional frequency will cause some of the protons in the low energy state to flip to the high energy state. For protons at field strengths used for CMR, the Larmor frequency is in the “very high frequency” or VHF range of radio frequencies commonly used for FM radio and broadcast television. Radiation in this frequency range is non-ionizing, contributing to the inherent safety of MRI in comparison to radiographic techniques. Energy is proportional to frequency, and the RF energy used in MRI is many orders of magnitude lower than X-ray radiation, and is not known to cause the increased risk of DNA damage that is observed with the use of X-rays. Despite its lower energy, the MRI signal is detectable because there are so many hydrogen nuclei available in the body to contribute to the signal.

**The Boltzmann distribution**

From the quantum mechanical point of view, hydrogen spins are found in either one of the two available energy states. However, there are only slightly more spins in the low energy spin state compared to the high energy state, and the excess spin number is, according to Boltzmann equilibrium probability, directly proportional to the total number of spins in the sample and the energy difference between states. The relation between the number of spins in high energy state \( (N^-) \) and the number of spins in the lower energy state \( (N^+) \) is given by the expression:

\[ N^- / N^+ = e^{-E / kT} \]

where \( E \) is the energy difference between the spin states; \( k \) is Boltzmann’s constant, \( 1.3805 \times 10^{-23} \text{ J/K} \); and \( T \) is the temperature in Kelvin. The energy of a proton, \( E \), is directly proportional to its Larmor frequency \( \nu \) (in Hz), such that \( E = h\nu \), where \( h \) is Plank’s constant (\( h = 6.626 \times 10^{-34} \text{ J/s} \)). Following the Larmor equation (above), this results in a direct relationship between proton energy \( E \), and magnetic field \( B_0 \), \( E = h\gamma B_0 \). When the energy delivered to the system matches the energy difference
between the spin states, spins from the stable lower energy states jump up to the unstable higher energy states. As these spins fall back into the lower energy state, they emit a detectable signal; this is the resonance phenomenon.

Only those excess spins in the low energy state are available for excitation, and able to generate MRI signal when they return to equilibrium position. There are only approximately nine more spins in the low energy state compared to the high energy state for each 2 million spins at 1.5 T field strength. Given that each ml of water contains nearly $10^{23}$ hydrogen atoms, the Boltzman distribution predicts over $10^{17}$ spins contributing to the MRI signal in each ml of water. As the magnetic field strength increases, the number of excess spins in low versus high energy state increases, and with it the magnitude of the MRI signal. The larger number of excitable spins leads to improvement in image quality (signal-to-noise ratio [SNR] and/or resolution) and is the driving force for imaging at higher magnetic field strengths like 3 T and 7 T.

The RF pulse and signal reception
When a specimen or subject is placed in the high magnetic field of an MRI system, the number of spins in the low energy level exceeds those at the high energy level (Figure 1.1 b) creating a net magnetization aligned in the direction of $B_0$. Externally applied radio frequency (RF) energy with a frequency that matches the precessional or Larmor frequency of the spins will cause some of the protons in the low energy state to jump up to the high energy level. In terms of the net magnetization, the magnetic field component of the RF wave, $B_1$, which is perpendicular to the direction of $B_0$, and of μT order of magnitude, will tilt the longitudinal magnetization ($M_L$) to an angle that depends on the strength of the applied $B_1$ field and the duration of the RF pulse, which is usually from one to several milliseconds. A 90° RF pulse will rotate the net magnetization totally from the longitudinal plan ($z$) into the transverse plane ($xy$). It is this transverse component of the net magnetization that generates the MR signal detectable by a receiver coil. The MR signal is captured in the form of an induced voltage in a receiver antenna, or “coil”, placed perpendicular on the transverse plane. The precession of the transverse component of the magnetization, $M_{xy}$, generates an oscillating current in the receiver coil according to Faraday’s law of induction.

In summary, signal generation in MRI follows a few basic steps. As spins are subjected to the strong magnetic field, the net magnetization aligns with the direction of the applied field in the longitudinal ($z$) direction. The RF pulse with a frequency that matches the precessional frequency of the protons tilts the net magnetization from the longitudinal to the transverse plane ($xy$). Afterwards, the precession of spins around the axis of the main magnetic field induces a “resonant” signal in a receiver coil placed perpendicular to the transverse plane (Figure 1.2).

Relaxation
Through application of the RF pulse, which provides the energy necessary for spins to jump from the low to the high energy level, the protons are raised up to an excited, unstable state. While the magnitude of the MR signal depends on the net magnetization $M_{xy}$, the duration of the induced voltage is a function of the relaxation time constants $T_1$ and $T_2$, and/or $T_2^*$ of the sample. These relaxation parameters are different for different tissues and pathologies, and as such are primary sources of image contrast in CMR.

$T_2$ and $T_2^*$ relaxation
A RF pulse that tilts the net magnetization into the transverse plane also brings the spins into phase coherence with each other, resulting in a maximum current in the receiver antenna. As time passes, the spins that were initially precessing in phase with each other will lose phase coherence, resulting in a decrease in the net magnetization (Figure 1.2) and induced voltage. The loss of phase coherence is called transverse or spin–spin relaxation and is characterized by the $T_2$ time constant. The rate of loss in phase coherence among the individual protons is influenced by the chemical environment experienced by each. The presence of each spin slightly affects the local magnetic field, and as such the precessional frequency of the surrounding spins. Due to this spin–spin interaction protons will lose phase coherence and the transverse
The basics of cardiac MR

Figure 1.2 Conversion of longitudinal magnetization, $M_z$, into transverse magnetization, $M_{xy}$, by a 90° RF pulse, results in an initial phase coherence of the spins causing magnetization ($M_{xy}$) will decay exponentially from $M_z$ at a rate defined as $T_2$, described by the relationship:

$$M_{xy}(t) = M_z(0) \exp(-t/T_2)$$

The transverse relaxation is highly dependent on the molecular structure of the sample. Small molecules in amorphous medium demonstrate a long $T_2$ because fast and rapidly moving spins average out the intrinsic magnetic field inhomogeneities. Conversely, larger macromolecules that are subject to constrained molecular motion, exhibit much shorter $T_2$ due to the accumulation of phase differences among spins, which are not canceled by rapid diffusion.

The spin–spin interaction is not the only factor responsible for the time-decay of transverse magnetization and acquired MRI signal. Extrinsic magnetic inhomogeneities, such as imperfections of the main magnetic field or susceptibility differences between adjacent tissues, also contribute to dephasing and loss of phase coherence among spins. The time-decay of signal in this case is characterized by the time constant $T_2^*$, which is always shorter than $T_2$. However, the $T_2^*$ signal loss caused by static, extrinsic magnetic field inhomogeneities is corrected in spin echo sequences by the use of 180° RF refocusing pulses. RF refocusing can reverse the phase difference induced by static field inhomogeneity and re-establish phase coherence.

**T1 relaxation**

The end of the RF pulse begins the return to equilibrium. Immediately after the RF pulse, the excited spins will undergo relaxation through the same energy coupling process. The return of excited spins to the low energy, equilibrium state, which is accompanied by the recovery of the longitudinal magnetization ($M_z$), is part of the spin-lattice relaxation process. The rate of $M_z$ recovery is a function of the relaxation time constant $T_1$, which by definition, is the time necessary to recover 63% of the equilibrium magnetization $M_z$ after a 90° RF pulse:

$$M_z(t) = M_z(1 - \exp(-t/T_1))$$

The return to equilibrium is directly related to how fast the excited spins release their energy to the tissue (lattice). This process depends significantly on the physical properties of the tissue. The energy transfer is possible only when the precessional frequency of spins overlaps the vibrational frequencies of the molecules embedded into the lattice. Depending on their physical characteristics (size) the vibrational frequency of the molecules span different frequency ranges. The less efficient this system is at transfer of energy from the excited spins to the lattice, the longer the $T_1$ recovery time will be.

Moreover, $T_1$ relaxation is dependent on the main magnetic field strength. At higher magnetic field the precessional frequencies of spins increase,
and as such there is lower spectral overlap with the molecular vibrational frequency spectrum of the sample, resulting in an increase in spin lattice relaxation time with $B_0$. The only exception from this rule is offered by free water, which has a vibrational frequency range that covers a large spectrum of precessional frequencies. However, for a specific tissue, there is always the following relationship among relaxation times, $T_1 > T_2 > T_2^*$, regardless of the magnetic field strength.

**Contrast agents**

Tissue contrast in MRI is fundamentally based on tissue specific parameters such as proton density (PD) and relaxation times $T_1$, $T_2$ and $T_2^*$ and can be further influenced by diffusion, perfusion, flow and motion. Contrast agents offer another important source of tissue contrast essential to many cardiac imaging techniques. Contrast agents generally work by shortening both $T_1$ and $T_2$, but with a predominant effect on either one or the other depending on the specific agent. $T_1$ shortening contrast agents enhance the MR signal (positive contrast) by increasing the signal in $T_1$ weighted images. The reverse is true for a predominantly $T_2$ contrast agent; shortened $T_2$ leads to decreased signal (negative contrast) in $T_2$ weighted images.

While the underlying processes by which contrast agents function is complex, their effects on $T_1$ and $T_2$ can generally be described in a simplified way by the equations:

$$\frac{1}{T_1} = \frac{1}{T_1^0} + r_1 C$$
$$\frac{1}{T_2} = \frac{1}{T_2^0} + r_2 C$$

where $T_1$ and $T_2$ are the tissue relaxation times after contrast agent administration, $T_1^0$ and $T_2^0$ are the relaxation times prior to contrast agent injection, $C$ is the contrast agent concentration and $r_1$ and $r_2$ are the longitudinal and transverse relaxivities of the contrast agent. However, $r_1$ and $r_2$ are field strength dependent and the linear relationship between relaxation time shortening and contrast agent concentration is no longer valid at high concentrations.

Most contrast agents used for clinical CMR are the paramagnetic chelates of gadolinium (Gd$^{3+}$). Gadolinium has unpaired orbital electron spins and a very large magnetic moment. Gadolinium shortens the $T_1$ relaxation time by allowing free protons to become bound and to create a hydration layer, which helps energy release from excited spins and accelerates the return to equilibrium magnetization. A number of CMR applications are dependent on exogenous contrast agents, including angiography, first-pass perfusion, late gadolinium enhancement (LGE), and characterization of tumors and masses.

**Image encoding**

**Magnetic field gradients**

The Larmor equation is at the heart of image encoding. The main magnetic field, $B_0$, generated by the MRI system is engineered to be as homogeneous as possible. Homogeneity of about 1 part per million over a roughly spherical region of $\frac{1}{2}$ meter in diameter is typically achieved, depending on the particular magnet design. Within this homogeneous volume, all protons precess at the same frequency (disregarding tissue susceptibility differences and other sources of field distortion). By precisely controlling the strength of the magnetic field as a function of both location and time, the frequency and phase of precession also become functions of location and time. Using this principle, the MR signals coming from different locations within the body can be distinguished from one another, and an image can be formed. Special gradient coils are embedded within the bore of the MRI system to create controlled, linear variations in the $B_0$ field strength in each of the three orthogonal directions in the Cartesian $(x,y,z)$ spatial coordinate system (Figure 1.3). By applying current to these coils simultaneously in appropriate ratios, a linear gradient in the magnetic field can be generated in any arbitrary direction. This linear change in magnetic field translates into a linear change in resonant frequency depending on location in that direction.

**Slice selection**

In order to generate a magnetic resonance signal that can be detected, the magnetization must be
tipped away from the longitudinal axis and into the transverse plane by RF excitation, as described earlier. The process of slice selection limits RF excitation to a plane of tissue of any desired thickness. Recall that a spatially linear gradient or ramp in the magnetic field establishes a linear relationship between proton precessional frequency and location. In order to excite or tip the magnetization of precessing spins, an RF pulse must oscillate at the precessional frequency of those spins. Physically, RF pulses are of finite amplitude, duration, and bandwidth. The amplitude and duration of the RF pulse will control the resulting flip angle; the longer the pulse and higher the amplitude, the greater the tip angle of the net magnetization. The RF pulse center frequency can be shifted to match a specific location along the gradient, and the bandwidth of the pulse can be limited to selectively excite the protons with a narrow range of frequencies around the center frequency, as shown in Figure 1.4. Thus a slice of arbitrary thickness and location along the direction of the slice select gradient can be selectively excited to generate the signal used to form the MR image. The direction of the slice selection gradient, and therefore the orientation of the slice, can also be arbitrarily chosen by appropriate combination of the physical x, y, and z gradient fields. Following slice selective excitation, the signal detected by the MRI receiver coil will come from the excited slice only. The amplitude of the signal emitted by the slice is directly proportional to its thickness; this sets the lower practical limit on slice thickness at about 2 mm. Thinner slices can be achieved by 3D encoding, which is addressed in the section on phase encoding.

**Frequency encoding**
The process of slice selection excites the slice or slab of tissue that will generate the MR signal; the next steps of frequency and phase encoding serve to encode this tissue into individual discrete two-dimensional picture elements (pixels), or three-dimensional volume elements (voxels). Once again, linear field gradients and the Larmor relationship between field strength and precessional frequency are used to encode spatial location information into the MRI signal. After a slice-selective RF pulse tips the magnetization into the transverse plane, an MR signal is emitted by all of the spins contained within the slice and some method of encoding is required to distinguish the signals coming from the individual voxels. A linear magnetic field gradient is switched on in one of the in-plane directions, perpendicular to the slice select gradient. This gradient has the effect of frequency encoding. While this gradient is on, preces-
sional frequency has a linear distribution along the gradient direction, and thus every location along the gradient can be distinguished by the frequency of the signal it emits. The MR signal is detected through the receiver coils and digitally sampled using an analog-to-digital converter (ADC) during the application of a constant frequency encoding gradient. This detected signal is the sum of all of these frequency components. Fourier transform is used to separate out the individual frequency components in the detected signal, and thus decode the signal from the entire slice into individual signals coming from discrete locations along the frequency encoding gradient.

Frequency encoding can also be described in terms of spatial frequency, and this alternative description is also helpful to understand phase-encoding, the method used to encode the other in-plane dimension of the image. Spatial frequency expressed as cycles/cm is directly analogous to the perhaps more familiar concept of temporal frequency expressed in units of cycles/sec or Herz. Whereas temporal frequency pertains to a time varying signal, spatial frequency can be used to describe a signal varying with position, for example, an image. An individual spatial frequency then describes a sinusoidal variation in pixel intensity across an image. A complex image can be expressed as the linear combination of many spatial frequencies. Lower spatial frequencies determine the gross features and contrast in the image, while higher spatial frequencies determine image details and sharpness. The Fourier transform can be used to go back and forth between image space and spatial frequency, or “k-space”, in the same manner it is used to describe the frequency component of a time-varying signal.

Before the frequency encode gradient is switched on, all of the spins are precessing at the same frequency and in phase with each other. As soon as the frequency gradient is switched on, the spins begin to precess at a frequency linearly dependent on position, and this linear distribution of frequencies causes a sinusoidal distribution of phase across the slice in the direction of the frequency encoding gradient (Figure 1.5). It is the integral or

![Figure 1.4](image-url)
accumulated area under the frequency encoding gradient pulse that determines the instantaneous sinusoidal distribution of phase. This sinusoid across space describes a single spatial frequency, often referred to as $k_x$. As time progresses while the gradient remains on, the area under the frequency encoding gradient increases and progressively higher spatial frequencies are mapped out in the distribution of phase. Thus, each digital sample of the MR signal corresponds to the signal attributable to a unique spatial frequency component in the image, that is, each sample corresponds with a distinct value of $k_x$.

**Phase encoding**

The third spatial dimension (second in-plane dimension) must also be encoded in order to distinguish the signal from each individual voxel and complete the process of image formation. Phase encoding is also based on the Larmor equation, and on discrete sampling of the spatial frequency content of the image. Frequency encoding, as described earlier, samples spatial frequency components in rapid succession as the area under the frequency encoding gradient pulse accumulates over time. Phase encoding, however, is instead typically accomplished by applying a series of gradient pulses of successively increasing amplitude, each designed to encode a single specific spatial frequency component, $k_y$, of the image (Figure 1.6). The phase encode gradient pulse amplitude is incremented to encode a different spatial frequency component $k_y$ prior to each frequency encoding gradient. This completes the concept of two-dimensional spatial frequency encoding. The matrix of sampled image data represents the two-dimensional spatial frequency content of the image and is often referred to as $k$-space, and the process of image data acquisition can be thought of as filling of $k$-space. Each phase encoded line of data corresponds to a specific spatial frequency in the phase encoded direction, and contains all spatial frequencies in the frequency encoding direction. With the sampling of each MR signal, all values of $k_x$ are encoded for a single value of $k_y$. Thus, each phase-encoded line of data corresponds to a raster line in $k$-space. The two-dimensional Fourier transform is utilized to convert this spatial frequency information into the image domain (Figure 1.7). Phase-encoding can also be applied in the slice direction to encode thinner sections of tissue than possible using selective excitation alone. 3D data acquisition incorporates the process of phase encoding into the slice direction as well as one in-plane direction.

**Basic pulse sequences**

The pulse sequence defines the sequence of events on a microsecond scale controlling all factors
The understanding of pulse sequences can be gained by looking at the five basic components of any CMR pulse sequence: magnetization preparation, echo formation, k-space trajectory, k-space segmentation, and image reconstruction. Some of the options used in CMR within each of these categories are listed in Table 1.1.

**Magnetization preparation**

Magnetization preparation refers to the various methods available to impart sensitivity of the pulse sequence to specific characteristics of the tissue within the imaged slice. For example, the inversion
The basics of cardiac MR

For delayed-enhancement imaging post-contrast, since time is available in between IR pulses to allow for signal recovery, the SR pulse is more appropriate for perfusion imaging when multiple slices are prepared and imaged each cardiac cycle. The double-inversion or black-blood preparation is commonly used to suppress the blood signal \[1,2\]. It works by effectively inverting the blood outside of the imaged slice, without effecting the magnetization within the imaged slice. When the inverted blood flows into the slice, the pulse sequence can

Table 1.1 CMR pulse sequences can be broken down into the components listed in the five columns of the table. Each application involves a different combination of these components to achieve specific imaging goals.

<table>
<thead>
<tr>
<th>CMR pulse sequences</th>
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</thead>
<tbody>
<tr>
<td><strong>Magnetization preparation</strong></td>
</tr>
<tr>
<td>• Inversion recovery</td>
</tr>
<tr>
<td>• Saturation recovery</td>
</tr>
<tr>
<td>• Double inversion recovery (black blood)</td>
</tr>
<tr>
<td>• Fat suppression</td>
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<tr>
<td>• T2-preparation</td>
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<tr>
<td>• Diffusion weighting</td>
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<tr>
<td>• Tagging</td>
</tr>
<tr>
<td>• DENSE</td>
</tr>
<tr>
<td>• Magnetization transfer</td>
</tr>
<tr>
<td>• Velocity encoding</td>
</tr>
<tr>
<td>• T1, T2, T2* mapping</td>
</tr>
<tr>
<td><strong>Echo formation</strong></td>
</tr>
<tr>
<td>• Spin echo</td>
</tr>
<tr>
<td>• Turbo Spin Echo</td>
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<tr>
<td>• SPACE</td>
</tr>
<tr>
<td>• Gradient Echo</td>
</tr>
<tr>
<td>• Steady-State Free Precession (SSFP)</td>
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<tr>
<td>• Echo Planar Imaging (EPI)</td>
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<tr>
<td>• Spin echo EPI</td>
</tr>
<tr>
<td>• Gradient echo EPI</td>
</tr>
<tr>
<td><strong>k-space trajectory</strong></td>
</tr>
<tr>
<td>• Cartesian</td>
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<tr>
<td>• Linear</td>
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<tr>
<td>• Centric</td>
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<tr>
<td>• Spiral</td>
</tr>
<tr>
<td>• Radial</td>
</tr>
<tr>
<td>• PROPELLER</td>
</tr>
<tr>
<td><strong>k-space segmentation</strong></td>
</tr>
<tr>
<td>• Non-segmented</td>
</tr>
<tr>
<td>• Segmented</td>
</tr>
<tr>
<td>• Single-shot</td>
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<tr>
<td><strong>Image reconstruction</strong></td>
</tr>
<tr>
<td>• Partial Fourier</td>
</tr>
<tr>
<td>• Parallel Imaging</td>
</tr>
<tr>
<td>• SENSE</td>
</tr>
<tr>
<td>• SMASH</td>
</tr>
<tr>
<td>• GRAPPA</td>
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<tr>
<td>• TSENSE/TGRAPPA</td>
</tr>
<tr>
<td>• k-t methods</td>
</tr>
</tbody>
</table>

Figure 1.8 The inversion (a) and saturation (b) recovery magnetization curves of fat, myocardium and blood. Observe the increased magnetization difference, resulting in better T1 contrast between different tissue types when inversion recovery sequence is used as compared with saturation recovery technique.

recovery (IR) technique precedes data acquisition with a 180° RF pulse to provide high sensitivity to differences in T1 (see Figure 1.8). Saturation recovery (SR) preparation using a 90° RF pulse is commonly used in first-pass perfusion imaging. It provides moderate T1-weighting that is not as strong as the IR technique, but does not require a wait period in between pulses to allow magnetization to recover since longitudinal magnetization is essentially set back to zero with each saturation pulse. While IR imaging has been very successful for delayed-enhancement imaging post-contrast, since time is available in between IR pulses to allow for signal recovery, the SR pulse is more appropriate for perfusion imaging when multiple slices are prepared and imaged each cardiac cycle. The double-inversion or black-blood preparation is commonly used to suppress the blood signal \[1,2\]. It works by effectively inverting the blood outside of the imaged slice, without effecting the magnetization within the imaged slice. When the inverted blood flows into the slice, the pulse sequence can
be timed to acquire data just as the blood signal is crossing through the zero or null point in its T1 recovery curve. Velocity encoding can also be considered as a magnetization preparation, although unlike the others listed in the table it incorporates specific gradient pulse design to control the motion and flow sensitivity of the pulse sequence rather than a separate preparation module [3].

Echo formation
A few basic methods of echo formation are employed in CMR, each with distinct advantages and disadvantages that have led to specific applications for each. Spin Echo, including Turbo- or Fast Spin Echo and SPACE [4], is used primarily as a black-blood method for cardiovascular morphology and tissue characterization based on T1 or T2 changes. Gradient Echo is commonly used for delayed-enhancement and first-pass perfusion, in combination with IR and SR prep pulses, respectively. Gradient echo cine is used for phase velocity mapping, tagging, and in some cases for visualization of valve function. Most cine imaging applications, however utilize steady-state free precession [5] (SSFP or TrueFISP), due to its inherently high blood-to-myocardium contrast, high signal-to-noise ratio, and high imaging efficiency [6]. Echo Planar Imaging (EPI) has found widespread application as a method for perfusion imaging due its high efficiency [7].

k-space trajectory
CMR applications are dominated by Cartesian k-space sampling, that is, a linear raster scanning of k-space accomplished by conventional phase encoding each line of data. Virtually all CMR sequences in broad clinical use employ this conventional sampling strategy. Some alternative trajectories that have been employed in CMR are shown in Figure 1.9. The spiral trajectory has some advantages in speed and insensitivity to motion, and has been utilized for coronary artery imaging [8]; however it is highly sensitive to field inhomogeneities, and has not seen widespread application for that reason. Radial imaging has some efficiency advantages over Cartesian sampling, and is experiencing some gain in popularity for cine imaging due to its ability to achieve higher spatial resolution for a given number of acquired lines. PROPELLER [10] combines some of the advantages of Cartesian and radial acquisitions, and while it is popular for neuro-imaging, CMR applications have not yet been fully developed.

k-space segmentation
Segmentation refers to the strategy of segmenting data acquisition over multiple cardiac cycles [11]. The degree of segmentation can range from one k-space line per cycle (non-segmented), up to acquisition of all of the lines needed to reconstruct an image (single-shot). Any level of segmentation can be used with each of the basic methods of echo formation (gradient echo, spin echo, SSFP, EPI). Overall acquisition time is inversely related to the number of lines acquired per image each cardiac cycle (lines per segment), that is, the more lines per segment, the shorter the scan time. The trade-off is in temporal resolution; the more lines per segment, the poorer the temporal resolution. Modern CMR sequences for cine, flow, and delayed-enhancement are designed to acquire enough lines per segment to reduce the scan time to a reasonable breath-hold. The success of segmented imaging depends not only on patient breath-hold, but also on a
regular cardiac rhythm to ensure that the k-space data from each cardiac cycle is capturing the heart in the same respiratory and cardiac positions. In patients with severe arrhythmia or an inability to breath-hold, real-time or single-shot methods are commonly used due to their insensitivity to respiratory motion effects.

**Image reconstruction**

Partial Fourier or partial k-space acquisition has been used for a number of years as a means of reducing scan time at the expense of signal-to-noise [12]. More recent advances in image reconstruction methods have played a large part in the improved image quality and efficiency of CMR. Parallel Acquisition Techniques (SENSE [13], SMASH [14], GRAPPA [15], and TSENSE [16]) have become an integral part of virtually all commonly applied CMR pulse sequences. These methods allow reconstruction of full field-of-view and full resolution images while sampling only a fraction of the full k-space data matrix. This results in a significant time savings that can be directly beneficial as shortened scan time, or traded for higher spatial or temporal resolution. This entails a direct trade-off of signal-to-noise ratio, so the acceleration attainable using parallel imaging is generally limited to a factor of two or three, but that can make the difference between a 10 second or a 20 second breath-hold, and so represents a significant gain in imaging performance.

Table 1.1 lists some of the many possibilities for each of these pulse sequence components. Elements from each column of the table can be combined to construct pulse sequence variations for specific applications. For example, double inversion recovery, turbo spin echo, and segmented, linear, Cartesian k-space trajectory with GRAPPA parallel imaging reconstruction is a common application for T2-weighted imaging of the heart. Or, saturation recovery, gradient echo EPI with centric, Cartesian, single-shot trajectory and TSENSE reconstruction [17] is commonly used for first-pass perfusion imaging. Delayed-enhancement imaging is commonly performed using inversion recovery, gradient echo, with linear, Cartesian, segmented k-space acquisition [18]. The list of possibilities goes on, and while there is not space in this chapter to delve into the details of the numerous combinations in common use in CMR, hopefully Table 1.1 helps to illustrate the wide range of sequence combinations available, and even some possibilities that have not yet been investigated. Most modern MR systems have very flexible interfaces and pulse sequence control software that allow the user to easily mix and match components from these categories. Unfortunately, for every useful combination there are many more that have no value, contributing to the complexity of CMR and the need for every CMR practitioner to gain some basic understanding of the underlying physics.

**Cardiac and respiratory synchronization**

**ECG triggering and gating**

In addition to the categories outlined in Table 1.1, CMR sequences can be further subdivided with respect to depiction of cardiac motion: dynamic or static. Dynamic methods include any cine techniques designed to represent the heart or flow patterns at multiple phases throughout the cardiac cycle. Static methods generate images of the heart at a single-phase of the cardiac cycle. Static techniques are generally applied to depict cardiovascular anatomy, or to characterize tissue by generating images sensitive to any of a variety of contrast mechanisms. In either case, synchronization with cardiac motion, generally using an ECG signal, is necessary to time each image to a specific phase of the cardiac cycle. This is not the case in real-time cine methods that acquire dynamic images asynchronous with the cardiac cycle. Some sequences, like first-pass perfusion and dynamic 3D MR angiography, can be thought of as hybrids between dynamic and static imaging. These techniques create a dynamic series of images, but each image depicts a different cardiac cycle, not a different phase of the cardiac cycle.

The R-wave of the ECG is typically used to generate a trigger signal indicating the beginning of the cardiac cycle with the initiation of ventricular systole. CMR pulse sequence events are timed relative to that trigger to acquire data at specific time points within the cardiac cycle. Static images are acquired using prospective triggering. That is, one or more lines of k-space data are acquired beginning at a specific time-delay relative to the trigger
pulse. Dynamic cine images can be acquired by either prospective triggering or retrospective gating. With prospective triggering, phases of the cardiac cycle are defined by a fixed time after the R-wave, regardless of the duration of each individual cardiac cycle. With retrospective gating, each cardiac phase is defined as a certain percentage of the cardiac cycle, allowing the actual duration of each phase to vary flexibly with variation in cardiac cycle.

**Respiratory motion compensation**

Respiration causes variation in the position of the heart from beat to beat, and leads to motion artifact in segmented acquisitions scanned over multiple cardiac cycles. There are four basic strategies to deal with respiratory motion artifact: signal averaging, breath-holding, respiratory gating, and single-shot imaging. In many common CMR applications like cine, velocity mapping, late gadolinium enhancement, and black-blood imaging, segmented acquisitions that are fast enough to be performed in a reasonably short breath-hold are widely available and commonly used as the standard method. In small children unable to voluntarily breath-hold, signal averaging is successfully used to average out respiratory motion artifact. Advances in gradient hardware continue to drive advances in pulse sequences. This is especially true in CMR where acquisition speed is critical to avoid the deleterious effects of motion. Gradient amplitude and switching speeds have hit the limits of physiological stimulation. RF receiver systems continue to advance in the number of channels used to increase the capacity of parallel imaging techniques. Multi-channel array coils designed specifically for cardiovascular applications can improve SNR and parallel imaging performance.

**References**


**MRI hardware**

The main field-generating components of the MRI system include the main magnet (B₀ or B₁ field), the RF transmitter coil (B₁ field), and the gradient coils (Gₓ, Gᵧ, Gₚ fields). Additional second-order shim coils are also often employed to achieve a more homogeneous B₀ field. Separate computer systems are typically used to control the MRI field-generating units (measurement-control system), reconstruct the acquired data, and provide an interactive interface to link the user to the MRI system control and the reconstructed images.

Advances in hardware continue to drive advances in pulse sequences. This is especially true in CMR where acquisition speed is critical to avoid the deleterious effects of motion. Gradient amplitude and switching speeds have hit the limits of physiological stimulation. RF receiver systems continue to advance in the number of channels used to increase the capacity of parallel imaging techniques. Multi-channel array coils designed specifically for cardiovascular applications can improve SNR and parallel imaging performance.
CHAPTER 2

Technical aspects of pediatric cardiac MR

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Introduction
The application of cardiac magnetic resonance (MR) imaging to pediatric patients with heart disease has been a significant advance in care over the last two decades. However, for a successful study, important changes have to be made in the way that scans are performed relative to adults. The age at which a patient ceases to be pediatric and becomes an adult is generally accepted to be in the mid to late teens. In practice, the overwhelming majority of teenagers with normal intellectual and emotional development can be imaged successfully using techniques suitable for adults, and therefore the contents of this chapter apply principally to patients aged 12 years or less.

The changes required encompass the environment of the scanner suite, the set-up, the use of sedation or anesthesia in patients unable to cooperate, and the need for additional physiological monitoring in such cases. The smaller body habitus, faster heart rates, and reduced circulation times of smaller children require alterations in the sequences used for imaging. Artifacts from certain implanted devices used almost exclusively in pediatric cardiac patients can necessitate changes in study protocols in individual patients.

The scanner suite
MRI in pediatric cardiac patients involves the imaging of structures only a few millimeters diameter, necessitating excellent spatial resolution. Such image acquisition needs to be gated to heart rates of perhaps 150 beats per minute in neonates and small infants. This poses great demands on the MR scanner in terms of gradient strength and the speed at which these gradients can be manipulated. Advances in electrical engineering and field homogeneity in more modern MR scanners allow for improved image quality and shorter scan times, and the use of such modern scanners should be considered mandatory for pediatric cardiac cases. Those facilities fortunate to have access to multiple MR scanners of differing capabilities should schedule pediatric cardiac cases on the scanner with the highest imaging performance.

Children have a reduced ability to control their body temperature, especially neonates and small infants, who are particularly susceptible to cold. Many adult MR scanner facilities are deliberately kept at a temperature far too cold for a neonate to tolerate for any length of time, and special care should be taken to keep such patients warm at all times. This concern extends to the area outside the scan room itself. Any significant prolongation of the anesthesia induction or IV access in a small child carries a significant risk of hypothermia.
Lighting in the scan room should be of good quality, especially over the patient table area, so that assessment of the patient can be rapidly performed in the room. It should also be capable of being dimmed, to facilitate sleep in infants or sedated patients as required.

A suitable décor in the scan room and associated areas can be very helpful for the older pediatric patients who may be apprehensive at the prospect of a study while awake. Pediatric hospital facilities can give consideration to colorful wall murals and the concealment of unneeded accessory equipment from the patient.

**Coil selection**

Coil selection should be appropriate to maximize the signal to noise ratio. Neonates and very small infants may fit entirely within an adult knee or head coil, and the reduced distance between the coil and the patient can compensate for the reduction in image quality inherent in the use of a coil not specifically designed for cardiac or body imaging. In older infants, a flexible coil, or the full adult body or cardiac coil should be used. The weight of an adult coil is too great to be tolerated by a small child; it should be elevated on foam pads on either side of the patient to sit just clear of the anterior thoracic wall. This technique can also be used to reduce artifact caused by motion of the coil in a patient with a marked precordial heave, irrespective of the age of the patient.

Coil elements that are not contributing to an individual image should be switched off to reduce image noise.

**Anesthesia and sedation**

The degree of co-operation required of the patient in a cardiac MR scan is greater than that required for any other type of MR scan; cardiac scans can be lengthy, require no significant movement and repeated breath-holds at the same point of the respiratory cycle over a period of perhaps 45 minutes to an hour. The patient is asked to do this in a very strange environment which many adult patients find intimidating. The use of either sedation or general anesthesia is required, therefore, to allow for performance of cardiac studies in those children too young to co-operate adequately. There is an important difference between the two techniques in that sedated patients continue to breathe throughout, and image acquisition has to be altered substantially as a result, whereas a paralyzed and ventilated patient under anesthesia can effectively “breath-hold” by the simple expedient of asking the anesthesiologist to temporarily cease ventilation.

There is no definitive cut-off age below which children will be unable to co-operate adequately to allow for a successful cardiac MR study, but in general, most children aged 10 can do so. The younger the child is below this age, the less likely it is that the scan will be completed successfully, although a limited study with much reduced time in the scanner may be possible in a child as young as 7. Prior preparation of the child outside the scan room is important; the input of an informed play therapist, or the presence in the scan room of a supportive parent or other regular caregiver can reduce patient anxiety.

The environment of the MR scan room is a challenging one for the anesthesiologist or the pediatrician/nurse sedation specialist. The patient will be contained mostly within the scanner and direct visual inspection during the study is not possible without retrieval of the patient from the bore and removal of the coil. A number of centers have moved to video monitoring with MRI compatible cameras placed in critical positions to visualize the patient. Extensive physiological monitoring of subjects with reduced levels of consciousness using equipment specifically designed to operate in an MR scan room is mandatory. Limb-lead ECG, pulse oxymetry, and blood pressure must all be monitored, inspiratory and expiratory gas analysis employed in anesthetized patients, and temperature monitoring should also be available. ECG leads may have to be positioned in unusual places to obtain a good trace if the heart is in an unusual position (e.g., dextrocardia), or if the intracardiac anatomy is considerably distorted because of the underlying congenital lesion (e.g., univentricular hearts, severely dilated RV, severely hypoplastic LV).
CHAPTER 2 Technical aspects of pediatric cardiac MR

Monitoring outputs should be available wherever the anesthesiology/sedation team is positioned during the study as well as in the scanner control room with careful attention paid as to which is the master and which the slave monitors. There should be direct verbal communication between the anesthesiology/sedation and imaging teams at all times, and preferably direct visual contact also.

The ideal location of the anesthesia equipment within the MR scanner facility is a matter of some debate. Special anesthetic machines designed to operate within the MR scan room are available though at considerable increase in cost compared to standard equipment and anesthetic gases may be supplied in non-ferrous cylinders that can be taken safely into such environments. However, such a set-up inevitably increases the risk that non-MR compatible components or gas cylinders will be inadvertently introduced into the MR scan room, either as part of routine maintenance or during a sudden breakdown, with potentially disastrous consequences for a patient or staff members [1,2]. For this reason, many facilities prefer to position the anesthetic equipment directly outside the scan room, with the gas lines passing through wave guides installed for this purpose. This arrangement has a much reduced risk of inadvertent introduction of unsafe equipment into the scan room, and also makes the communication between the anesthesiology and imaging teams much easier, though at the cost of increased compliance in the anesthetic circuit.

There is usually a minimum distance that monitoring and anesthetic equipment designed to operate in an MR scan room must be kept from the magnet, within which it may not operate correctly, and might even be attracted into the scanner bore. Careful establishment of this distance from the manufacturer is mandatory before the equipment is first introduced into the scan room [3]. Consideration should be given to the use of physical restraints to prevent incursion of the equipment within such a distance, and thus avoid accidents [4].

The staffing of the anesthesiology team should also be considered. MR facility staff are usually very careful as to the safety procedures required to work without incident in the presence of a high magnetic field environment, but anesthesiology staff usually do not experience such in their training; it is preferable, therefore, that a small staff of anesthesia caregivers who work in the facility on a frequent basis be used. Pediatric cardiac patients should always be sedated or anesthetized by staff specifically trained to do such because of the complex cardiorespiratory physiology that can be present. Irrespective of whether the patient is merely sedated or fully anesthetized, a staff member with appropriate training should be assigned the sole task of monitoring the patient throughout the study; this person should have no role in image acquisition.

It is customary to commence anesthesia or sedation in the MR facility, and to remove the patient to an appropriate recovery area within or without the facility when the study is complete. However, on occasions when the induction of anesthesia is thought likely to be more hazardous than normal, or where difficulties in intubation are anticipated, then consideration should be given to commencement of anesthesia in an operating theatre suite or intensive care unit, with transfer of the patient to the MR facility once stable anesthesia has been achieved. Patients who require ongoing infusion therapy during their study are best served by addition of lengthy microbore tubing extensions to their IV lines, allowing the infusion pump to remain outside the scan room during the study, with passage of the tubing through a wave guide to maintain the infusion during the scan. Extension of the tubing is best done in the ward prior to the study, when time is not at such a premium.

Neonates and very small infants may be successfully studied while asleep, and such a state can be achieved by careful attention to feeding at an appropriate time prior to the study (usually 20–30 minutes), and care to ensure that the child is kept in a quiet and dimly-lit environment for some time before the study commences. Oral sucrose at 0.2 ml/kg, given on a pacifier or via syringe, can also be used to calm the baby. The use of swaddling and commercially-available vacuum-shaped support bags etc. also helps to
reduce patient motion. Imaging sequences that allow for continuous breathing throughout must be used.

The decision as to whether to use sedation or anesthesia to perform a cardiac MR scan is one which requires consideration of a number of factors, including the age of the patient, the amount of the information that must be acquired and thus the likely length of the study, local availability of MR scanner time, anesthesiology staff and funding for such along with the availability of specialized sedation teams which include nurses and pediatricians. A failed study is a poor use of inevitably limited healthcare resources, and, if the child is distressed by the experience, may have long-term consequences in terms of future MR scans for that patient (many pediatric cardiac patients will need repeated imaging over their lifetimes). The patient and caregiver may have traveled long distances to a specialized hospital for the study, with considerable disruption to family life, and a further trip to repeat a failed study under anesthetic is wasteful for the family also.

The choice between anesthesia and sedation is a matter for individual institutional and patient preference. Anesthesia is more controllable in terms of the onset, duration, and depth of impaired consciousness, and thus has an advantage in scheduling when MR scanner access is at a premium. Sedation use has been associated with reduced image quality in some studies [5] but not in others [6]; and in some institutions, is far more likely to fail than anesthesia [5], though failure rates can be reduced to close to zero [6] by careful use of expert personnel and strict sedation regimes [6–9]. Imaging times for studies performed under anesthesia can be shorter in theory because of the ability to “breath-hold” the patient as required for image acquisition; in practice, the scanning time difference is marginal at best. Anesthesia is also probably marginally safer than sedation. Though there is a lack of data to support this assertion, it is widely believed that anesthesia is safer than sedation for percutaneous cardiac intervention, with sedation being used for small children and anesthesia for older children and adults. In general, sedation is used for patients who require a more controlled environment, while anesthesia is used for patients who require a more controlled level of consciousness.

Contrast agents

Gadolinium (Gd)-based contrast agents may be used in pediatric cardiac MR studies for MR angiography, assessment of a cardiac mass, or for a delayed enhancement study for a variety of applications including possible cardiomyopathy and myocarditis, assessment of myocardial perfusion or infarction etc. Contrast dose should be adjusted for body mass, with a maximum dose of 0.2 mmol/kg for angiography in those where there is marked admixing of the pulmonary and systemic circulations.

The use of a double-barrel injector pump is fairly standard in MR angiography, but not all pumps available are suitable for such work in neonates and small infants. Some injector pumps can only deliver a minimum dose of 1.0 ml and for very small infants, this may exceed the maximal recommended dose. If the preferred method of angiography timing is to perform a timing sequence while a small test bolus of contrast is administered, the test bolus alone may represent a maximal dose if such a pump is used. A pump with a minimum contrast delivery of 0.1 ml, increasing in 0.1 ml increments, should be used.

Children are susceptible to all the usual side-effects of Gd contrast agents, including that of nephrogenic systemic fibrosis (NSF), a recently described complication of Gd administration in patients with impaired renal function, with an incidence in adults estimated at 3–5% in those with a GFR of less than 30 ml/min/1.73 m² [16,17]. NSF has not been reported in children <6 years of age. This is a severe and usually progressive condition in which the patient develops characteristic fibrotic skin lesions, with subsequent involvement of the muscles and joints, and then other organs such as the lungs, myocardium, liver, testes, diaphragm etc. It results frequently in severe
disability, and may be fatal. NSF has been reported as a complication of all Gd contrast agents available at present, but appears to be more frequent with the use of gadodiamide (Omniscan), gadopentetic acid (Magnevist) or gadoversetamide (Optimarck). Infants under the age of one year have a reduced GFR compared to older children and adults, and theoretically, therefore, are at increased risk of this particular complication, though this is not established as yet. Careful consideration should be given as to whether Gd contrast is absolutely necessary in children under the age of 1, or those with impairment of renal or hepatic function. Once again, this is theoretical since no report of NSF in children <6 years of age has been reported. A half-dose of contrast may be considered sufficient, and some authorities recommend that those contrast agents that appear to have a higher risk of causing NSF should only be used in infants under 1 year of age after careful consideration [18].

**Respiratory motion**

Respiratory motion can be eliminated in a number of ways. Breath-holding is used in awake children, and in those under anesthesia who are paralyzed. An alternative in children under sedation, or while asleep, is to perform imaging with multiple repetitions or averages (usually 3–4) and hence reduce the respiratory motion artifact with this “averaging.” Such images can be excellent quality, however, institutions not familiar with the technique may not obtain image quality as good as those performed with breath-holding, and there is little additional benefit in increasing the repetitions beyond 3–4, but they can be of reasonable quality (Figure 2.1a–b). Image acquisition is prolonged proportionally to the increase in the number of repetitions. The amount of diaphragmatic excursion has a significant effect on image quality, and this technique works best in very small infants only – even by 9 months of age, in certain institutions, there is a significant deterioration in image quality obtained by this means (Figure 2.1c). It should be noted that imaging acquired using the “averaging” technique is more representative of the overall function of the heart (e.g., ventricular function, measuring flow in the great vessels) since it averages all this data over inspiration as well as expiration; breath-holding is obviously not physiologic.

An alternative is the use of navigator gating, in which the diaphragm position is tracked while image data is acquired continuously, but image reconstruction uses only data acquired when the diaphragm is at a particular pre-determined position. The dome of the right hemi-diaphragm is tracked usually, and data acquired at end-expiration used, thus minimizing the time needed for image acquisition because this is the point at which the diaphragm is positioned most frequently (Figure 2.2). Navigator-gating is reasonably robust, but at the cost of fairly lengthy acquisition times. It is used mainly for 3D imaging of the coronary arteries, useful in patients with anomalous coronary anatomy, conditions such as Kawasaki’s arteritis which involve the coronary arteries, or those who have had re-implantation of the coronary origins as part of a surgical procedure, such as an arterial

**ECG signal**

The ECG signal has been replaced by the use of vectorcardiography (VCG), which is less susceptible to distortion from flowing blood in the thoracic aorta acting as a conductor [19]. This can be transmitted to the imaging computer in a variety of ways; increasingly wireless transmission is used. Alternatively, the ECG signal from the anesthetic monitoring equipment can be used to generate a pulse signal contemporaneous with the R-wave to the MR scanner. Peripheral pulse gating may also be used, but requires a good peripheral circulation, and will often be unsuccessful in small children, especially if they are exposed to cold. Peripheral pulse gating differs from the VCG in that the pulse waveform is delayed by 200–300 ms after the R-wave, and sequence timing for prospectively gated sequences must be adjusted as a consequence. Peripheral pulse gating is especially useful in patients with many ectopic beats, because the ectopics do not produce a peripheral pulse, and are therefore ignored by the scanner. If even this fails, then the use of non-triggered steady state free precession (e.g., true FISP) sequences allows for some information to be acquired.
switch operation for transposition of the great arteries.

**Sequence adjustment**

The smaller size of anatomical structures in infants and neonates requires some adjustment of image factors in order to optimize image quality (Figure 2.3). Reduction of slice thickness reduces the SNR significantly, and 3 mm thickness is recommended as a minimum. Spatial resolution should ideally be as high as possible, but SNR may also be reduced substantially if the field of view is too small. The use of over-sampling rather than an increase in the field of view helps to keep the spatial resolution high while avoiding wrap artifacts, though the length of image acquisition is increased. Increasing the number of image repetitions can partly compensate for a reduced SNR, but at the expense of a proportional increase in acquisition time. Keeping the bandwidth as low as possible has a tradeoff but will increase signal as well.

The timing of sequences may be affected by the faster heart rates in very young children. Those sequences that are retrospectively gated, such as steady state free precession (SSFP) cine sequences, have a true temporal resolution that is determined by the length of the cardiac cycle, and the TR. Temporal resolution can be improved by minimizing the TR, and by reduction of the number of lines of k-space to be acquired in each cardiac cycle.
CHAPTER 2 Technical aspects of pediatric cardiac MR

Figure 2.1 Continued (c) The same child as in (b), but with the child being ventilated rather than breath-held, and using a free-breathing sequence with five repetitions; note the reduced quality compared with (a) because of increased diaphragmatic excursion in a larger child.

Figure 2.2 A track of the position of the right hemidiaphragm over time during a navigator-gated acquisition to image the coronary arteries. Only image data acquired when the diaphragm is positioned at end-expiration, that is, between the green lines, is used for image construction.
PART I  The basics of cardiac MR

Note that the number of images in a cardiac cycle that can be reconstructed from such a sequence is determined independently by use of data interpolation; a large number of such images makes a cine loop appear very smooth and pleasing to the eye, but the true temporal resolution is unrelated to this [20]. Reduction of the TR in retrospectively-gated sequences is the most important change to achieve good image quality in images in patients with fast heart rates.

Those sequences that are prospectively gated, such as turbo-TSE sequences, rely on a set time period between the excitation pulse and the multiple refocusing pulses. The short R-R interval in small children may necessitate the use of a “double-gating” technique, when the excitation pulse is applied at the start of one cardiac cycle, and the refocusing pulses are deliberately applied during the following cardiac cycle (Figure 2.4). This allows for tissue contrast to be unchanged from those seen in adult patients, and allows for tissue characterization of cardiac masses etc. It requires double the number of cardiac cycles to acquire the image however and this may be a limiting factor in those children who are breath-holding voluntarily.

Whenever possible, the use of parallel imaging techniques such as SMASH [21], SENSE [22], and GRAPPA [23] should be used to reduce image acquisition times. This is possible only on certain sequences, and with certain coil selections, dependent on scanner manufacturer. It should be noted that with some coils, while using parallel imaging, signal to noise will decrease so the imager
in general, however, this is performed in two sets of image acquisitions. The addition of parallel imaging techniques has allowed for reduced acquisition times sufficient for imaging to occur in a single breath-hold, thus reducing respiratory artifact. This allows for a rapid acquisition to estimate the peak velocity in a vessel, enabling the aliasing velocity to be accurately set at approximately 1.3–1.5 times the peak velocity, ensuring optimal precision in the velocity measurement while avoiding aliasing.

However, if a breath-holding technique is used for a definitive flow measurement, it is vital to consider the effect of a breath-hold on the circulation. Some children with complex cardiac disease have central systemic veins that connect directly to the pulmonary arterial circulation, via Glenn anastomosis or Fontan conduits, with no ventricle. Pulmonary flow in such patients is partly dependent on respiration [26]; normally flow is fairly continuous and at low velocity (rarely exceeding 0.5 m/s), with a biphasic pattern during the cardiac cycle. The changes in intrathoracic pressure which occur in patients who are breath-holding alter the flow in a Fontan circulation, and therefore flow measurements in the central veins or pulmonary arteries may not reflect true clinical effects. The use of longer acquisitions with free breathing, which needs to weigh this factor with the decrease in time.

**Flow measurement**

Flow imaging is performed with a sequence which includes a flow-encoding gradient pulse in a gradient echo cine sequence, and a reference data set which uses gradient moment nulling to remove all phase shift from constant velocity generated by other sources of phase shift such as local field inhomogeneity; subtraction of the two sets of data results in a “phase” image in which the signal in a voxel is proportional to the direction and velocity of the contents of the voxel [20,24]. Such sequences can be performed with breathing, allowing for some temporal respiratory averaging over the cardiac cycle or with breath-holding.

Flow imaging can be used to quantify a cardiac shunt [25], by measuring flow in the aorta and the main pulmonary artery. Ideally, this should be done on the same acquisition, so eliminating errors due to a change in the cardiac output between two separate acquisitions. Positioning of such an imaging slice can be problematic, but if the volume of flow is the important measurement, then this is not altered by the imaging slice being positioned at a small angle to the ideal, perpendicular to the direction of flow. Peak velocity measurements will not be accurate in such a scenario however. In general, however, this is performed in two sets of image acquisitions.

The addition of parallel imaging techniques has allowed for reduced acquisition times sufficient for imaging to occur in a single breath-hold, thus reducing respiratory artifact. This allows for a rapid acquisition to estimate the peak velocity in a vessel, enabling the aliasing velocity to be accurately set at approximately 1.3–1.5 times the peak velocity, ensuring optimal precision in the velocity measurement while avoiding aliasing.
effectively averages out the effects of respiration on the Fontan circulation, should be considered mandatory.

**Contrast angiography**

Performance of a contrast angiogram is an excellent way to obtain an overall view of the thoracic vasculature, the cardiac connections and lesions thereof. It is usually possible to include the whole thorax and much of the abdomen within the field of view. Angiographic sequences may be performed with or without prospective ECG-gating; the former usually requires increased slice thickness, and hence a reduction in spatial resolution, because of the inherently longer acquisition time. Vascular structures that are remote from the heart can be seen well without ECG-gating, but those that are directly adjacent to the heart such as the central pulmonary veins, or the very medial lobular pulmonary artery branches, will appear blurred if ECG-gating is not used, as will the intracardiac anatomy. There is good evidence that the use of ECG-gating improves diagnostic information in this circumstance [27]. The choice of whether to use ECG-gating depends on the region of interest in a particular patient therefore (Figure 2.5).

The timing of the contrast acquisition can be performed in two ways. A small test dose of contrast can be given, and a low-resolution 2D T1-weighted sequence performed repeatedly to detect the arrival of the contrast in a particular structure, and to allow for calculation of an appropriate timing delay between contrast administration and the image acquisition. This allows the contrast

![Figure 2.5](image)

**Figure 2.5** Examples of the benefit of using an ECG-trigger during contrast angiography in the thorax. In (a), a coronal MIP image shows the ventricular septum and the differing contrast concentrations in the left and right sides of the heart, with no artifact from cardiac motion in an infant of 6 months of age. In (b), the right upper pulmonary vein, and left heart are seen well in a child of 9 years, with no cardiac motion artifact (note the anomalous pulmonary venous drainage from the apical segment of the right upper lobe to the SVC).
bolus to be timed to arrive at the time of central k-space filling, and thus achieve optimal image contrast. Angiographic sequences vary as to the temporal position of central k-space filling within the sequence, and it is important for the operator to verify this in advance for the particular sequence in use. An alternative method of timing is for the full contrast bolus to be given, and the 2D T1-weighted sequence is used to trigger the full angiographic sequence as the arrival of the contrast bolus within a pre-determined structure is detected; in this technique, an angiographic sequence with central k-space filling at the start of the acquisition is selected.

The volume of contrast used is weight-dependent, and thus will be extremely small in neonates and small infants; the contrast is followed by a saline bolus to advance it through the systemic venous circulation. The temporal length of the contrast bolus can be adjusted by slowing the rate of injection, but image quality is better if the bolus is still a relatively short, concentrated one, timed to arrive in the vessels of interest at the time of central k-space filling, rather than a longer and therefore inherently more dilute bolus.

MR angiography is usually performed as a subtraction technique, with an acquisition performed prior to administration of contrast used as a means of subtracting out underlying soft tissue signal, leaving a 3D data set of the vessels with contrast within them. A further angiographic sequence performed immediately after the contrast acquisition can occasionally provide additional information: if there have been problems with the contrast injection due to operator error or injector pump malfunction, this delayed acquisition may be sufficient to allow for a diagnosis to be made rather than the study be a failure.

Improved acquisition speeds, with the use of parallel imaging and data-sharing techniques, in conjunction with a reduction in through-plane spatial resolution, allows for angiographic data sets to be acquired in time periods of less than a second [28]. Multiple such time-resolved acquisitions can be used to investigate abnormal blood flow through the systemic and pulmonary circulations [29].

It is important to examine the extrathoracic vasculature when this is included in the field of view; this may allow other important and previously unrecognized additional non-cardiac diagnoses to be made (Figure 2.6).

Artifacts
Cardiac MR imaging is susceptible to all the usual imaging artifacts inherent in MR imaging. However, there are a number of artificial devices, used almost exclusively in patients with congenital cardiac disease, whose presence will affect the images obtained. There is extensive literature available as to the safety of performing MR imaging in such patients [30, 31], but these rarely comment on the amount of artifact seen.

The amount of artifact varies with the composition of the device, and the sequence used. Most metallic devices are made of material that causes a small amount of local artifact only; these include some arterial stents, ASD and PDA closure devices. Embolization coils made of stainless steel cause a very large amount of artifact (Figure 2.7), and their use is contra-indicated in the thorax and upper abdomen in congenital cardiac patients because this effectively prevents follow-up with MRI; they have been superseded with coils made of platinum for this reason. Changing the sequence used can make a significant amount of difference to the amount of artifact seen; in general the greater the magnetic gradients used, and the faster they are changed in a sequence, the greater amount of artifact a particular device will cause. Thus the artifact on a turbo-TSE sequence will usually be less than that seen on a SSFP cine sequence, which in turn will be less than that seen on an MR angiogram (Figure 2.8).

Sample settings
Table 2.1 lists some sample settings for imaging infants and children with congenital heart disease. This table is not meant to be an exhaustive list of all the types of cardiac MR techniques nor should these settings be taken as unchangeable, but rather they are guidelines and starting points which should be tailored to the patient. The table, in specific, is created for a Siemens system. Users of other systems should adapt these general guidelines to their own vendor’s specifications.
Figure 2.6 Examples of unsuspected and important extracardiac pathology found incidentally during cardiac MR imaging. In (a), there is crossed fused ectopia of the kidneys on the right side, with associated abnormal ureteric course. In (b), there is hydronephrosis of the left kidney, which was found later to be due to pelvi-ureteric junction obstruction. Both patients were neonates with heterotaxy syndrome; both images are from contrast MRA acquisitions.

Figure 2.7 Artifact from steel embolization coils in the internal thoracic arteries on scout images, preventing further cardiac MR imaging. (a) coronal, (b) sagittal.
Figure 2.8 Artifact from an Amplatzer 5 mm septal occluder, placed across an extracardiac Fontan conduit fenestration on an (a) T1-weighted TSE; (b) T2-weighted HASTE and (c) TrueFISP cine acquisition; the amount of artifact varies with the sequence used.
Table 2.1 Sample settings for various CMR techniques (all settings are for infants and children who cannot breath-hold).

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Setting</th>
<th>Parameter</th>
<th>Setting</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stack of Turbo Spin echo images throughout the chest</td>
<td>TR (ms)</td>
<td>500 (entire slab)</td>
<td>OS</td>
</tr>
<tr>
<td></td>
<td>TE (ms)</td>
<td>23</td>
<td>Rectangular FOV</td>
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<tr>
<td></td>
<td>FOV (mm)</td>
<td>150</td>
<td>Number of slices</td>
</tr>
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<td></td>
<td>Thickness (mm)</td>
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<td>Turbo factor</td>
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<td></td>
<td>Distance factor</td>
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<td>Echo spacing (ms)</td>
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<td></td>
<td>Matrix</td>
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<td>Flip angle (deg)</td>
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<tr>
<td></td>
<td>NEX</td>
<td>4</td>
<td>Bandwidth</td>
</tr>
<tr>
<td>Stack of static steady state free precession images throughout the chest</td>
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<td>166</td>
<td>Segments = 29</td>
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<tr>
<td></td>
<td>TE (ms)</td>
<td>1.59</td>
<td>Trigger delay =</td>
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<tr>
<td></td>
<td>FOV (mm)</td>
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<td>334ms</td>
</tr>
<tr>
<td></td>
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<td></td>
<td>Matrix</td>
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<td>Partial phase Fourier</td>
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<td></td>
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<td>Thickness (mm)</td>
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<td>Phase partial Fourier</td>
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<tr>
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<tr>
<td></td>
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<td>Bandwidth</td>
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<td>Bandwidth</td>
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References


Assessment of morphology

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Introduction
Accurate planning for cardiac magnetic resonance (CMR) morphologic assessments requires an understanding of the role of CMR in congenital heart disease. Two-dimensional echocardiography may have poor acoustic windows which can limit this technique although in some instances, patients can undergo surgery with just echocardiography alone [1]. Extracardiac structures such as the great arteries and great veins, complex intracardiac connections (baffles, superior–inferior ventricles), and airway-vascular inter-relationships may be particularly difficult to profile by echocardiography, even preoperatively in the neonate and young infant. In the past, these patients would have undergone diagnostic cardiac catheterization or have gone to surgery without the optimal amount of information; however, CMR has gradually replaced invasive angiography for these diagnoses. The ability to couple function and flow information, true 3D angiography, and the lack of ionizing radiation makes CMR the modality of choice for imaging morphology.

Since many patients undergoing CMR will have had prior echocardiograms, the CMR examination is generally more targeted than for echocardiography. Often, some of the patient’s anatomic lesions have been identified prior to arriving in the MRI suite, but there usually remain unanswered questions to totally characterize the lesion. Consequently, details of previously identified anatomic findings from prior clinic, echocardiogram, surgical or catheterization notes are extremely helpful in planning the CMR examination. In some cases, direct review of the echocardiogram is warranted. A chest X-ray, if available, should always be reviewed for the presence of clips, coils, stents, and even pacemakers/retained pacing leads; metal artifacts may preclude an interpretable study if they are too close to the anatomy of interest.

The most important preparatory step in planning the MRI examination is communication with the patient’s cardiologist. Patients with congenital heart disease often have multiple abnormalities. Some of these defects may have already been well characterized by other modalities. It is important to make sure that the imaging time is properly devoted to the unresolved issues rather than duplicating prior work although confirming presumed findings may also be important in many situations; a systematic approach to the patient and the lesion is always the approach in the MRI examination. In addition, one must keep the mindset of the patient’s practitioner. For example, when a patient is referred for an aortic arch evaluation, the referring physician wants to know more than whether there is a coarctation or not. He (or she) wants to know whether to manage expectantly, to refer for surgery or to attempt intervention in the catheterization laboratory. The imaging protocol must be designed with these questions in mind.
As a general rule, the goals of CMR in patients with congenital heart disease are three-fold:
1. Characterize location and anatomic severity of the primary lesion and define the extent of the secondary lesions.
3. Identify both associated and coincident defects.

Characterizing anatomic location and severity is simply achieved by profiling the defects in orthogonal planes or in true 3D reconstructions such that hemodynamic and physiologic significance can be inferred. Associations must also be defined and consistent with the primary lesion (e.g., bicuspid aortic valve with coarctation of the aorta or left ventricular hypertrophy in the presence of aortic stenosis). Both static and dynamic images should be obtained. The best pulse sequences for this purpose will be discussed later.

Lesion severity can also be assessed by its functional consequences. For example, left ventricular mass is an objective functional measure of left-sided obstruction. Similarly, the ratio of pulmonary to systemic blood flow is a marker left to right shunt and pulmonary relative to systemic vascular resistance. In parallel hemodynamic circuits, such as the pulmonary arteries or veins, quantitative flow measurements are better markers of stenosis significance than pressure gradients (whether directly measured or inferred). Severe branch pulmonary artery stenosis may produce little flow acceleration in the affected vessel if there is compensatory dilation of the parallel limb. Pulse sequences for functional and flow analysis will be discussed in other chapters.

The third goal simply reflects the complexity of these individuals. Even patients that have had comprehensive cardiac evaluations and surgery may have incomplete or incorrect diagnoses. While focused examinations and tailored protocols are the rule, a systematic approach is still essential, analogous to segmental techniques used in ultrasonography [2]. Protocol planning MRI on new patients should therefore include enough “big picture” imaging to identify previously unsuspected abnormalities. It is not terribly uncommon to discard a carefully scripted protocol based upon unanticipated findings discovered in preliminary “survey” image acquisitions.

In this chapter, a general framework for anatomic characterization of congenital heart disease is provided. The initial section outlines the “imaging toolbox”, interleaving specific examples. Next, exam planning and execution are discussed. Lastly, two clinical examples are given, integrating different CMR imaging techniques to provide a comprehensive diagnosis.

**Methodology**

Although there are dozens of cardiac pulse sequences for anatomic imaging, many with manufacturer-specific features, they can crudely be broken down into three classes:
1. Black-blood imaging.
2. White-blood cine and static imaging.
3. 3D gadolinium-enhanced angiography.

Sequences for evaluating functional parameters such as tissue-strain, blood velocity and flow, perfusion and viability will not be discussed in this chapter.

**Black-blood spin-echo imaging**

Black-blood spin-echo techniques are used to generate static images with high signal-to-noise ratio, resolution, and image contrast. Images must be gated to the heart rate so repetition time (TR) is restricted to integer multiples of the cardiac period. Image contrast is controlled by adjusting the echo time (TE). Cardiac muscle has a normal T2 of around 30–40 ms. Thus, echo times less than 20 ms are T1-weighted and greater than about 60 ms are T2-weighted. Most anatomic characterization is performed with T1-weighted images; T2-weighted imaging is reserved for questions regarding tissue characterization. As a result of the radiofrequency refocusing, spin-echo techniques are inherently more robust to metal artifact. Metal artifact can be problematic in post-operative patients where surgical clips cause local image voids and anatomic disruption. Black-blood techniques are particularly useful when the relationships between airway and vessels must be elucidated [3–8]. Figure 3.1 demonstrates four examples. Panel (a) demonstrates a double aortic arch with a severely constricted trachea (arrow) while panel (b) illustrates a focal proximal left bronchial stenosis without any associated vascular abnormality. In panel (c), the left
pulmonary artery (arrow) is stenotic, constrained by a large Damus–Kaye–Stansel anastomosis anteriorly and the left bronchus posteriorly. Lastly, panel (d) demonstrates a widely patent trachea and left bronchus following division of a vascular ring (left arch, right descent, aberrant subclavian).

Black-blood images are also vital for characterizing masses, thrombus, abdominal situs, hematomas or other soft tissue details [8–12]. Figure 3.2 demonstrates T1 and T2 weighted images of a giant intramuscular mass occupying most of the left ventricle. The tumor was isointense on T1-imaging.

Figure 3.1 Airway-vessel relationships. (a) Axial standard spin echo image demonstrating a double-aortic arch with dominant right arch and tracheal compression (arrow). (b) Coronal, breath-held, double-inversion recovery fast spin-echo image of isolated, severe, proximal left bronchial stenosis (arrow). (c) Sagittal standard spin-echo image demonstrating a double-inlet left ventricle (LV) after Damus–Kaye–Stansel (DKS) anastomosis and Blalock–Taussig shunt with left pulmonary artery stenosis (arrow). The left pulmonary artery was distorted by enlarged DKS anastomosis anteriorly while left bronchus and descending aorta prevented posterior expansion. (d) Breath-held double-inversion recovery fast spin-echo image illustrating a right aortic arch with left descending aorta and aberrant subclavian artery from a diverticulum of LV after Kommerell who was after release of a vascular ring. The trachea and left bronchus (arrow) are well profiled and without stenosis.
There are two basic black-blood implementations, standard spin echo and fast spin echo. Standard spin echo uses a single heartbeat per phase encoding step. Since each phase acquisition is relatively short compared to the repetition time, multiple slices can be obtained through interleaving. As a consequence, each slice is obtained at a different delay from the cardiac trigger; this fact must be considered when interpreting the resulting anatomy. Scans must be performed with the patients freely breathing and special approaches are needed to reduce respiratory artifact. Some scanners allow data acquisition to be restricted to end expiration (so-called respiratory triggering). The respiratory signal may come from abdominal bellows or from a navigator echo. Using this technique, image quality is high but imaging efficiency is low, leading to long acquisition times. Some platforms do not support respiratory triggering for standard spin echo imaging. An alternative approach is to use multiple acquisitions of the same image (respiratory averaging) and/or to order phase acquisition intelligently with respect to respiratory variation (respiratory ordered phase encoding). Respiratory compensation algorithms may require an even number of averages. We typically use four excitations, leading to an acquisition time of 5–8 minutes, depending on heart rate. As a rule, respiratory compensation and respiratory triggering work reasonably well in young, sedated patients who have sinusoidal breathing patterns, but often fail miserably otherwise.

Although standard spin echo was once a workhorse of cardiac MRI, we now limit its use to term and premature infants where signal to noise is paramount and respiratory artifacts are low. Slice thicknesses as low as 1.5 mm can be obtained in these patients, with Gibb’s ringing being the limiting factor. Infants often have heart rates greater than 120 beats per minute, lowering T1 contrast and decreasing the number of available slices. To correct for this, the scanner may be adjusted to trigger on every other R-wave, doubling the effect TR, slices acquired, and scan time. In order for standard spin echo techniques to produce “black

Figure 3.2 (a) Axial, precontrast, T1-weighted, breath-held double inversion recovery fast spin-echo image demonstrating a huge intramural mass involving the left ventricle. Tumor is isointense with ventricular muscle but more heterogeneous on both pre and post-contrast (not shown) scans. (b) Axial, T2-weighted, breath-held double inversion recovery fast spin-echo image in the same scan orientation. Tumor is slightly brighter (longer T2) than ventricular muscle but demonstrates considerable heterogeneity. LV: left ventricle.
blood", blood must flow through-plane in the time between the 90 and 180 degree pulse. Since blood vessels tend to run superior–inferior, inadequate blood nulling can be problematic in coronal and sagittal imaging planes.

The second class of black-blood techniques, so-called fast spin-echo techniques, use a train of refocusing pulses to collect multiple phases per excitation, accelerating the acquisition by a factor of the echo-train-length (ETL). The optimum echo train length is primarily limited by cardiac motion; the echo-train duration is like an open camera shutter. As a result, image acquisition is typically initiated during diastole when cardiac motion is minimized. In addition, it is necessary to decrease the ETL as heart rates rise (and diastole shortens). A useful rule of thumb is to adjust ETL to keep the image acquisition time for a single slice between 12 and 16 seconds. The chief advantage of fast spin echo is that it enables single slice acquisition in a breath-hold, eliminating respiratory artifact. It also allows targeted (a few slices), high resolution imaging in a much shorter time than standard spin echo. Image contrast for fast spin echo is controlled by reordering phase acquisition such that the center of k-space is collected near the desired TE. Longer TEs, typically around 40 ms, are used with fast spin echo compared with single spin echo creating a mixed T1, T2 contrast. Since there is little through-plane blood flow during image acquisition, fast spin-echo techniques are not inherently "black-blood". Consequently, cardiac fast spin echo is often used in conjunction with an inversion pulse to null the blood pool. Default value of the inversion time is around 150 ms but may need to be shortened if there is circulating gadolinium contrast. Most "black-blood" techniques now use the "double inversion" technique to generate even better image quality. A "non-selective" inversion pulse is used to flip the protons 180 degrees and destroy spins throughout the body; then a "selective" inversion pulse is used to flip the protons in the slice position of interest back to 0 degrees restoring the spins at that position. This has the effect of making the cavity of the heart or great vessels very dark as blood flowing into the slice position of interest have their proton spins destroyed; only tissue within the slice position of interest gives off signal. This yields even better image quality.

Fast spin echo techniques are the preferred black-blood imaging modality for all patients who can breath-hold. They may also be used in free-breathing patients by increasing the number of acquisitions to 3 or 4. Fat-ghosts from the moving anterior chest wall can be ameliorated by placing an anterior saturation band.

**White-blood cine and static techniques**

While static, black-blood pictures can answer some questions, they may incompletely characterize lesion severity. Black-blood techniques for anatomy have generally been supplanted by white-blood static techniques, generally of the steady state free precession type. A full volume set of the entire thorax may be obtained in <30 seconds with the single shot technique while with segmented approaches and multiple averaging, a typical volume set may take 2.5 to 3 minutes. In contrast to black-blood techniques, all the static steady state free precession images are obtained at the same point in the cardiac cycle (generally end-diastole); obtaining images having high contrast between the cavity which is bright and the soft tissue which is dark. Steady state free precession techniques have high signal to noise. An occasional drawback is when there is turbulent flow in diastole – signal from the cavity is lost and structures cannot be visualized (e.g., pulmonary arteries in a Stage I Norwood operation). In these particular instances, dark blood techniques are then used.

Any sonographer can relate the difficulty of interpreting ultrasonic still-frames taken out of their dynamic-context and CMR is no different. Cine CMR images offer the same physiologic insights provided by echocardiography, even though they are not always acquired in real-time. For this reason, cine images represent the backbone of CMR despite their slightly lower resolution and contrast at times compared with black-blood imaging. Cine imaging offers improved temporal resolution because it uses gradients to refocus echoes instead of slower radiofrequency excitations. Radiofrequency pulses also deliver too much heat to be used for cine imaging. There are two basic variations of fast gradient echo imaging. Low flip angle, spoiled gradient echo techniques (known as SPGR or FLASH) have been routinely used for
nearly two decades. In contrast, relatively newer techniques use the higher flip angle steady-state free precession techniques (SSFP). In “classic” cine mode, one phase is acquired per heartbeat producing sampling rates of 100–300 Hz for single slice acquisitions. However, classic cine is so slow (several minutes per slice) that it has been replaced by segmented acquisitions. Segmented techniques typically divide the cardiac cycle in temporal segments where multiple image phase-encodes (or views) are binned together. The imaging time per slice is reduced by the views per segment (VPS) binned, allowing images to be collected in a single breath-hold. The tradeoff between imaging time and temporal blurring follows the same principles as for ETL in fast spin echo imaging. The intrinsic temporal resolution of the cine sequence is simply the VPS multiplied by the TR. As a result, faster heart rates require lower VPS for accurate anatomic characterization.

SPGR represents the older of the two techniques and has generally been replaced by SSFP but still retains some clinical utility [13–17]. Its advantages are its robustness and its flow sensitivity. SPGR techniques are well-conditioned and may succeed when poor shimming or respiratory motion cause other techniques to fail. SPGR techniques are T1-weighted and blood only appears bright because spins flowing into the imaging slice are naive. Slowly moving blood darkens slightly because of saturation. Blood flowing into the imaging plane is bright because it contains non-saturated spins. This process is also known as flow-mediated enhancement. Blood saturation effects create the appearance of streamlines, even in slow-flowing blood such as systemic and pulmonary veins. In contrast, blood that accelerates to the point of turbulence produces signal voids in the image because of spin randomization. The combination of flow-related enhancement and turbulent signal void produce patterns similar to those observed with color Doppler in echocardiography and help to highlight hemodynamically significant stenoses.

Figure 3.3 demonstrates a patient with known pulmonary stenosis who underwent an MRI to evaluate her branch pulmonary arteries. While these were normal, an unsuspected high-grade stenosis was observed at the junction between the superior vena cavae and the right atrium. Sagittal SPGR imaging demonstrated a discrete tissue web with a pinpoint communication and a continuous high velocity turbulent jet extending into the RA (Figure 3.3a,b); paraspinous venous collaterals were also observed, accounting for the inferior vena cava (IVC) dilation. Because of the thin and mobile nature of this obstruction, this lesion was invisible on the 3D-contrast enhanced magnetic resonance angiography (CEMRA), but was also evident on gated black-blood imaging (not shown). This patient underwent successful balloon angioplasty which eliminated a 12 torr mean gradient. Figure 3.3c,d demonstrate a patient with restenosis of a pulmonary venous anastomosis following repair of total anomalous venous drainage. SPGR imaging nicely characterizes a turbulent jet that extends through the open mitral valve in the left ventricle. Some phasic variation is seen; phasic variation of turbulent jets is reduced or even eliminated with increasing lesion severity. In this patient, anastomotic re-obstruction was also identified by echo with a mean gradient of 12 torr. The primary purpose of the MRI was to exclude more distal venous occlusion as well as to assess pulmonary artery anatomy.

SPGR can be used to characterize severity of either valvular or vascular stenoses [16]. SPGR yields best images with breath-holding, however, it works relatively well with either respiratory-triggering or respiratory-averaging. The primary limitation of SPGR is its relatively poor image contrast and low signal to noise ratio because of the low-flip angle excitation.

SSFP offers many advantages to SPGR. In SSFP, spoiling is eliminated yielding TRs that are nearly 50% shorter than for SPGR. This allows better temporal and spatial resolution during breath-hold imaging. SNR is higher because SSFP can use larger flip angles (typically 75 to 90 degrees instead of around 20 degrees). Image contrast is much higher and represents a product of T1 and T2 weighting. As a result, SSFP yields better coronal and sagittal imaging than SPGR because it is less vulnerable to saturation of in-plane blood flow. This is illustrated in Figure 3.4 which demonstrates SSFP imaging of an extracardiac Fontan connection (panel (a)). The Glenn anastomosis is profiled in the same slice. SSFP is also particularly useful in evaluating myocardium, masses, and pericardial
contrast, speed, and resolution, SSFP has largely replaced SPGR for myocardial imaging as well as extracardiac anatomic characterization in adults and adolescents [20,21]. SSFP does have a few disadvantages, however. Its ultrashort TE produces less flow sensitivity to vascular or valvular obstructions, particularly in low-flow lesions such as atrial septal defects and

structures [18,19]. Figure 3.4 (panel (b)) demonstrates a short axis SSFP image from a patient with a one month history of pericarditis. Notice how well the pericardium, effusion, and myocardium are distinguished. A thick rind (5–6 mm) is demarcated posteriorly with focal adhesions, later confirmed by tagging (this patient later underwent pericardial stripping). As a result of its superior

contrast, speed, and resolution, SSFP has largely replaced SPGR for myocardial imaging as well as extracardiac anatomic characterization in adults and adolescents [20,21].

SSFP does have a few disadvantages, however. Its ultrashort TE produces less flow sensitivity to vascular or valvular obstructions, particularly in low-flow lesions such as atrial septal defects and
CEMRA is absolutely essential for charactering small vessel disease such as peripheral pulmonic stenosis or aortico-pulmonary collaterals [23–27]. CEMRA offers comparable diagnostic accuracy as cardiac catheterization in evaluating the number and location of aortico-pulmonary collaterals in children [24,25] and adults with pulmonary atresia [27]. Figure 3.6 demonstrates a patient with suspected Alagille’s syndrome and peripheral pulmonic stenosis. Proximal PAs are very dilated but prune immediately in second and third order branches (panels (a)–(c)). Both the right and left side are heavily involved. Vessels have an irregular, “beaded”, appearance for alternating dilations and constrictions. Flow profile by phase contrast velocity measurements (not shown), often exhibits “ringing” and systolic prolongation. Panels (d)–(f) demonstrate the utility of 3D reconstruction to provide good spatial “sense” for pulmonary arterial supply in a patient with pulmonary atresia and multiple pulmonary aortopulmonary collaterals. A 3.5 mm modified Blalock-Taussig shunt supply blood to continuous true pulmonary arteries, however the right pulmonary artery remains hypoplastic (panel (d)). Three aortopulmonary collaterals are identified (panels (e), (f)). Based upon these data, a right unifocalization was proposed.

pulmonary venous stenoses. A second limitation is that when needed, longer TR and TEs degrade image quality and worsen temporal resolution. Lastly, SSFP is troubled by metal artifact (such as sternal wires) and by flow artifacts near regions of rapid blood acceleration [22]. These artifacts are worse in infants and young children because blood acceleration in the great arteries is larger than for adults. Three-dimensional local shimming improves but does not eliminate these artifacts.

3D gadolinium-enhanced angiography
CEMRA is a true 3D acquisition technique. Imaging voxels are more isotropic than for 2D acquisitions, making CEMRA studies well suited for multiplanar reformatting and volume-rendering. Thus CEMRA is an ideal technique for characterizing tortuous vascular connections such as complex arch and pulmonary artery and vein variants (Figure 3.5a–c). Three dimensionality is also helpful in surgical visualization and planning in more ordinary lesions such as small aortic aneurysms (Figure 3.5d).

CEMRA, because of its excellent SNR, has the highest resolution of any of the cardiac imaging sequences. Although standard 2D imaging may be adequate for diagnosing large vessel disease,
The physical principles behind CEMRA are straightforward. CEMRA uses large flip angles and the shortest possible TR and TE, saturating protons from all tissues having a long T1. Intravascular gadolinium, however, has short enough T1 to yield bright vessels against virtually no background tissue signal. The short TR and TE, combined with fractional frequency and/or phase sampling, allow the use of 3D phase encoding in a single breath-hold. Acquisitions are often not cardiac

**Figure 3.5** (a) Cervical right arch with left descending aorta. There are separate takeoffs of all four head vessels with an unusually dilated right subclavian artery; fibrous continuity between the left subclavian and left carotid were observed at surgery. The transverse arch is severely hypoplastic. Innominate vein passes posteriorly and inferiorly of its normal location. (b) Total anomalous venous drainage with decompression to inferior spinal venous plexes. There is tremendous systemic venous collateralization, posteriorly, without significant azygous or hemiazygous drainage. (c) Truncus arteriosus with aorta arising from the right of a common trunk. The aortic arch is normal but the pulmonary arteries are massively dilated. (d) Patient with proximal aortic aneurysm, most likely as a result of prior aortic cannulation. IV: innominate vein; LSCA: left subclavian artery; PVC: pulmonary venous confluence; RSCA: right subclavian artery.
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Figure 3.6 (a–c) 3D CEMRA in the coronal plane for a patient with suspected Alagille’s syndrome and peripheral pulmonic stenosis. Central pulmonary arteries are massively dilated with diffuse pulmonary arteriopathy in the second and third order branches. (a) Near interruption of the right pulmonary artery (RPA) as it trifurcates into upper, middle, and lower branches (arrow). (b, c) Alternating dilations and constrictions (arrows) in the left lower pulmonary artery (LPA) creates a “beaded” appearance to the vessel. (d–f) Coronal 3D-CEMRA in a patient with tetralogy of Fallot, pulmonary atresia and multiple aortopulmonary arteries after Blalock-Taussig shunt (BTS) to small native pulmonary arteries. (d) Shunt (BTS) inserts into the pulmonary artery bifurcation (arrow). The right pulmonary artery (RPA) failed to grow following the initial procedure and is nearly isolated. The left pulmonary artery (LPA) has grown well and has relatively normal arborization. (e) 10 mm coronal maximum intensity projection image demonstrating three aortopulmonary collaterals from an anterior perspective. Collaterals #1 and #3 supply the right upper and right lower distributions, posteriorly. (f) A 3D reconstruction from a posterior perspective illustrating the relative spatial relationships of the three collaterals and the true pulmonary arteries.

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triggered, so CEMRA is not useful for intracardiac diagnoses; however, most manufacturers do have the capability of using ECG triggered CEMRA and this can allow for intracardiac diagnosis.

CEMRA represents an “off-label” use of gadolinium. Dosing guidelines are empiric and based on adult studies. Gadolinium is cleared by glomerular filtration which varies proportionally to body surface area, rather than patient weight. Hence, one would expect that children could tolerate greater per-kilogram dosing compared with adults. Previous studies have typically used 0.2 mM to 0.5 mM gadolinium per dose with an exam maximum of 0.5 mM/kg [24–26]. Power injection is quite helpful in larger patients but we use hand injection at 0.5–1 cc per second in infants and small children with tenuous venous access. For infants less than 7 kg, we typically dilute the contrast up to a total volume of 3 cc and inject at 0.5 cc/second. Contrast injection should be followed by saline flush of 5–10 cc. Systemic venous anomalies or known venous occlusions should be considered when placing the IV. For example, a lower extremity IV may not opacify the pulmonary arteries in a patient with a Glenn shunt (occasionally the Glenn shunt will be visualized on the “levophase” or late
systemic venous return phase). Bilateral upper extremity IVs may be necessary in patients with dual caval systems to evaluate upper extremity venous thromboses or collaterals.

CEMRA image quality depends upon synchronization of the central portion of k-space with peak contrast enhancement. Many systems now allow the operator to track contrast progress using a projection, “bolus tracking” or “fluoro” mode [28]. This allows the high resolution CEMRA examination to be triggered during maximum contrast enhancement. Other systems allow data acquisition to be triggered from a sentinel region of interest (for example in the aorta). Timing boluses can be used to estimate the contrast delivery curve, however, the injection conditions during the test bolus often don’t match the actual angiogram. Contrast delivery is delayed in lower extremity injections, smaller bore IVs, low cardiac output states and in patients with expanded venous capacitance (Fontan patients). In contrast, gadolinium delivery through a central line produces almost immediate right-sided opacification. Despite all of these considerations, excellent results can still be obtained using empiric knowledge of the pulse sequence and the injection technique/site.

Proper bolus synchronization also requires an understanding of the CEMRA phase ordering. Standard, fully-sampled acquisitions capture the center of k-space at the acquisition midpoint. Fractional acquisitions move the center of k-space earlier, decreasing the necessary delay between contrast injection and image collection. Centric or elliptico-centric techniques acquire k-space from the “inside-out” [29]. These approaches require nearly peak enhancement prior to scan initiation or vessels will exhibit central non-enhancement. In infants and children, standard rectilinear phase order (full or partial acquisitions) should be used.

CEMRA images are typically collected over multiple temporal phases. Not only does this insure at least one interpretable anatomic snapshot, even if bolus timing was imperfect, but temporal information can also be useful clinically. Early contrast enhancement is indicative of low resistance pathways, such as AV fistula, while delayed contrast enhancement can be observed with stenoses or extravascular abnormalities. Figure 3.7 demonstrates angiogram images from an otherwise healthy 5-year-old boy with unexplained cyanosis and a positive contrast-echocardiography study. To evaluate for possible early pulmonary venous return, the CEMRA was acquired with the center of k-space occurring within 4 or 5 seconds of injection. Patient was free-breathing so image quality is poor, however right pulmonary vein (RPV) opacification is clearly prominent when left pulmonary veins remain unenhanced (panel (a)). Second pass (panel (b)) delineates the location and extent of the fistulous vessels in the posterior right lung. The posterior aspect of the left lung exhibits some minor changes as well. Panels (c) and (d) represent first and second pass angiograms from a patient with Marfan’s disease and chest pain. First pass fills the true aorta and the proximal portion of the dissection through the entry point at the base of the innominate artery. Aortic lumen was compromised by intimal dissection throughout the entire transverse arch, having a semilunar appearance on cross-sectional cuts (not shown). The origin of the left subclavian was also compromised by direct extension of the intimal flap into this vessel. Second pass (panel (d), roughly 24 seconds after injection) highlights contrast retention within the massive pseudoaneurysm; head vessels and true arch lumen are relatively less enhanced.

Recently, parallel imaging and view-sharing strategies have greatly shortened CEMRA acquisitions. While not yet approaching the temporal resolution of X-ray angiography, it comes close and these techniques clearly separate arterial and venous phases with high resolution in three dimensions [30,31]. Imaging techniques such as TWIST and Keyhole produce high temporal and spatial resolution 3D images that can not only produce 3D reconstructions but also contain important temporal information. The trade-offs between temporal and spatial resolution must be driven by the clinical question as well as the local capabilities of the MRI scanner.

Despite its many advantages and technical improvements, CEMRA is predominantly used in an ungated manner, blurring structures that move during the cardiac cycle. Thin structures, such as membranes, folds, or indentations, are particularly vulnerable to being underappreciated compared with longer segment obstructions. This effect can be particularly prominent in the pulmonary arter-
Figure 3.7 (a, b) Coronal CEMRA in a four-year-old patient with persistent hypoxia and positive contrast echocardiogram. (a) First phase of the CEMRA (triggered at initiation of injection) opacifies the right pulmonary veins (RPV) even before the left pulmonary artery (LPA) is completely filled. (b) The second phase of the CEMRA (postero-cranial angulation) demonstrates persistent contrast opacification of a ball of abnormal vessels posteriorly in the right lung (labeled fistulae). Similar, but more subtle changes are seen in the posterior aspect of the left lung as well. (c, d) Sagittal CEMRA in patient with Marfan syndrome after aortic root replacement who presented with painless dissection. (c) 3D reconstruction of first phase of CEMRA (16-second acquisition, 8-second delay time). The tubular ascending graft is undisturbed but the native aortic lumen from the graft to the descending aortic is semilucent because of intimal dissection (arrow). Dissection passes through origins of the innominate and left subclavian artery and the entry point to the pseudoaneurysm is at the base of the innominate. (d) 3D reconstruction of CEMRA taken immediately following the previous image (16 second acquisition). Contrast has cleared the native aorta (labeled Ao) but is retained in the giant pseudoaneurysm (An).
ies because they are relatively more distensible than the aorta. Imagine the photograph of someone waving their arm while the camera shutter is open for many seconds; stationary or nearly stationary components will be sharp but the thin arm will be thicker and appear to reside in multiple locations. Figure 3.8 demonstrates an example of this phenomenon. A size disparity is observed in the right and left pulmonary artery and there appears to be a mild proximal left pulmonary artery stenosis. However gated double-inversion recovery images (panel (b)) demonstrate only a 2–3 mm communication between the MPA and LPA. White blood cine imaging (not shown) demonstrated a corresponding turbulent jet extending deep into the PA consistent with discrete obstruction. Phase contrast flow imaging confirms the hemodynamic severity of this lesion (panel (c)) with a nearly 10:1 flow differential and near abolition of pulsatility in the left pulmonary artery. Discrete coarctation from a thin posterior shelf, or a venous web can likewise be underestimated; for example, the venous web previously demonstrated in Figure 3.3a was invisible on the angiogram. Therefore it is important to integrate information from all of the available imaging approaches when assessing hemodynamic significance.

Orientation and voxel size for CEMRA acquisitions should be determined by the anatomy of interest. Imaging in principle planes generally produces the shortest echo and repetition times; images are easily reformatted into arbitrary orientations. Image resolution is highest in the frequency direction, followed by the phase and slice directions. Choice of optimal imaging plane should match the image resolution with the anatomic question to be addressed. Zeropadding or other interpolation algorithms introduced by the scanner do not represent real resolution gains, although they can subjectively improve image quality.

Planning and executing the examination

The most important “anticipatory” decisions are the choice of sedation (if any), imaging coil, and imaging protocol (including use of contrast). Choice of sedation and imaging coil are discussed in Chapter 2, but will interact with sequence parameters through respiratory motion and SNR. The decision to use imaging contrast depends on whether the question is anatomic or functional and on the location of the suspected pathology. For example, serial functional evaluation of the heart in a patient with thalassemia does not justify the risks and discomforts of IV gadolinium.

All exams begin with a series of “localizers”, which tend to be low resolution, ungated images

Figure 3.8 (a) 3D reconstruction of axial CEMRA performed in a patient with truncus arteriosus (type 1 1/2) and total anomalous pulmonary venous drainage status post homograft repair. There is size disparity between the right and left pulmonary artery (LPA) and suggestion of mild proximal LPA narrowing (arrow). (b) Gated, oblique, free-breathing, respiratory-averaged (4 averages) double inversion-recovery image oriented along the LPA. There is a fold between the LPA and main pulmonary artery (MPA) with only a 2–3 mm communication (arrow). White blood cine imaging in the same orientation demonstrated considerable anterior-posterior motion of the obstruction during the cardiac cycle and a long-systolic jet of turbulence (not shown). (c) Relative flow-profile in the right (grey line) and left (black line) pulmonary arteries respectively. Pulsatility is nearly abolished in the LPA and there is nearly a 10:1 net flow differential. ms: milliseconds; m/s: meters per second.
that serve as a starting point for more definitive imaging. Each vendor has different sequences that are suited for this task. Some platforms offer real time localization, but image quality is variable and these techniques are not universally available. After initial scout images, our approach has been to collect a stack of axial cine (either free breathing SPGR or SSFP) images covering the top of the arch to the dome of the diaphragm. Although this is time consuming (4–6 minutes), it generally establishes 90–95% of the diagnosis and alerts the operator to previously unrecognized abnormalities. An alternative, as previously mentioned, is an axial stack of static SSFP throughout the thorax which takes 2.5–3 minutes. While multiplanar reconstruction is used on these images to define planes for cine imaging and velocity mapping, an axial stack of HASTE images are obtained to collect a set of dark-blood images to complement the bright-blood ones.

Subsequent sequences should generally be acquired in order of importance because scans occasionally must be terminated prematurely (mechanical failure, patient instability). In fact, some exams could consist of simply a scout and a 3D angiogram, similar to computed tomography, if the clinical question is focused or the patient is unstable. If delayed hyperenhancement imaging is desired, contrast injection should occur relatively early in the exam. The only caveat is that double inversion recovery fast spin echo imaging is easier to use prior to contrast injection. Typically, we perform our imaging in the following order: scouts, axial cine, targeted cines, double IR, angiography, function, flow and delayed hyperenhancement imaging.

The standard segmental approach should be used on all images to discern morphology. In general, the route of flowing blood should be used all the times to evaluate anatomy so as not to miss any important structural features. Atrial, ventricular and great artery relationships must be defined including looping of the ventricles (D vs. L) and position of the great vessels (D vs. L). The inter-segmental connections such as venous, atrioventricular and ventricular-arterial connections must clearly be delineated. Within the heart, abnormal connections such as ventricular or atrial septal defects should be sought along with valve morphology (e.g., bicuspid aortic valve). Extracardiac structures such as collaterals (e.g., aortic-pulmonary) must be searched for and structures such as arterial-venous malformations can be inferred by utilizing “time resolved” 3D gadolinium sequences or visualized directly.

Integrated examples

To illustrate the methodologies described above, we will present two clinical cases integrating information from different pulse sequences. Subsequent chapters will delve into specific anatomical problems in more detail. The first patient was a former 2.4 kilogram infant with heterotaxy syndrome with situs ambiguous, levocardia, unbalanced complete atrioventricular (AV) canal, D-malposed great arteries with pulmonary atresia and infradiaphragmatic total anomalous pulmonary venous drainage (Figure 3.9). He had undergone surgical anastomosis of the pulmonary venous confluence to the common atrium and a modified Blalock–Taussig shunt to the main pulmonary artery. The patient initially did well post-operatively but was reintubated 3 weeks later for respiratory distress and pulmonary edema in the left lung. An MRI was requested to evaluate for pulmonary venous obstruction.

The patient was imaged intubated and ventilated. The use of the knee-coil (2.5-fold higher SNR relative to the head and cardiac coils) allowed a field of view between 14 and 18 cm and a slice thickness of 1.6–3 mm. A stack of axial cine images was performed which demonstrated an unbalanced canal without significant atrioventricular valvar insufficiency (panel (a)). There was also no evidence of pulmonary venous obstruction (panel (b)). Since the patient’s heart rate was 150 beats per minute (BPM), fractional echo time was used to improve temporal resolution. Since this was an initial “survey”, images were collected free-breathing with two averages.

Breath-held coronal double-inversion recovery-images were performed through the whole chest. We favor this approach in heterotaxy patients because it yields excellent discrimination of the situs and great vessel abnormalities. Panel (c) demonstrates a midline liver and dual hepatic venous drainage into ipsilateral atria. Interrupted IVC toazygous continuation was easily profiled as well as a small hemiazygous communication to a left sided
and the true RPA. This most likely represented a ductal constriction and explained the left-sided pulmonary edema observed on chest X-ray.

The final example is a 21-year-old patient with transposition of the great arteries and ventricular septal defect who had undergone a Mustard procedure at 6 months of age (Figure 3.10). The patient was referred for routine follow-up and was asymptomatic. The exam consisted of a 3-plane localizer, 4-chamber and short axis SSFP images, sagittal SGPR images, phase contrast velocity measurements in aorta and pulmonary artery and hepatic veins from a midline liver drain into their ipsilateral atria. Bronchial tree is well-profiled. A hemiazygous vein drains to the left atrium through a remnant of a left superior vena cava (L-SVC), however the superior portion of the L-SVC is atretic. (d) Same technique, more anteriorly, highlighting widely patent Blalock-Taussig shunt (BTS) to the main pulmonary artery. Superior vena cavae (SVC) enters the top of the right atrium. (e) Axial, breath-held double-inversion recovery fast spin echo image is used to size the pulmonary venous anastamosis. (f) 3D reconstruction of axial CEMRA images demonstrating acquired, long-segment discontinuity of the right pulmonary artery (arrow). Proximal pulmonary veins (PV) are unobstructed.

caval remnant (not shown). Left bronchus is epiarterial and right bronchus is hyparterial (panel (c)). Modified Blalock-Taussig shunt to the main pulmonary artery was well-delineated (panel (d)).

A few axial double inversion recovery images were collected to size the pulmonary venous anastamosis (panel (e)). Both the initial axial cine and the coronal inversion recover images suggested an interruption of the right pulmonary artery at its origin. This was confirmed on contrast-enhanced angiogram (panel (f)) which revealed nearly a centimeter gap between the pulmonary bifurcation and the true RPA. This most likely represented a ductal constriction and explained the left-sided pulmonary edema observed on chest X-ray.

The final example is a 21-year-old patient with transposition of the great arteries and ventricular septal defect who had undergone a Mustard procedure at 6 months of age (Figure 3.10). The patient was referred for routine follow-up and was asymptomatic. The exam consisted of a 3-plane localizer, 4-chamber and short axis SSFP images, sagittal SGPR images, phase contrast velocity measurements in aorta and pulmonary artery and hepatic veins from a midline liver drain into their ipsilateral atria. Bronchial tree is well-profiled. A hemiazygous vein drains to the left atrium through a remnant of a left superior vena cava (L-SVC), however the superior portion of the L-SVC is atretic. (d) Same technique, more anteriorly, highlighting widely patent Blalock-Taussig shunt (BTS) to the main pulmonary artery. Superior vena cavae (SVC) enters the top of the right atrium. (e) Axial, breath-held double-inversion recovery fast spin echo image is used to size the pulmonary venous anastamosis. (f) 3D reconstruction of axial CEMRA images demonstrating acquired, long-segment discontinuity of the right pulmonary artery (arrow). Proximal pulmonary veins (PV) are unobstructed.
A 3D CEMRA in the coronal plane. A small residual ventricular septal defect (VSD) with a pulmonary to systemic flow ratio (Qp/Qs) of 1.2 by phase contrast velocity mapping was demonstrated (panel (a)). There was biventricular hypertrophy with preserved systolic function. Four chamber SSFP images demonstrated wide open pulmonary venous connection (panel (b)) and pulmonary outflow (panel (c)). A small baffle leak (panel (d)) and unobstructed inferior systemic venous baffle were seen on sagittal SPGR images. Superior limb tapered with the atria with mild flow acceleration and no turbulence (not shown). The superior limb tapering was also captured by the 3D CEMRA (panel (e)). The pulmonary vasculature was quite abnormal by CEMRA with dilated proximal pulmonary arteries (PAs) and pruned distal vessels. This represents irreversible vascular disease from the relatively late surgical repair. It is concordant with the mild left ventricular hypertrophy present despite any significant outflow tract obstruction. This example demonstrates how seamlessly cardiac MRI blends anatomic and functional characterization of congenital heart disease to provide a comprehensive description of a patient’s congenital heart disease.
CHAPTER 3 Assessment of morphology

References


Assessment of ventricular function and blood flow

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Introduction

Cardiac magnetic resonance imaging (CMR) is the premier imaging modality to assess ventricular function in and blood flow in congenital heart disease (CHD). Important parameters in these types of lesions such as ejection fraction (EF) and end-diastolic volume (EDV) can be accurately measured despite the bizarre ventricular shapes encountered in CHD. Although there is overlap in CMR capabilities with other imaging modalities and procedures such as cardiac catheterization (e.g., cardiac output) and echocardiography (e.g., average velocity in a vessel), CMR has some very special features which are routinely utilized in everyday practice that cannot be duplicated (e.g., averaging ventricular function over multiple heartbeats directly into the image). It has also been shown that CMR can add greater accuracy and reproducibility to readily accepted standard measures of ventricular function and blood flow (e.g., ventricular volumes and mass [1], cardiac output); this has enabled research studies, for example, to markedly decrease the number of patients needed to attain specific endpoints. CMR has been used to accurately assess ventricular volumes and mass, for example, for many years [2–6] with high reproducibility and decreased variability [7,8]. It is considered the “gold standard” even in the echocardiographic community [9–11] where between 1997 and 2005, at least 51 publications compared echocardiography with the “gold standard” of CMR.

Only a general overview of CMR techniques useful in assessing ventricular function and blood flow in CHD is presented in this chapter; whole textbooks related to ventricular function and blood flow along with its assessment by CMR can and have been published. They are beyond the scope of this section. The reader is referred to other more comprehensive treatments of this subject.

CMR techniques

CMR is an extremely broad-based versatile modality with multiple techniques which can be utilized in evaluating ventricular function and blood flow. Each technique can have a unique contribution in evaluating ventricular function and blood flow or can be used in a complementary fashion to use as an internal consistency check on the data. One of the strengths of CMR is not only its quantitative nature, but that there are different techniques that add a check on each other for accuracy.

There are a few generalities that can be applied to the variations in the various techniques which will be discussed.

• Images can be obtained with breath-holding or with “averaging” if the patient cannot hold his breath; “averaging” is obtaining the imaging data multiple times and embedding in the image which allows for the respiratory motion to be mostly
compensated for. Images can be obtained with breath-holding in 5–25 seconds while “averaging” usually takes 20 seconds to 2 minutes.

- As an alternative to breath-holding and “averaging,” “real time” imaging can be performed [12]. The scanner creates images as quickly as possible with or without the ECG; with parallel imaging (see below), temporal resolution can be as great as 30–40 seconds. In addition, “interactive” real time imaging can be performed where a real time window shows the heart beating while other windows are used as reference planes; this is similar to performing “sweeps” as in echocardiography except the reference planes are used instead of the transducer on the chest/abdomen and the real time window is equivalent to the echocardiographic monitor. In general, there is tradeoff of spatial for temporal resolution.

- Arrhythmia rejection: In general, nearly all imaging depends upon using the ECG; this has obvious consequences in patients with arrhythmias. Arrhythmia rejection forces the scanner to use imaging data from only a range of heart rates (e.g., 80–90 beats per minute) so that premature beats or beats with pauses do not hinder the scan.

- Non-parallel vs. parallel imaging: With non-parallel imaging, only one coil is used to acquire data while with parallel imaging, multiple coils are used which speed up the imaging process. The disadvantage for some coil configurations is that signal to noise is decreased.

- Retrospective gating vs. prospective triggering: In retrospective gating, the ECG is recorded as the scanner continuously obtains imaging data. At the end of the scan, software fits the imaging data into the most appropriate portion of the ECG to create the image. In prospective triggering, the scanner uses the QRS of the ECG as a signal to begin imaging; there is a necessary “dead-zone” at the end of each cardiac cycle for the scanner to await the next QRS complex. The advantage of retrospective gating is that it covers the entire cardiac cycle.

- The number of lines of imaging data obtained per heartbeat can be varied. With increasing number of lines obtained, the scan becomes shorter (used for breath-holding for example) since fewer heartbeats are needed to create the image. The disadvantage is that even though the entire scan time is shorter, the amount of time scanning per heartbeat is longer; this may not “freeze” the motion of the heart and blurring will occur.

- Combining the last two concepts above, retrospective gating creates an “interpolated” image in between measured images; placing more than one “interpolated” image in between two measured images decreases the accuracy of the data.

**A note on the challenges of CMR for ventricular function and blood flow in infants and children**

As with any imaging modality, and CMR is no exception, tradeoffs are made in the technical part of the imaging to make it quicker and simpler. For example, spatial resolution can be increased at the cost of temporal resolution and visa versa. These tradeoffs deserve increased attention in infants and children since they require a high temporal resolution because of high heart rates as well as high spatial resolution because of their small size. These tradeoffs in the adult patient are much less of an issue because of the larger size and lower heart rates, but can only partially be taken advantage of in children. Adult patients can also hold their breath which allows for specialized sequences to be taken advantage of; since children under 7–9 years of age cannot usually cooperate and therefore need medication to remain motionless in the scanner (deep sedation or cardiac anesthesia), these specialized sequences are useless. Since this is the mainstay of adult CMR, different approaches need to be applied and special sequences developed to successfully image these small patients.

Assessment of ventricular function and blood flow by CMR must be interpreted in light of the considerations just mentioned. For example, since some small patients need to undergo deep sedation, ventricular performance parameters may be lower than if the patient was awake. In addition, because CMR approaches to the sedated patient include multiple “averaging” of imaging data, the function is “averaged” over a 30 second to 2 minute period and embedded in the image which is different from the “instantaneous” picture of the heart obtained from echocardiography or angiography; in that instance, the “averaging” must be done in the physician’s mind.
**Techniques used to assess ventricular function**

There are six general types of CMR which are in common use today in varying degrees to assess ventricular function (Figures 4.1 and 4.2):

1. General cine CMR [13]
2. Myocardial tissue tagging (e.g., SPAMM-SPatial Modulation of Magnetization) [14,15]
3. T2* for myocardial iron assessment [16]
4. Stress CMR [17]/coronary flow reserve [18]
5. Phase encoded velocity mapping techniques [19–21]
   - Blood
   - Myocardial velocimetry
6. Perfusion [22]/viability [23]

Only 1–4 will be discussed in any detail in this section. Phase encoded velocity mapping will be discussed in greater detail in the blood flow section; however, it is mentioned in this section because of the ability of the technique to obtain stroke volume, cardiac index and regurgitant fraction which are obvious parameters used to assess ventricular function. Myocardial velocimetry, however, will be discussed in greater detail for completeness. Myocardial perfusion and viability will be discussed in greater detail in Chapter 5, but it is included in this section since it has obvious implications for ventricular performance.

**General cine CMR [13]**

(Figures 4.1 and 4.2)

There are two types which are the “workhorse” of ventricular function—(a) steady state free precession (SSFP) (Figures 4.1–4.3) [24]; and (b) gradient echo/FLASH [13] (Figure 4.3, bottom images). Blood appears signal intense and other tissues appear less so. With SSFP imaging, the contrast between blood and tissue is greater than in gradient echo imaging, and relies on this for its excellent image quality. Images can be obtained fast (SSFP is quicker than the gradient echo sequences) and

![Figure 4.1 Cine CMR – steady state free precession. The top panels are typical short axis views and the bottom panels are typical long axis views; the left panels are systole and the right panels are diastole. The images are from a 10-year-old patient, after tetralogy of Fallot repair.](image-url)
PART I  The basics of cardiac MR

Multiple, sometimes hundreds of heartbeats, “averaging” the ventricular performance into the image. Contrast this to angiography and echocardiography for example, where each image represents the ventricular contraction at that instant in time. The physician reading the echocardiographic study or the angiographic images must then view the many heartbeats and “average” the ventricular performance in his or her mind. In CMR, this “averaging” is documented in the image itself!

A very important concept with cine involves the presence of turbulent blood flow which will display a signal void in the turbulent region [27] (Figure 4.3). This will be elaborated on more in the techniques utilized to assess blood flow section (p. 63), and is mentioned here simply because this is

Figure 4.2 Cine CMR used to assess cardiac index. These images are from the same patient in Figure 4.1, after tetralogy of Fallot repair. After axial images of the heart are obtained, multiple, contiguous, cine CMR images are performed across the entire ventricle at the same temporal resolution. The data are then sorted by time. By measuring end-diastolic (upper series of images) and end-systolic (lower series of images) endocardial borders on all slices, stroke volume can be calculated. Ventricular mass is the difference between the volumes traced by the endocardial border and the epicardial border. Only the left ventricle and endocardial borders are traced for simplicity. Short axis images on the right are near the base and ones on the left are near the apex.

Although there is a move to increase imaging speed, there are advantages in taking time. CMR is unique in that images are generally created over
CHAPTER 4 Assessment of ventricular function and blood flow

Shape, which are needed for calculations of volumes in other imaging modalities are avoided. This is extremely important in the ventricles found in CHD, with the highly variable shapes. In addition, it is usually when the ventricular shape changes to a form where geometric formulae do not apply (e.g., a failing ventricle with aortic stenosis, where it becomes rounded and globular in shape) where the clinician is most interested in assessing ventricular volume and mass. CMR has been validated and used to evaluate right and left ventricular geometry and performance in multiple studies including CHD [1–4,6].

(A) Assessment of wall motion: Using cine CMR, as with echocardiography, global and regional wall

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Figure 4.3 Cine CMR to visualize turbulent flow. The top two panels are steady state free precession images from a 7-year-old with mitral insufficiency; note the jet of turbulence in diastole (arrow) from the 4-chamber (left) and long axis views (right). The lower left and middle panels are gradient echo images of a 5-year-old with a bicuspid aortic valve and aortic insufficiency; note the central jet of aortic insufficiency in the 2-chamber view (left) and left anterior oblique view (middle). The lower right image is an off-axis gradient echo cine CMR view from an 8-year-old with a conoventricular ventricular septal defect – note the turbulence (loss of signal) across the defect (arrow). LV: left ventricle; RV: right ventricle.
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ejection fraction, wall thickening, etc. can be obtained.

(B) **Assessment of ventricular volumes, ejection phase parameters and mass (Figures 4.1, 4.2 and 4.3):** To obtain ventricular mass and volumes (with subsequent calculations of ejection fraction and cardiac index), multiple contiguous sets of cine CMR sequences are performed through the ventricle at the same temporal resolution (Figure 4.2). The following protocol is used:

1. Localizers are performed to locate the heart in the chest.
2. Anatomical survey: Two different approaches are used, one more time consuming than the other:
   - Preferably, a full set of contiguous transverse images are obtained as a general survey of
anatomy in order to interpret the functional information in the context of the anatomy. These images are used as localizers to set up for the short axis views needed to obtain ventricular volumes and mass.

- If time saving is important (this approach takes <1 minute), a set of ultrafast gradient echo or SSFP sequences are performed to define the short axis:
  a) From the localizers in #1, a plane perpendicular to the axial localizer intersecting the atrioventricular valve plane and the apex is obtained.
  b) A plane perpendicular to the localizers in a) intersecting the atrioventricular valve plane and the apex is again obtained. This will yield a 4-chamber view.
  c) The short axis views are obtained perpendicular to the long axis of the ventricle.

3. If the first approach in #2 is used, multiplanar reconstruction (MPR) for localization of the ventricular short axis is then performed which yields a more accurate definition of the short axis. MPR is a software package which can stack the axial images contiguously and subsequently “reslice” it in any plane. To obtain the short axis of the ventricle (Figures 4.1 and 4.2), the following method is used similar to the second approach in #2:
   - A short axis plane of the atrioventricular valves, viewing these structures en-face, is obtained.
   - From this short axis, atrioventricular valve plane, a second plane perpendicular to the center of both valves is created angled to varying degrees, from anterior/inferior to posterior/superior (in general).
   - From the newly created plane above, a third plane perpendicular to this resulting in a long axis view of the ventricle is used to find the apex of the heart.
   - From this long axis view, a plane bisecting the atrioventricular valve and apex will yield a “true 4-chamber view.”
   - From the 4-chamber view, short-axis slices are simply the plane parallel to the long axis of the ventricle.

4. Using FLASH or SSFP techniques, short axis slices extending from atrioventricular valve to apex are used to obtain ventricular volumes. Usually 8–12 short-axis levels are obtained at temporal resolutions of 15–30 phases throughout the cardiac cycle, depending upon the heart rate. It is important to note that a cine of the 4-chamber view should be obtained before embarking on a stack of short axis slices as the position of the atrioventricular valve plane at end-diastole is needed to obtain the full volume data set.

This entire part of the exam can be as short as 6–7 minutes including localizers; the actual short axis cines can be obtained, in certain instances, in one breath-hold. Three-dimensional cine sequences have even been developed where any plane can be prescribed, off line, from the cine data set.

The cine data are then sorted by time resulting in multiple full volume data sets. By tracing (done semiautomatically on most software systems) the endocardial borders on all images at the phases of interest (usually end-diastole, which is defined at the first phase after the R wave on the ECG trace and end-systole, which is usually defined as the phase with the smallest cavity area), ventricular volumes are obtained (Figure 4.2). This is the product of the measured areas and the slice thickness, with the results summed across all slice levels using Simpson’s rule [1]. Ejection phase indices are then calculated:

- Stroke volume is the difference in the cavity volumes at end-diastole and end-systole.
- Ejection fraction is stroke volume divided by the end-diastolic volume.
- Cardiac index is the product of stroke volume and the average heart rate during image acquisition divided by body surface area.
- Ventricular time-volume curves can be created (Figure 4.5).

By tracing the epicardial borders and performing the same exercise outlined above with the endocardial borders, total ventricular volume is obtained (i.e. ventricular mass and cavity volume). When ventricular cavity volume is subtracted from this total ventricular volume at end-diastole, ventricular mass is obtained.

(C) Assessment of valvar stenosis and insufficiency (Figure 4.3): See Techniques used in assessing blood flow (p. 63) for detailed delineation of how this is performed.
PART I  The basics of cardiac MR

Myocardial tissue tagging [14,15,28–31] (Figures 4.4, 4.6, 4.7)
A unique capability of CMR is the ability to purposely tag tissue magnetically, whether it be myocardium or blood. This is generally performed by modifying a cine CMR sequence on most machines (for myocardial tissue, some use spin-echo) which, by design prior to imaging, destroys all the spins in a given plane resulting in a linear signal void (dark lines – Figures 4.4, 4.6 and 4.7). For example, in Spatial Modulation of Magnetization (SPAMM for short, Figure 4.4), there are:

• multiple radiofrequency pulses of 130° separated in time and
• a series of gradient radiofrequency pulses which produce saturated spins (the hydrogen atoms become incapable of producing a signal) in two sets of parallel lines perpendicular to each other. Subsequently, a standard cine CMR sequence follows which divides the wall into “cubes of magnetization” as it has been termed. The rotation, translation and deformation of the cubes can then be tracked to assess wall strain, motion and regional wall thickening (Figures 4.4 and 4.7) in both 2- and 3-dimensions. As a side benefit, since the sequence is a gradient echo one, turbulent flow within the ventricle or across a valve will be observed. Tagging can be either systolic or diastolic (Figure 4.4) and images can be obtained with a high temporal resolution if necessary. High temporal resolution, however, tends to demonstrate stripe degradation and progressive image blurring as the number of acquired images increases, “smearing” the “cubes of magnetization” in the later images. This can be compensated for by performing a number of separate acquisitions with trigger delays at a given slice level (which increases the acquisition time) and combining the images and sorting them temporally. In practice, however, this is rarely necessary as 6–7 images in systole or diastole are sufficient to track the motion and deformation with little tag degradation. With 3-Tesla systems, stripe persistence is much greater than with 1.5 Tesla scanners and stripes may persist without degradation throughout the cardiac cycle.

Qualitatively, the “cubes of magnetization” can be imaged to view subtleties of regional wall motion abnormalities; however, detailed analysis yields more interesting results. There are two ways in which this is performed (Figure 4.6):

(A) Tracking the grid intersections: The initial step in analyzing SPAMM images is to track the magnetically tagged grid intersections on all phases which can be done manually or semi-automatically. A triangular grid is created for each image utilizing Delaunay triangulation which ensures uniform, non-overlapping triangles and the following analysis can then be performed:

• Strain analysis: Myocardial regional deformations can be characterized using finite strain analysis on the deforming triangles [32,33], the methodology of which has been validated in a phantom [34]. The mathematics of strain analysis is beyond the scope of this article and is outlined in the appendix in a previous investigation [3]. Strain tensors can be calculated and the local deformations can described by two principal strains, $E_1$, and the orientation of the principal axes of these strains. The first principal strain, $E_1$, is defined as the most negative strain and the second principal strain, $E_2$, which is orthogonal to $E_1$, is defined as the most positive strain. In diastole, for example, $E_1$ can be thought of as “radial thinning” strain whereas $E_2$, can be thought of as “circumferential lengthening” strain. The strains can then be mapped onto a grayscale (Figure 4.6) or color-coded and superimposed onto the anatomic image.

• Wall motion calculation: The center of each triangle can be used to calculate regional wall motion (linear as well as rotational movements) in relation to any point in space, which is usually the center of mass of the ventricular cavity. The mechanics of wall motion can be characterized by parameters such as wall twist (using angles), radial wall motion or wall thickening or thinning, which can be measured by the distance between “cubes of magnetization” as tracked through the phase of interest. Similar to strain data, wall motion data can be displayed graphically as well as quantitatively. In one type of graphical representation of systolic wall motion shown in the figure, “dots” represent the triangular centroid location at end-diastole and the “tails” represents the subsequent systolic motion. For the analysis, the
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Figure 4.5 Ventricular time-volume curve with calculated maximum volume rates of change (dV/dT) in systole and diastole. cc: cubic centimeters; LV: left ventricle; msec: millisecond; sec: second.

Figure 4.6 Analysis of tagged images. The intersection points of the tagged images can be used to track wall motion. In the upper left panel, a 2-year-old single left ventricle patient’s systolic motion is shown in short axis – the dots represent the starting point at end-diastole and the tails represent the subsequent motion in systole. Note the discoordinated motion with areas of twist and no twist (arrows). Strain components are derived from the deformation and coded into a grayscale or color scale map (middle). This scale is mapped onto the anatomic image (upper right) which is another single left ventricle patient and analysis performed by a software package called VIDA. “Brown” regions represent more strain while yellow regions represent less strain. The images in the lower left and right are the mid-short axis of the left ventricle of a patient with tetralogy of Fallot at end-diastole (ED) and mid-systole (MS) analyzed using a software package called HARP. “Blue” areas are regions of more strain.
myocardial wall is typically divided into anatomic regions.

(B) Analysis in k-space: The development of the ability to analyze tagged image in k-space has allowed a much more rapid analysis. HARP (Harmonic Phase imaging) [35], developed at the Johns Hopkins School of Medicine takes the tagged images and converts them into their 2-dimensional Fourier transform which has spectral peaks. When a single peak is isolated, the inverse Fourier transform can create a HARP image, which has imaginary and real components to it. Through analysis of these HARP images, ventricular strain and wall motion can be measured [36,37].

“One dimensional” tagging can also be performed where just one set of parallel lines are laid down on the myocardium (Figure 4.6) [38]. Some investigators have utilized this type of tagging for strain measurements and have obtained excellent results, similar to the SPAMM technique (Figure 4.4). This type of tagging has also found great utility in labeling relatively thin structures, such as the right ventricular myocardium with subsequent analysis of regional myocardial shortening both qualitatively as well as quantitatively. To be performed correctly for this purpose, the tags are laid down perpendicular to the direction of myocardial motion (e.g., in the “4-chamber” view, tags are laid down perpendicular to the long axis of the ventricle). For quantification, the distance between the signal poor areas of the tag are measured at end-diastole and end-systole, and a regional shortening fraction is obtained.

$T_2^*$ for myocardial iron assessment (Figure 4.8) [16]
The measurement of $T_2^*$ in the relaxation process of the protons can be used to assess the amount of myocardial iron present as well as the liver. The exact details of how this is performed are beyond the scope of this book. In brief, by obtaining multiple images at various echo times (TEs) utilizing a gradient echo sequence, the myocardium and liver become increasingly dark with longer TEs. Because iron is ferromagnetic, the magnetic properties of the myocardium and liver change with increasing iron concentration by decreasing the measured $T_2^*$ (which makes the myocardium even darker). Values <20 milliseconds are at risk for decreased ventricular function. The patient population that this is generally used for are those with thalassemia and sickle cell disease although this has been applied to a number of other patient populations with chronic anemia. Chelation therapy in these patients is generally modified using this information.

**Figure 4.7** One-dimensional myocardial tagging (stripes). Similar to grid tagging, 1-dimensional myocardial tagging lays down one set of parallel lines and allows for labeling thin myocardium such as the normal right ventricle (RV). This can be used to determine regional wall motion and a regional ejection fraction. The set of panels on the left are a 4-chamber view at end-diastole (ED) (top) and end-systole (ES) (bottom) of a 10-year-old who presented to rule out RV dysplasia. Note how the deformations can be visualized. The panels on the right demonstrate how a regional shortening fraction (the right numbers) and regional wall thickening (left numbers) may be measured on the RV free wall (top panel – end diastole; bottom panel – end systole).
**Stress CMR/coronary flow reserve**
This will not be discussed extensively since it has had limited use in pediatrics; however, the reader should be familiar with these techniques as they may come into more use in the future.

**Stress CMR [17]:** This is similar to other imaging techniques which utilize either physiologic (via exercise) or pharmacological methods to stress the patient and is indicated when the resting state does not accurately reflect the clinical state of the patient. Any of the above three CMR techniques (cine CMR, myocardial tissue tagging or phase encoded velocity mapping methods) are used in conjunction with stress, depending upon what the desired parameter to be measured, although generally, cine CMR in the short axis is the most important set of images. Typically, baseline imaging is obtained prior to the application of the stress and subsequently, imaging is performed again in the same views at or immediately following peak stress. Modifications of CMR parameters are performed to compensate for the increased heart rate and motion of the patient (if exercising). A comparison is then performed between the unstressed and stressed states to determine the effect of the stress on ventricular function. Regional wall motion abnormalities and changes in cardiac index, for example, may be assessed using this method. In the CMR scanner, exercise can be performed utilizing a non-ferromagnetic bicycle and less preferably, utilizing hand-grips. For pharmacological manipulation, dobutamine is generally used.

**Coronary flow reserve [18]:** The ability of the coronary artery system to increase its blood supply to due to increased metabolic demands is an important functional parameter. If mismatch occurs, ventricular function may suffer. Administration of the coronary vasodilator, adenosine, can be used to increase coronary blood flow. The difference between coronary blood flow with and without adenosine is one indicator of the ability of the heart to increase its blood supply. CMR, utilizing phase encoded velocity mapping of the blood (see section below), can be used to measure coronary blood flow in the native state as well as with adenosine administration. Some studies have used phase encoded velocity mapping of the coronary artery directly but measuring coronary venous flow via the coronary sinus has also been reported and can be easier to perform.

**Phase encoded velocity mapping techniques [19–21] (Figure 4.9)**
Both phase encoded velocity mapping of the blood and myocardial velocimetry (Figure 4.9) fall under this category and utilize the same principles. Blood phase encoded velocity mapping and the elementary physics of this technique will be discussed more extensively in the Blood flow section of this chapter (p. 63). It is mentioned in this section because of the ability of the technique to obtain stroke volume, cardiac index and regurgitant fraction which are obvious parameters used to assess ventricular function. Myocardial velocimetry, however, will be detailed in this section.

Myocardial velocimetry is the CMR equivalent to Doppler tissue imaging in echocardiography; myocardial tissue velocities can be recorded. In myocardial velocimetry, however, each pixel can encode velocities in three orthogonal planes hence creating a 3-dimensional velocity map of the myocardium (Figure 4.9). Because of this, myocardial velocimetry can determine 3-dimensional velocities in any part of the myocardium; with Doppler tissue imaging, only the myocardium traveling in the direction of the Doppler beam can be accurately recorded. Any deviation in direction of myocardial motion causes an error using the cosine of the angle. Both myocardial velocimetry in CMR and Doppler tissue imaging in echo suffer, however, from the same drawback – the velocity of a specific piece of myocardium is not measured, but rather a 3-dimensional space is identified and the velocity of myocardium moving into and out of that 3-dimensional space is measured. Only CMR myocardial tagging truly measures this non-invasively. Although new software is becoming available, myocardial velocimetry is not routinely used as it is difficult to analyze.

**Perfusion/viability**
This will not be discussed extensively since it will be covered in detail in Chapter 5.

**Perfusion (Figure 4.10) [22]:** Regional myocardial perfusion is an obvious important parameter in
viability (Figure 4.11) [23]: Infarcted myocardium is more of an issue in adults than it is in CHD. Nevertheless, native lesions such as anomalous left coronary artery from the pulmonary artery or operations which scar the myocardium (e.g. most repaired tetralogy of Fallot) may manifest myocardial scarring. Gadolinium is avidly taken up by scarred myocardium and can remain in the scarred tissue for an extended period of time while it is subsequently “washed” out by coronary blood flow in perfused myocardium. The signal intensity–time curves separate and the infarcted myocardium curve remains highly signal intense after 5–10 minutes whereas normal myocardium becomes much less so. Hence the alternative name of “delayed enhancement CMR.” CMR pulse sequences take advantage of this property to image infarcted myocardium which is unique in non-invasive imaging. This infarcted myocardium is signal intense while the non-infarcted myocardium is dark (signal intensity differences up to 500%). The technique has been shown to accurately delineate the presence, extent and location of acute and chronic myocardial infarction. This imaging can be performed in two ways (see Chapter 5 for details):

- Using a delay time (typically called the TI time) which generally falls in the range of 250–350 msec...
in the sequence. This delay time must be chosen correctly to optimize the difference between infarcted and normal myocardium.

- Phase sensitive reconstruction of the inversion recovery data which will provide consistent contrast between normal and scarred tissue over a wide range of TIs.

**Techniques used to assess blood flow**

There are four general types of CMR which are in common use today in varying degrees to assess blood flow (Figures 4.12–4.16):

1. phase encoded velocity mapping of blood [19,20]
   - through plane (Figure 4.12 and 4.14)
   - in-plane (Figure 4.13)

2. blood tagging (Figure 4.15) [39]
3. cine CMR (Figure 4.3) [13]
4. time resolved gadolinium (Figure 4.16).

**Phase encoded velocity mapping of blood (Figures 4.12 and 4.14)**

This technique may be used to measure flow and velocity in any blood vessel; for example, relative flows to each lung may be obtained by utilizing through-plane velocity maps across the cross-section of the right and left pulmonary arteries. An internal check is using a through-plane velocity map across the cross-section of the main pulmonary artery and ensuring that the blood flow to the right and left pulmonary arteries equals the blood flow in the main pulmonary artery. It is also common to utilize phase encoded velocity mapping
The principle underlying phase encoded velocity mapping will be delineated in this chapter in a simplified approach. The interested reader is referred to major CMR textbooks for greater detail. In this discussion, it is key to understanding what “phase” means in the term “phase encoded velocity mapping.” When tissue is excited by energy, the subsequent signal that gets released when the protons relax (for example, a sine wave) can be described by its frequency, its amplitude (the strength of the signal) and its phase (where in the sine wave cycle the signal is at a given period of time). Two waves can have the same frequency and amplitude but be in different points in their cycle (e.g., identical waves but shifted in time); they are out of phase. Another way to understand this is that the same part of each cycle occurs at a different period of time (e.g., the peak of sine wave “A” occurs prior to the peak of sine wave “B”).

The principle underlying phase encoded (also called phase shift or phase contrast) CMR very simply is that moving tissue within a magnetic field changes phase after a radiofrequency pulse imparts energy to it. More exactly, whenever anything moves along the axis of an applied gradient, the phase of the spinning vectors in that object becomes altered relative to the stationary object. This selec-
Figure 4.11 Viability. The patient is a 1-year-old who had a neonatal insult and infarcted apical and some mid-short axis myocardium. Waiting 5–10 minutes after gadolinium injection, the delayed enhancement sequence is performed – viable myocardium is black as tissue perfusion “washes out” the gadolinium. Infarcted myocardium maintains the gadolinium longer and is signal intense (arrows). The top left image is an off-axis 4-chamber view and the top right image is a short axis view demonstrating the infarcted regions. A specialized technique called “phase sensitive” viability imaging (lower image) does not require the 5–10 minute waiting time.

...labels... moving tissue; the technique utilizes a “bipolar” radiofrequency pulse which goes from positive to negative and then from negative to positive performed with two sequences back to back. The other principle to understand is that before any radiofrequency pulse is applied, protons are tilted depending upon where they are in the magnetic gradient. Therefore, when the first radiofrequency pulse is applied, both stationary and moving tissue accumulate phase (are further tilted); when the second radiofrequency pulse of equal but opposite intensity is applied immediately afterwards, stationary tissue lose their phase and accumulate a net phase of zero (their tilt goes back to their original position since they experienced an equal and opposite radiofrequency pulse and haven’t moved position). Moving tissue, however, does not revert to their original tilt since they have moved in the magnetic field and are at a different position of the magnetic gradient. They therefore accumulate a phase shift. To summarize, this will yield a zero phase change for stationary objects in both sequences whereas there will be a net accumulation of phase in moving tissue. By subtracting, pixel by pixel, the phases of one sequence from the other, background phase changes of stationary objects are cancelled out and the phase shift of the moving tissue is amplified. Then, usually, the “phase difference” method is used to map the phase shift angles into signal intensities. Flow is calculated by the formula:

\[ \Delta \text{phase} = g \times v \times T \times A_g \]

where \( g \) = gyromagnetic ratio, \( v \) = velocity, \( T \) = duration of the gradient pulse and \( A_g \) is the area of each lobe of the gradient pulse.
Figure 4.12 Through plane phase encoded velocity mapping. This technique is used to measure flow as well as velocity into and out of the plane of the image. Both an amplitude (anatomic) as well as phase image (flow encoded image) are obtained routinely. Directionality is encoded as either signal intense (white) or signal poor (black). The left panel is an amplitude image at the level of the aortic (Ao) valve in a patient with a bicuspid Ao valve which is plainly seen with fusion of the right and left coronary cusps. The middle panel is a phase encoded image which measures velocity into and out of the imaging plane; if velocities exceed the VENC (in this case 1.5 m/s) which is the maximum velocity that can be measured (similar but not exactly like the Nyquist limit in Doppler echocardiography), directionality in the blood appears reversed (i.e., white in the middle of the black flow). The right panel is the same as the middle panel except the VENC was increased to 2.75 m/s; note how all the blood in aorta is black.

Figure 4.13 In plane phase encoded velocity mapping. This technique is used to measure velocity in the plane of the image similar to Doppler echocardiography. Similar to through plane, both an amplitude (anatomic) as well as phase image (flow encoded image) are obtained routinely. Directionality is encoded as either signal intense (white) or signal poor (black). The images are from a 12-year-old with a native coarctation of the aorta. The left and middle panels are amplitude images at end-diastole (left) and mid-systole (middle); note the turbulence (loss of signal) at the coarctation site (arrow). The right panel is the phase image where white is encoded as flow towards the head and black is flow towards the feet. Note the thin black jet at the coarctation site (arrow) corresponding to the turbulence in the middle panel.
CHAPTER 4 Assessment of ventricular function and blood flow

Figure 4.14 Time-volume curves using phase encoded velocity mapping. (a) Aortic (Ao) and pulmonary (MPA) curves of a patient with a ventricular septal defect and a pulmonary (Qp) to systemic (Qs) flow ratio of 2.8; time is in milliseconds (ms). (b) Inflow patterns across the mitral valve in a normal individual (upper left panel), a 4 year old after anomalous coronary artery repair with poor ventricular function (upper right panel) and a 6 year old single ventricle patient after Fontan (lower panel). Note the loss of the normal E/A wave ratio in the upper right and lower panels. cc: cubic centimeters; ml: milliliters; sec: second; vs: versus.
Figure 4.15 Blood tagging. Placing a saturation band (arrows) over blood destroys all the spins in the blood and turns it dark on the image thereby "labeling" or "tagging" the blood. The patient is a 3-year-old with an incomplete atrioventricular canal (ostium primum atrial septal defect). The top left panel demonstrates the defect (*) and volume overload of the right atrium (RA) and ventricle (RV). The upper right panel shows the image once a saturation band is placed on the RA – white blood from the left atrium (LA) is seen to cross the defect shunting left to right. The lower left panel shows the image once a saturation band is placed on the LA – dark blood from the left atrium (LA) is seen to cross the defect confirming shunting left to right.

A VENC (velocity encoding) is used to tailor the strength of the radiofrequency pulse to the anticipated velocities to be measured (the equivalent in echocardiography would be the Nyquist limit). Using the VENC and the signal intensity, the velocity of moving tissue in each pixel can then be encoded. This can occur with either blood (hence blood phase encoded velocity mapping) or with myocardial tissue (hence myocardial velocimetry described above).

There are two ways to encode velocity in the image spatially:
- In "through plane" phase encoded velocity mapping, each pixel encodes velocity into and out of the plane of the image (Figure 4.12).
- In "in-plane" phase encoded velocity mapping, similar to Doppler echocardiography, velocities are encoded in the plane of the image and not into and out of it (Figure 4.13). Unlike Doppler, the velocities are encoded in either the Y- or X-direction of the image.

This type of flow encoding is advantageous in that each pixel can encode velocity in three orthogonal planes. Motion in one direction is mapped onto the anatomic image as increased signal intensity while motion in the other direction appears dark and signal poor and stationary tissue appears gray. Color can be added to make it similar to Doppler echocardiography.

To determine the flow utilizing through plane phase encoded velocity mapping, the cross-sectional area of the vessel perpendicular to flow is used. Software enables the identification of the region of interest on the anatomic or "magnitude"
should be taken, of course, to make sure that it is indeed the proper cross-sectional area; angled views not perpendicular to flow or views at a level not where the maximum flow would be will underestimate the maximum velocity. This is similar to Doppler echocardiography where Doppler interrogation angled obliquely to the jet of interest or the 2-D sector not in the area of maximum velocity will underestimate the maximum velocity.

Encoding velocity parallel to flow (“in-plane” flow encoding, Figure 4.13) is predominantly used to measure peak flow velocities which is similar to what Doppler echocardiography measures. It has an advantage over the through plane technique in measuring maximum velocities in that velocities can be measured all along a jet of interest in the direction the jet is pointing. The jet is aligned by rotating the entire field of view to make one side exactly parallel to the jet. The peak velocities can then be translated into pressure gradients via the simplified Bernoulli equation \( \Delta P = 4v^2 \) where \( \Delta P \) is the pressure gradient in mm of mercury and

![Figure 4.16 Time resolved 3-dimensional gadolinium imaging. The upper panels are three phases of gadolinium flow from a patient with double outlet right ventricle (S,L,L) after a double switch (arterial switch and Senning operation). The pulmonary (left), the systemic arterial (middle) and late recirculation (right) phases are shown. Note how well the perfusion to both lungs is visualized and how equal it is. Arrows on the right panel identify the proximal portions of the upper and lower limbs of the systemic venous pathway. By contrast, the lower panel is a pulmonary phase from a patient with tetralogy of Fallot after repair – note the diminutive left pulmonary artery (arrow).](image-url)
\( v \) is the peak velocity in meters/second). The phase maps on present day scanners can give a temporal resolution of about 15–20 milliseconds.

Both in-plane and through plane velocity mapping reliability is a function of a few factors such as slice thickness ("partial volume" effects may induce inaccuracies in velocity calculations) and the angle of the jet (the jet needs to aligned perpendicular to the direction of phase encoding, similar in some sense, to Doppler flow measurements).

These techniques have a few limitations. If the VENC is not chosen properly, errors may occur. For example, if the VENC chosen (maximum velocity detectable) is too low, velocities in the patient will exceed the ability of the CMR scanner to encode them, similar to aliasing and exceeding the Nyquist limit in Doppler echocardiography. If the VENC chosen is too high for the velocity measurement, the dynamic range will be lost and a less accurate measurement will be obtained. This is similar to Doppler interrogation of venous flow where the spectral scale is set to a maximum velocity of 5 meters/second. Another way to think of this kind of inaccuracy is the difference when measuring a ½ cup of fluid (4 oz) in an 6 oz measuring cup (appropriate setting of the VENC or Nyquist limit) versus a gallon measuring cup (inappropriate setting of the VENC or Nyquist limit).

Phase encoded velocity mapping of blood has many applications in CHD which can be divided into three different categories – (1) flow quantification; (2) velocity measurement; and (3) flow visualization:

1. **Flow quantification**

   **(A)** Cardiac output: Since the vast majority of moderate to severe CHD requires some form of intervention, either surgical or catheter based, the measurement of CO can be an important parameter to assess (e.g., single ventricles, the right ventricle of a patient with transposition of the great arteries after atrial inversion operation such as Senning or Mustard procedures. This can be used as an internal check to the ventricular volume measurements.

   **(B)** Regurgitant volumes and regurgitant fractions are important parameters in the assessment of ventricular function and blood flow in patients pre- and postoperatively in CHD (e.g., in a patient after tetralogy of Fallot repair with a transan- nular patch [40] or in a patient with a bicuspid aortic valve and severe aortic insufficiency). The regurgitant fraction of a semilunar valve is easily obtained by utilizing through plane phase encoded velocity mapping just above the semilunar valve and measuring the forward and reverse area under the flow-time curve. The regur- gitant fraction is simply the area under the reverse flow (regurgitant volume) divided by the area under the forward flow (forward volume) multiplied by 100. To obtain an atroventricular valve regurgitant fraction, a combination of techniques are used; cine CMR to measure the stroke volume of the ventricle and phase encoded velocity mapping to measure the amount of forward flow through the semilunar valve. The difference between the two (assuming no semilunar valve insufficiency) is the regurgitant volume of the atroventricular valve. Antegrade flow across the leaky atroventricular valve and forward flow across the associated semilunar valve can be used as well (but less reliably).

   **(C)** Shunt flow can be calculated simply by placing velocity maps across the aorta and main pulmonary artery and measuring flow (e.g., Qp/Qs) [41]. Measuring flow in both branch pulmonary arteries can add an internal check on the amount of pulmo- nary blood flow.

   **(D)** Relative flow to each lung [42]: Maldistribution of flow to right and left lungs is a prominent feature of a number of CHD lesions (e.g., after repair of trans- position of the great arteries utilizing the Le Compte maneuver, the left pulmonary artery may get “stretched” as it crosses the ascending aorta and cause decreased flow to that lung). Relative flow to each lung is generally assessed utilizing phase encoded velocity mapping in each branch pulmonary artery. The flow in the main pulmo- nary artery must equal the sum of the flows to each lung in the absence of collaterals and is used as an internal check. Care must be taken to place the VMAP in the branch pulmonary artery proximal to the takeoff
of the first branches to ensure this blood flow is included.

(E) Collateral flow [43]: In patients with relatively long standing coarctation of the aorta, for example, aortic collaterals may develop to bypass the obstructed segment. The amount of collateral flow can be determined by CMR by placing a phase encoded velocity map across the aorta just distal to the coarctation site and one across the aorta at the level of the diaphragm. In the normal circumstance, flow at each level will be very similar or the flow at the level of the diaphragm slightly lower (because of flow to the intercostal arteries). In the presence of a coarctation of the aorta with collaterals, however, flow just distal to the coarctation site in the aorta will be lower than flow in the aorta at the level of the diaphragm since collateral flow will present in the latter and not in the former. This technique can, therefore, quantify this amount of collateral flow.

2. Velocity measurements

(A) Pressure gradients: Valvar stenosis along with great artery stenosis occur in a myriad of CHD lesions where it is important for the clinician to determine pressure gradients (e.g., bicuspid aortic valve, coarctation of the aorta). The determination of gradients by CMR is similar to the way it is determined by Doppler echocardiography, using the simplified Bernoulli equation. A maximum velocity is measured, typically in the vena contracta, and the gradient is simply the product of 4 and the velocity (in meters/second) squared. Measurement of maximum velocities may be performed in two ways: (i) “in-plane” velocity mapping directed parallel to the obstruction to flow and (ii) “through plane” velocity mapping perpendicular to flow. Both have their strengths and weaknesses.

3. Flow visualization

(A) Septal defects: Atrial or ventricular septal defects may be identified by in plane phase encoded velocity mapping techniques. Orienting the velocity encoding direction in the direction of flow is necessary for successful visualization. Flow is bright in one direction and dark in the opposite direction.

(B) Flow directionality: Isolation of the subclavian arteries may occur naturally or may be iatrogenically created (e.g., a subclavian flap angioplasty to repair coarctation of the aorta). The subclavian usually obtains blood by retrograde flow in the vertebral artery along with some collateral flow. Some patients may be at risk for a “subclavian steal” phenomenon in this instance and CMR flow techniques may be used to identify retrograde flow in the vertebral arteries. In the normal situation, when a “through plane” phase encoded velocity map is placed in the neck, both carotid and vertebral arteries will be labeled as either bright or black on the images. In cases where there is isolation of a subclavian artery and flow in the ipsilateral vertebral artery is retrograde, three of the four head vessels (carotid and vertebral arteries) will be encoded in one direction and the ipsilateral vertebral artery will be encoded in the opposite direction, proving the physiology.

Blood tagging [39] (Figure 4.15)

As noted in the previous section on myocardial tissue tagging, CMR is unique in its ability to tag tissue non-invasively. Blood may be tagged the same way myocardial tissue is tagged. This type of tagging has been called “bolus” tagging or more generally blood tagging and is a gradient echo sequence similar to myocardial tissue tagging. Instead of a “grid” or a series of parallel lines (1-D tagging) being laid down on the myocardium, it utilizes a radiofrequency pulse to produce saturated spins along a single line of variable thickness designated by the user (a black stripe on the image) across a blood vessel or, for example, a heart chamber. Unlike myocardial tissue tagging, however, a saturation pulse precedes each phase of the cardiac cycle; with myocardial tissue tagging, the saturation pulse is only applied immediately preceding the first phase. The tag on the blood from the previous phase generally fades away prior to the next phase. Blood flow displaces this band of saturation, whereas stationary structures (e.g. chest wall and spine) maintain the saturation band’s original position (Figure 4.15). Each image...
represents blood displacement between tagging and image acquisition.

If a thin band is placed over a blood vessel, velocity can be calculated at any point along the band by measuring the displacement of the saturation band relative to the stationary structures; the time between saturation band being placed and image creation is known and it is a simple calculation to divide the distance of tag displacement by the time. Essentially, this bolus tagging enables direct visualization of velocity profiles in a blood vessel in-vivo. Similar to phase encoded velocity mapping of the blood, individual regions of the blood vessel may be studied to evaluate flow dynamics. These flow dynamics are not limited to the primary flow patterns (e.g., the forward or reverse flow); even secondary flow patterns (blood flow perpendicular to the primary flow) have been visualized by placing the saturation band on the blood vessel in long axis, parallel to the direction of blood flow – and then the blood vessel is imaged perpendicular to the blood flow (e.g., across the cross-sectional area of the vessel). The resultant cine demonstrates blood flow “twisting” (as in the aorta) or lack thereof in the axial plane, representing the secondary flow pattern. The rotation of the blood can be quantified by measuring the angle between bands on successive images.

Blood tagging can be used in many imaginative ways to demonstrate where blood is coming from or going to. For example, it can be used to demonstrate flow across an intracardiac shunt (Figure 4.15). If a thick saturation band is placed across the left atrium in a patient with an atrial septal defect, dark, signal poor “tagged” blood may be seen to cross into the right atrium. Conversely, if there is right to left flow, bright, signal intense blood may be visualized to cross to the left atrium. This technique can also be used, for example, to demonstrate retrograde flow in the vertebral arteries in patients with isolated subclavian arteries; a saturation band is placed over the vertebral artery and the dark, “tagged” blood is seen to enter the subclavian via a retrograde course; if the subclavian artery was not isolated, bright blood would be seen to enter the dark, vertebral region.

This technique was also used to demonstrate, in-vivo, the relative contribution of flow from each cavae to each pulmonary artery in single ventricle patients who underwent the Fontan procedure; after acquiring a standard cine of the branch pulmonary arteries where it meets the superior vena cava and Fontan baffle in an off-axis transverse plane, a saturation band was placed over blood originating from the inferior vena cava and the cine of the branch pulmonary arteries run again. Finally, a saturation band was placed over blood originating from the superior vena cava and a third cine of the branch pulmonary arteries run. Cines with saturations bands placed demonstrated decreased blood signal relative to the cines without the saturation bands; the decreased signal intensity from cines with and without saturation bands were proportional to the contribution of flow to each branch pulmonary artery from each cavae.

**Cine CMR (Figure 4.3)**
Visualization of the flow void in turbulent flow by cine can be used to assess valvular insufficiency or the presence of ventricular septal defects. Valvular insufficiency, for example, can be graded similar to color Doppler echocardiography. Multiple views should be obtained for both the flow void of valvar insufficiency as well as stenosis to optimize the ability to visualize this flow void; once the insufficiency or stenosis is visualized, slices in orthogonal directions are mandatory to fully evaluate the jet. This is simply qualitative and volumetric assessment of the regurgitant flow as well as the maximum velocity of the insufficiency and stenosis jet should be calculated using phase encoded velocity techniques. Caution must be used, however, when using the signal void to grade the regurgitation or stenosis. The size of the flow void is a function of a number of factors, including, for example, the echo time (TE), where longer TEs increase the size of the signal void. The rough parallel in echocardiography would be adjusting the Nyquist limit or the gain on color Doppler interrogation of the valve. In addition, steady state free precession imaging may underestimate the flow void seen on FLASH sequences and it is always best, if a flow void is present on steady state free precession imaging, to confirm its size on FLASH sequences.

**Time resolved 3-dimensional gadolinium (Figure 4.16)**
As noted previously, contrast enhanced CMR is the topic of another chapter but to be complete, it
should be noted that time resolved gadolinium imaging can be used to assess perfusion not only to the ventricle (perfusion imaging in the ventricular function section) but to the lungs as well as assessing shunts. As the contrast is injected, T1 weighted 3-dimensional images are obtained which can be as fast as less than a second per data set. The contrast is followed throughout the cardiovascular system. Absence of increased signal intensity in one part of the lung would indicate obstruction, possibly due to a pulmonary embolus. Right to left intracardiac shunts will also be plainly obvious.

**Conclusion**

CMR has evolved over the course of many years into an extremely useful tool for evaluation of physiology and function in many types of CHD, both in the pre-operative and post-operative setting. This chapter has only touched on some of the abilities of CMR in this area; however, it serves as a launching pad for both clinical and research applications. This chapter has not touched on applications in ventricular function and blood flow such as functional fetal CMR, interventional CMR or cardiac spectroscopy. Future advances in MRI technology such as oxygen sensitive MRI (detection of the oxygen content of tissues) [44], will certainly add even further understanding in the future into the physiology and function in CHD.

**References**


CHAPTER 5
Contrast cardiac MR – anatomy, physiology, viability and perfusion

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Introduction
Magnetic resonance imaging (MRI) has been an important diagnostic imaging tool to evaluate congenital heart disease for over two decades. In that time significant advances have occurred in both the hardware and software responsible for acquiring the images, as well as post-processing and analysis. Magnetic resonance imaging initially was directed at the qualitative evaluation of anatomic abnormalities of the cardiac chambers, pericardium, and arterial and venous great vessels. Subsequently, further technological advances have made magnetic resonance imaging a technique capable of evaluating the physiological parameters associated with congenital heart disease, and more recently providing greater insight into tissue characterization. Magnetic resonance imaging is particularly beneficial in assessing the anatomic abnormalities in congenital heart disease as it is a non-invasive and nearly risk-free diagnostic tool. The lack of ionizing radiation is of particular importance in congenital heart disease as these children may otherwise be exposed to repetitive procedures involving ionizing radiation with the potential for carcinogenesis.

This chapter will focus on the contribution that contrast enhancement plays in the pulse sequences employed in the imaging of congenital heart disease. Primary attention will be directed to the role of contrast enhanced magnetic resonance angiography in illuminating both the anatomy and physiology in congenital heart disease. In addition, the more recent introduction of delayed-enhancement imaging for the detection of myocardial viability and myocardial fibrosis, and contrast enhanced myocardial perfusion for ischemia detection will be explored.

Contrast enhanced magnetic resonance angiography

Technique
Contrast-enhanced magnetic resonance angiography is a “bright-blood” technique that produces a “luminogram” of the vasculature of interest. Gadolinium contrast agent is given intravenously and three-dimensional (3D) T1-weighted datasets are acquired, which are then reconstructed into volumetric displays using software packages in post-processing workstations. Data sets are typically acquired during repetitive breath-holds in adults, teenagers, and cooperative children, but also may be acquired during free breathing as needed in infants, younger children, and patients unable to perform breath-holds. These magnetic resonance angiograms are used to assess the course, caliber, contour, and relationships of the vascular tree (i.e., morphologic information), yet may also be acquired in rapid succession in an effort to determine functional information about shunts and blood flow [1].

Technological and methodological advances have provided the impetus for adoption of 3D...
contrast-enhanced magnetic resonance angiography for evaluating congenital heart disease in routine clinical practice. These include the introduction of: (a) high performance gradient hardware; (b) real-time magnetic resonance video fluoroscopy; (c) centric k-space encoding; and (d) parallel acquisition techniques.

- High performance gradient hardware has reduced repetition times (TR) and echo times (TE) to a few milliseconds or less, making it possible to acquire a high resolution contrast-enhanced angiographic volume, or even multiple volumetric acquisitions, within a breath-hold.
- Real-time magnetic resonance fluoroscopic monitoring provides a visual roadmap when timing the arrival of the gadolinium contrast agent bolus in the target vasculature. Such timing eliminates guesswork in timing, or the necessity for a test bolus acquisition. This may be particularly valuable in smaller patients when weight based administration of gadolinium contrast agent allows only small aliquots of contrast for use.
- Centric k-space encoding acquisition schemes allow the precise timing of peak contrast enhancement to correspond with the low-frequency, high contrast portion of the magnetic resonance angiogram acquisition. These k-space encoding schemes capture the arterial enhancement associated with the first pass of the gadolinium contrast, with minimal venous contamination even in scans with long acquisition times.
- Parallel acquisition techniques (i.e., SENSE, SMASH, and their variants) can dramatically accelerate data acquisition for almost any MRI pulse sequence, and, as a result, dramatically reduce acquisition time. There are some limitations with these techniques related to signal loss inherent to the data acquisition process, and may be particularly problematic with higher acceleration factors. These techniques are therefore most advantageous for acquisitions with high signal-to-noise ratios such as contrast enhanced magnetic resonance angiography [2].

These developments have made breath-held 3D contrast-enhanced magnetic resonance angiography a routine, non-invasive tool for assessing vascular morphology in adults with and without congenital heart disease, and has only more recently found increasing clinical use for evaluating the vascular morphology in pediatric patients. The rationale for this, alluded to above, is that many of the benefits of the improved techniques accrued during suspended respiration acquisitions. However, most infants and younger children are sedated and can not hold their breath during the contrast enhanced magnetic resonance angiographic acquisition, making it necessary to acquire data during free breathing.

In infants and younger pediatric patients, as a result of weight-based contrast restrictions, the volume of gadolinium contrast agent available for use is often only a few milliliters. If a test bolus is used it may represent a disproportionately large fraction of the total contrast dose, therefore limiting the amount of contrast used, and thus the amount of increased signal seen in the magnetic resonance angiogram.

Further, as a result of faster heart rates and circulation times, pediatric patient populations frequently have faster, but also more variable (and therefore less predictable) arterial-to-venous transit times. Thus, the interaction of a small bolus volume and rapid circulation places close time demands on accurately capturing the arterial phase of the contrast bolus. These factors in combination impose more stringent requirements on pediatric imaging, both from the scanner hardware and user interaction perspectives.

However, from at least one perspective pediatric vascular imaging is somewhat less technically demanding than in adults, namely, in the area of spatial resolution. In adults, when evaluating thoraco-abdominal vessels, there is often a critical need for high spatial resolution (preferably sub millimeter in-plane resolution) to evaluate the precise degree of stenosis in a branch vessel, such as a renal artery. In the pediatric patient population, however, the clinical question is not the evaluation of atherosclerotic disease, but the visualization of thoraco-abdominal vascular structures and branch vessels (and, in many cases, both arteries and veins), their inter-connections, and congenital vascular malformations. In the case of stenotic vessels precise quantification of the degree of stenosis typically is not as critical as it is in adults. As a result, the demands on spatial resolution in general may be less stringent than in adults. Contrast-enhanced magnetic resonance angiograms can thus be tailored to have more modest
spatial resolution while still maintaining rapid acquisitions necessary for the faster and less predictable circulation times.

Nonetheless, there are scenarios where maintaining spatial resolution is preferred, such as in identifying the presence and course, as well as stenoses involving aortopulmonary collateral arteries in patients with severe tetralogy of Fallot or pulmonic atresia. Neonates and infants, whose structures are inherently smaller, also need high spatial resolution. This may be problematic in some as data acquisition during free breathing is routine in pediatric imaging, and is common when imaging children younger than seven or eight years of age. Methods such as respiratory triggering, multiple number of excitations or averages, respiratory ordered phase encoding, and navigator echoes are all approaches employed to combat respiratory motion and therefore improve effective spatial resolution, though are not equally applicable for a first pass contrast-enhanced magnetic resonance angiogram.

Parallel acquisition techniques (SENSE/SMASH) are seeing an increased role and advantage when spatial resolution and speed of acquisition are paramount. For a given spatial resolution and coverage, SENSE/SMASH reduces the data acquisition time by a factor proportional to the acceleration factor, but does so without sacrificing the original spatial resolution [2,3]. In particular, Weiger et al. have shown in adults that SENSE/SMASH is particularly effective for contrast-enhanced magnetic resonance angiography, where the loss of signal-to-noise ratio inherent in the SENSE/SMASH acquisition can be partially offset by the intrinsic increase in signal-to-noise ratio associated with contrast enhancement [2]. In adults, time resolved contrast-enhanced magnetic resonance angiography also has been proposed as a means to obtain reliable arterial-venous separation without the need for bolus timing – particularly in imaging vasculature with a rapid circulation time (Figure 5.1). This same advantage in adults is available to the pediatric population where the free breathing, time resolved approach afforded by SENSE/SMASH techniques offers several practical advantages in imaging the great vessels and branch vessels. First and foremost the spatial resolution of the acquisition is maintained, or can even be improved. In addition, further benefits accrue by a reduction in motion artifact. For example, if a standard acquisition requires eight seconds, the acquired data set includes patient motion occurring over the entire 8-second duration. In contrast, with a parallel acceleration factor of 2 the acquired data would include only the motion artifacts occurring over a four second duration reducing the blurring incurred.

**Selected applications for magnetic resonance angiography**

**Aorta**

MRI is routine in the evaluation of great artery anomalies in infants and young children. In patients with prior surgery, and in teenagers and adults, obtaining adequate acoustic windows may be more problematic, and MRI is the preferred technique for accurate and non-invasive evaluation (Figures 5.2 and 5.3).

Coarctation of the aorta is the most frequently encountered abnormality of the great vessels and has been studied extensively using MRI. Spin-echo images typically are acquired in the axial plane, and oblique sagittal planes to include the ascending and descending thoracic aorta, arch, and arch branch vessels. When collateral vessels are present they may be seen as circular or serpentine signal voids in the mediastinum and paraspinous region. Alternatively and more commonly, contrast-enhanced magnetic resonance angiography is used to provide a complete 3D assessment of the aorta, site of coarctation, and extent of collateral arteries.

**Pulmonary arteries**

Pulmonic stenosis, whether valvar, supravalvar, or peripheral is a frequent feature of congenital heart disease in lesions such as tetralogy of Fallot, William syndrome, and hypoplastic left heart. Similar to aortic abnormalities, MRI is routine in infants and children, while in older children and adults, contrast enhanced magnetic resonance angiography has gained increasing appeal as it is not limited by suboptimal acoustic windows, intervening lung tissue, etc.

Peripheral pulmonic stenosis can be demonstrated with spin-echo images, however, current practice typically employs contrast-enhanced magnetic resonance angiography or newer 3D volumetric navigator-gated steady-state free-precession
Figure 5.1 The four images represent maximum intensity projection (MIP) images of individual, six-second acquisitions from a contrast enhanced magnetic resonance angiogram in a 25-year-old male with congenitally corrected transposition of the great arteries (CC-TGA). (a) The upper left image demonstrates the pulmonary phase where the right ventricle and pulmonary arteries are filled with gadolinium-chelate contrast agent. (b) The upper right image represents the intermediate phase of both pulmonary and left heart/aortic phase. (c) The lower left image demonstrates the left heart/aorta phase exclusively. In this image the pulmonary veins are better seen, as they are unobscured by the pulmonary arteries from the contrast that has already passed through the pulmonary arterial side. Note the right-sided descending aorta, and right-sided aortic arch and descending thoracic aorta, and mirror image arch vessel branching in this patient with CC-TGA. (d) The lower right image reveals early venous return from the head with contrast enhancement of both internal jugular veins and brachiocephalic veins, which then drain into the left sided superior vena cava.
acquisitions to rapidly and precisely delineate the main and branch pulmonary arteries. Contrast enhanced magnetic resonance angiograms easily identify areas of pulmonary artery aneurysm formation and stenosis [4] (Figure 5.4).

**Aortopulmonary collateral arteries**

Aortopulmonary collateral arteries typically develop in the setting of severe pulmonic stenosis or pulmonic atresia. These collateral vessels serve as the primary source of blood flow to the pulmonary parenchyma (Figure 5.5). However, it is critical to map all of the blood supply accurately including residual antegrade flow through the pulmonic valve (if present), significant bronchial arteries, or other shunts that may be present. Echocardiography is inadequate for this assessment for obvious reasons, and invasive catheter angiography traditionally has been performed preoperatively to provide this information. Recently, Geva et al. evaluated 32 such patients with contrast enhanced magnetic resonance angiography with direct comparison to invasive catheter angiography with good results [5]. They noted that contrast enhanced magnetic angiography determined all sources of pulmonary blood supply in these patients, and accurately identified the presence, size, and course of the aortopulmonary collateral arteries. As a result, the contrast enhanced magnetic resonance angiogram reasonably could be
Figure 5.3 Two images from a contrast enhanced magnetic resonance angiogram in a patient with CC-TGA (same patient as in Figure 5.1) (on the left an anterior view, and on the right a left anterior oblique view). The source images from the MRA were reconstructed using 3-D volumetric rendering, and nicely demonstrate the typical abnormal orientation of the great vessels seen in CC-TGA.

Figure 5.4 These images demonstrate a severe stenosis of the left pulmonary artery in a 48-year-old male with tetralogy of Fallot status post-repair. On the left is a 3-D volume rendered image in a relatively lateral orientation, while on the right is a thick slab maximum intensity projection in a similar orientation. The red arrows denote the proximal left pulmonary artery stenosis.
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Figure 5.5 3-D volume rendered reconstructions of contrast enhanced MRA in a 47-year-old female with unrepaired severe tetralogy of Fallot (images: left-shallow right anterior oblique orientation; middle-anterior view; right-shallow left anterior oblique). These images are limited to the descending thoracic aorta and demonstrate prominent aortopulmonary collateral arteries with peripheral stenoses. The origins of the aortopulmonary collaterals are denoted by the red arrows.

considered a viable alternative to the invasive catheter angiogram as a preoperative diagnostic tool. In addition to the lack of ionizing radiation, some of these patients needing coil embolization of selected aortopulmonary collateral vessels prior to surgery benefited from the non-invasive roadmap provided by reducing both overall procedure time and X-ray fluoroscopic time, see Chapters 12 and 16.

Coronary artery anomalies
Magnetic resonance angiographic techniques are valuable to evaluate anomalous coronary arteries non-invasively as there is no radiation or administration of potentially nephrotoxic contrast agents. In current practice the navigator-gated approach is most typically used as it allows the patient to breathe freely throughout a data-acquisition period that can range up to ten minutes. While a later chapter is devoted to coronary artery anomalies and disease, it is worthwhile discussing, at least in brief, the use of contrast enhancement in coronary artery magnetic resonance angiography.

To date there are a handful of published series evaluating the merits of coronary magnetic resonance angiography for anomalies relative to conventional angiography, though only a single study involves patients with congenital heart disease [6]. In these studies coronary magnetic resonance angiography had at least equivalent, and sometimes superior accuracy in defining the origin and proximal course of the coronary arteries.

The addition of contrast agent, whether extracellular or intravascular, has both advantages and disadvantages for coronary magnetic resonance angiography. The advantages include greater conspicuity of vessel edges, greater ability to precisely define the diameter of the vessel, and improved ability to precisely determine the length of the vessel, as well as identify its side branches [7]. Alternatively, the addition of contrast agent not only increases signal intensity within the coronary arterial system, but also within the venous system [7]. Contrast enhancement of the venous system complicates the interpretation of images, and may be particularly problematic in coronary artery abnormalities such as arteriovenous fistulas. At present there is only limited information in the use of contrast-enhanced magnetic resonance angiography for coronary anomalies. Viability imaging may be used to complement anatomy (see below).

Contrast enhanced myocardial viability imaging
For the past few decades myocardial viability has been determined by nuclear techniques (e.g.,
SPECT and PET), and more recently by dobutamine stress echocardiography. In the current decade MRI has made a dramatic appearance with the introduction of the “delayed-hyperenhancement” technique, also known as DE-MRI [8]. The use of contrast enhancement in myocardial infarction was studied throughout the 1980s and 1990s, but it was not until 1999 that Kim et al. published the initial manuscript describing what is now known as the DE-MRI technique [9]. This seminal study demonstrated the technique in a canine model, where the left anterior descending artery was occluded either transiently or permanently. DE-MRI of the explanted canine hearts was compared to TTC stained pathology specimens at 1 day, 3 days, and 8 weeks post-procedure, with close correlation demonstrated between regions of hyperenhancement on MRI and irreversible damage at pathology. As initially interpreted, the technique identified the presence and precise extent of chronic, irreversibly damaged myocardium as in chronic myocardial infarction. However, it soon became clear that the technique could also identify acutely irreversibly damaged myocardium as seen in acute and subacute myocardial infarctions.

DE-MRI is now a generally accepted technique for the evaluation of myocardial viability in ischemic disease. The clinical utility of DE-MRI for identifying non-viable myocardium also has been confirmed by direct comparison with multiple clinically established markers of myocardial viability, including contractile reserve, perfusion, metabolism, and even electromechanical mapping [10].

DE-MRI is valuable not only for ischemic related myocardial damage, but has been used successfully to identify infiltrative disorders, characterized by interstitial fibrosis, as in the restrictive cardiomyopathies. Examples include hypertrophic cardiomyopathy, amyloidosis, and sarcoidosis, to name a few [11] (Figures 5.6 and 5.7). As a result, DE-MRI

![Figure 5.6](image) Delayed-enhancement MRI in a 28-year-old male with amyloidosis. The red arrows in the vertical long axis image (on the left) and the mid-ventricular short axis image (on the right) highlight the diffuse, patchy hyperenhancement seen throughout the left ventricular myocardium. Amyloidosis is characterized by an inability to "null" the myocardium as a result of the diffuse infiltrative disorder.
The difference in signal intensity occurs as a result of an increased amount of gadolinium within non-viable tissue [12]. The mechanism for this differential contrast reflects the different volume of distribution, and the different wash-in and wash-out kinetics of gadolinium chelate between non-viable and viable tissue [13,14]. Gadolinium chelate has an initial, brief time in the intravascular system followed by a rapid distribution into the interstitial (extravascular and extracellular) space. Viable and non-viable tissues are substantially different in their degree of interstitial space: non-viable tissue has a large percentage of interstitium (up to 70–80%), while viable tissue has a much smaller amount of interstitium (approximately 15–25%) with the greatest component being intracellular. The increased amount of interstitial space allows a greater amount of gadolinium chelate per unit volume, and therefore greater shortening of T1 and higher signal intensity in non-viable as compared to viable tissue. In addition, non-viable...

**Figure 5.7** Delayed-enhancement MRI in a 12-year-old male with Duchenne muscular dystrophy. The red arrows in the vertical long axis image (on the left) and the mid-ventricular short axis image (on the right) denote the hyperenhancement in the myocardium with regions that are epicardial, transmural, and mid-myocardial. This pattern is not consistent with ischemic myocardial damage, but characteristic of infiltrative, non-ischemic cardiomyopathies.

**Technique**

The pulse sequence is a relatively simple one where, following intravenous administration of a gadolinium chelate, an inversion-recovery prepared, T1-weighted gradient-echo image is acquired. Viable myocardium is “nullled” (turns dark or black on the images) following an appropriately chosen inversion delay time after the inversion pulse. The ensuing image reveals viable tissue as dark, and non-viable, fibrotic or scarred tissue as bright white.
tissue has a relatively delayed wash-in and washout of gadolinium chelate, while viable tissue has more rapid wash in and washout of gadolinium [14,15]. This difference can be taken advantage of by waiting an appropriate amount of time to allow a proportionate washout of gadolinium chelate from viable tissue, while non-viable tissue continues to accumulate gadolinium, and thus appears brighter.

The DE-MRI technique was initially described as a gradient echo, T1-weighted, 2D-sequence with an inversion recovery pulse, and this sequence remains the mainstay of magnetic resonance viability imaging. Since that initial description though, multiple variations of the pulse sequence have been described, each of which has its own particular advantages:

- A steady-state free-precession base pulse sequence can be used instead of gradient echo, along with the inversion recovery pulse. This variation has been used in a variety of applications, and is notable for slight variations in inversion recovery time and accuracy of non-viable tissue determination, as compared to the gradient echo sequence.
- 2D phase-sensitive inversion recovery – this sequence is also 2D, and typically requires a single breath-hold per imaging slice. This sequence requires a longer breath-hold time than the standard 2D gradient echo based sequence, but eliminates the necessity for precise determination of an inversion recovery time for optimal nulling of normal myocardium [16]. As a result data acquisition is simplified for routine examinations.
- Single-shot – this sequence is also a 2D approach, but acquires a single imaging slice per heartbeat. The advantage is that patients who are unable to perform breath-holds or who have significant arrhythmias are still able to have viability imaging acquired [17]. The disadvantage is that the sequence has poorer spatial resolution and contrast resolution compared to standard techniques.
- 3D viability approaches have been developed to allow complete ventricular coverage in a single breath-hold. Such rapid imaging requires incorporation of parallel acquisition techniques (i.e., SENSE/SMASH) to accelerate the image acquisition. The limitations include a slightly poorer spatial resolution compared to the 2D techniques, but the advantages include lack of slice misregistration, which routinely occurs in 2D techniques, and no reduction in signal-to-noise and contrast to noise ratios. 3D approaches may also be combined with navigator gating to allow even higher spatial resolution acquisitions, or used in patients who are unable to perform breath-holds.

The DE-MRI technique, whether using 2D or 3D approaches, has multiple advantages for the determination of myocardial viability: imaging is typically performed under rest conditions; high spatial resolution is achieved; and myocardial viability information is routinely combined with cine-MRI data that is performed in identical slice orientations and positions for precise correlation of corresponding wall motion information. DE-MRI also requires no pharmacologic stress agent or exercise, in contrast to nuclear imaging and dobutamine stress echocardiography, which simplifies the examination. Routine DE-MRI exams can be performed in less than 30 minutes. DE-MRI has high spatial resolution, typically on the order of 1.5–2.0 mm in plane; in contrast, nuclear techniques typically have pixels with spatial resolution ≥10 mm. As a result, DE-MRI has a greater ability to define the transmurality of ischemic damage across the myocardium, and thus better defines the likelihood for functional recovery or improvement across a continuum. Finally, the correlation with cine-MRI is performed with sequences having equivalently high spatial resolution in imaging planes that correspond precisely to the acquired DE-MRI slices.

Interpretation of DE-MRI acquisitions is reasonably straightforward, and is performed in the context of cine-MRI findings:

- Normal, viable tissue is dark on DE-MRI images and has normal wall motion on cine-MRI.
- Non-viable or irreversibly damaged tissue is bright or mostly bright on DE-MRI images and dysfunctional on cine-MRI.
- Dysfunctional myocardium on cine-MRI that is dark or mostly dark on DE-MRI is usually viable.

In general, myocardial segments with ≤25% hyperenhancement have a high likelihood of functional improvement or recovery, while myocardial segments with ≥75% hyperenhancement have little to no likelihood of functional improvement or recovery following revascularization. In these cat-
Assessment of chronic disease

Myocardial viability has been based on nuclear techniques, and frequently has been considered a binary categorization. DE-MRI is altering that perception because of its high spatial and contrast resolution, which allows better delineation of the transmurality of irreversibly damaged myocardium. Subendocardial infarctions, manifested as subendocardial hyperenhancement, are easily differentiated from transmural damage, as well as from intermediate degrees of ischemic damage. The degree or percentage of infarcted tissue per myocardial segment is inversely correlated with recovery of function following revascularization, as shown in multiple studies [18–20].

In an animal model Hillenbrand et al. evaluated the relationship between hyperenhancement at 3 days and contractile recovery at 28 days following myocardial infarction. When hyperenhancement was <25% by segment the majority (87%) of segments demonstrated contractile improvement, as compared to segments with >75% hyperenhancement where functional recovery was unlikely. Intermediate degrees of hyperenhancement resulted in intermediate likelihood of contractile improvement [21].

In a study in humans, Kim et al. studied 41 patients with chronic ischemic heart disease and wall motion abnormalities, where both viability and wall motion imaging were completed pre- and post-revascularization. Kim et al. determined that myocardial segments with little to no bright tissue (irreversibly damaged) on baseline, pre-revascularization viability imaging were highly likely to recover function 2½ months following revascularization. Conversely, myocardial segments with a large percentage of bright tissue were unlikely to recover function [18]. This similar spectrum in potential for contractile recovery is consistent with results from other modalities assessing myocardial viability where hibernating myocardium demonstrates a continuum in its ability to recover.

The high spatial resolution of the technique also provides insights into myocardial disease processes that were previously undetectable. For example, patients undergoing percutaneous angioplasty and stenting frequently manifested small increases in creatine kinase-MB enzyme levels post-procedure that were felt to be of little clinical significance. The high spatial resolution of DE-MRI has provided direct visualization of discrete microinfarctions following percutaneous coronary procedures, thereby explaining the peri-procedural elevations in creatine kinase-MB enzyme levels [22].

DE-MRI has also proven valuable in acute myocardial infarction where regions of microvascular obstruction are detected. Both early and late DE-MRI images can identify the regions of no reflow within the centers of larger acute myocardial infarctions, which are also known as “microvascular obstruction”. Wu et al., using a precursor to the DE-MRI sequence, was the first to find evidence of microvascular obstruction in a series of 44 patients 10 ± 6 days after infarction, and noted that the presence of microvascular obstruction had a significant correlation with increased post-infarction complications [23].

Application to congenital heart disease

To date there have been primarily case reports and case series using DE-MRI in patients with congenital heart disease. The applications have been scattered, and include, but are not limited to, the visualization of a ventricular septal defect patch, identification of subendocardial fibrosis in a child with left ventricular non-compaction, and recognizing subendocardial fibrosis in another child with congenital aortic stenosis (Figures 5.8 and 5.9). The largest study had 40 patients with congenital heart disease who underwent surgery with a control group of 33 patients who had never undergone surgery [24].

At present the most suitable applications seem to be for post-operative evaluations. To that end two manuscripts have been published using DE-MRI in patients with systemic right ventricles. The first, by Hartke et al., noted that nine of 16 patients (56%) demonstrated hyperenhancement of the right ventricular myocardium, ranging from 5% to 80% of the total myocardium [25]. The presence of hyperenhancement was associated with increased rates of ventricular pressures and/or
reduced oxygen saturations, though the relationship was not linear. Alternatively, Fratz et al. studied 27 patients with systemic right ventricles (18 of whom were status post-atrial switch procedure), and found only one patient with hyperenhancement [26]. The discrepancies between these two closely temporally related studies suggest that there is much yet to learn about the presence or absence of myocardial scarring in patients with congenital heart disease, and more importantly what the clinical impact of such findings may be.

**Contrast enhanced myocardial perfusion**

Myocardial perfusion has been a mainstay in the diagnosis, management, and prognosis of patients with ischemic heart disease. Similar to viability imaging, nuclear techniques (i.e., SPECT and PET) have been a central component of ischemia assessment for the past few decades, yet both have limitations. SPECT is limited by relatively poor spatial resolution, has a not insignificant false positive rate based on soft tissue attenuation artifacts, and can impose significant radiation doses. Positron emission tomography has improved spatial resolution compared to SPECT, but remains encumbered by the necessity for cyclotron produced radionuclides, radiation dose, and relatively high cost. Coronary X-ray angiography, considered the gold standard for direct epicardial coronary stenosis determination, is invasive, encumbered by radiation, and is limited by the 2-D nature of the “luminogram” obtained.

Myocardial perfusion imaging with contrast enhanced MRI has advantages of increased spatial
the greatest clinical use since approximately 2000 coinciding with advances in MRI technology, as has been described previously. MRI myocardial perfusion imaging currently has two primary indications: the assessment of myocardial ischemia in patients with suspected coronary artery disease, and the identification of microvascular obstruction in patients with acute myocardial infarction.

**Technique**

The imaging sequence generally employed for myocardial perfusion imaging is a T1-weighted, segmented k-space gradient-echo or steady-state free-precession sequence. The gradient echo sequence may be performed with or without hybridization with an echo-planar readout to improve the speed of image acquisition. A series of short axis slices (3–8 depending on a variety of parameters, and the specific sequence employed) typically is acquired through the left ventricle under pharmacologic vasodilator stress and then repeated at rest. Depending on the sequence resolution (compared to nuclear techniques), lack of radiation, and, similar to nuclear techniques, provides functional information correlating with the degree of epicardial coronary artery stenoses. MRI myocardial perfusion differs from nuclear techniques in that it is a simple impulse response where the relative changes in myocardial signal intensity are evaluated during a bolus administration of gadolinium-chelate contrast agent under both rest and pharmacological stress (typically either adenosine or dipyridamole). The gadolinium-chelate contrast agent causes the myocardial signal intensity to increase in rough proportion to the amount of contrast agent passing through each region of myocardium [27]. Areas of normally perfused myocardium will have a more rapid increase in signal intensity under pharmacologic stress as compared with rest, while areas supplied by stenotic vessels will have slower and shallower rises in signal intensity under stress conditions.

Stress–rest myocardial perfusion imaging has been investigated since 1990, though it has gained
employed images are obtained every heartbeat, or every other heartbeat, during the dynamic passage of a compact bolus of gadolinium-chelate contrast agent.

T1-weighting is improved by the application of preparatory RF pulses applied at the beginning of the sequence. An inversion recovery preparation provides the greatest degree of contrast between normal and abnormally perfused myocardium, yet is sensitive to arrhythmias and therefore less amenable to quantitative assessment. As a result, most sequences employed today apply a saturation recovery preparatory pulse that renders the magnetization insensitive to arrhythmias, and allowing quantitative assessment of results, as desired.

The perfusion images are analyzed by evaluating the relative changes in signal intensity either qualitatively or semi-quantitatively to assess for ischemia. Qualitative assessment is performed as a dynamic visual analysis of the relative signal intensity in regions of myocardium corresponding to coronary artery distributions. A reduced rate of increase in signal intensity or an absolute decrease in signal intensity relative to normal remote myocardium ("hypoperfusion" defects) indicates significant coronary stenosis and ischemic tissue (Figure 5.10). The visual analysis is simple and relatively fast, and has demonstrated comparable sensitivity and specificity to nuclear techniques [28]. Semi-quantitative approaches are more time-consuming, making them less amenable for routine use, but have, in some studies, improved the accuracy over visual analysis. In a recent study, Al-Saadi et al. used the semi-quantitative analysis approach in a group of patients with stable disease, demonstrating a sensitivity and specificity of 90% and 83% for myocardial perfusion imaging compared with invasive coronary angiography [29].

![Figure 5.10](image1.png)

**Figure 5.10** T1-weighted, gradient-echo myocardial perfusion images in a short axis mid-ventricular orientation. The image on the left was obtained during adenosine pharmacologic stress and demonstrates dense hypoperfusion involving the anterior and anterolateral walls and majority of the interventricular septum (arrows) (left coronary distribution). The image on the right was obtained 15 minutes later under resting conditions and demonstrates uniform enhancement of all portions of the myocardium. This 42-year-old male had significant stenoses of the proximal LAD and diagonal branches resulting in pharmacologically induced ischemia, but normal blood flow under resting, baseline conditions.
Theoretically more robust parametric analyses similar to nuclear techniques are being developed that show promise for improved quantitation, but have not been tested in significant numbers.

Clinical validation of myocardial perfusion imaging
Since 2001 there have been at least nine single center studies demonstrating the clinical utility and accuracy of myocardial stress perfusion imaging in patients with suspected ischemia and coronary artery disease. Overall, the sensitivity, specificity, and accuracy have been at least equivalent, if not superior to SPECT [27]. More recently, an international multicenter trial compared stress myocardial perfusion imaging, at multiple contrast doses, to SPECT, with invasive coronary angiography as the gold standard comparator. In this study, performed at 18 centers and using multiple MRI vendors, myocardial stress perfusion imaging was equivalent to SPECT for the subpopulation of patients at optimal gadolinium chelate contrast dose, and superior to SPECT when compared across all doses of gadolinium chelate [30].

Application to congenital heart disease
Similar to myocardial viability imaging only limited data is available for myocardial perfusion imaging with MRI, most of which has been case reports and small case series in patients with congenital heart disease. A literature search reveals only a single publication where myocardial perfusion imaging, in most cases in combination with MRI viability imaging, was used to evaluate patients with congenital heart disease [31].

MRI myocardial perfusion imaging remains in its infancy in congenital heart disease. However, this is an important disadvantage when one considers the current routine use of SPECT imaging in congenital heart disease. Children and young adults are more sensitive to radiation exposure, and have a greater likelihood of developing soft tissue tumors as a result of repeated exposures to ionizing radiation. Substituting MRI myocardial perfusion imaging for at least some of these procedures has obvious potential benefits. In addition, MRI myocardial perfusion imaging has higher spatial resolution than nuclear techniques, providing better delineation of the smaller cardiac structures in children. The limitation in use would seem to reflect not the apparent clinical benefits, but instead the relative lack of available centers with sufficient resources and trained personnel to perform these studies.

References
The basics of cardiac MR

- **Phase-sensitive**
- **Prognostic significance**
- **Contrast-enhanced MRI**
- **Noninfarcted myocardium**
- **Transmural myocardial scars**
- **Evaluation of myocardial viability**
- **Measurement of myocardial infarction**
- **Perfusion cardiovascular magnetic resonance**
- **Delayed-enhancement cardiovascular magnetic resonance**
- **Evaluation of right ventricular function**
- **Noninvasive detection of myocardial ischemia**
- **Noninvasive detection of microvascular obstruction**

**References**

PART II
Cardiac MR of congenital and acquired pediatric heart disease
The normal cardiac magnetic resonance examination – ruling out congenital heart disease

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Introduction

The objective of this chapter is to present a cardiac magnetic resonance (CMR) methodology to verify that a patient does not have congenital or acquired heart disease (i.e., how to ensure that the examination is normal). The focus will be on the particular challenges presented in neonates and young children though the same methodology can be applied to adults as well. CMR can “rule out” congenital heart disease easily and has evolved into the “gold standard” for many applications in congenital heart disease such as vascular rings, assessment of aortic and pulmonary anatomy, ventricular function, blood flow and myocardial viability. In addition, evaluation of coronary artery anomalies has also become a mainstay of CMR in pediatrics with the advent of advanced hardware and software capabilities. The wide field of view CMR offers complements echocardiography. In patients with limited echocardiographic windows, such as larger patients or those who have had thoracic surgery and secondary scarring of the pleural spaces, CMR is especially important to provide a complete intracardiac and extracardiac examination. Additionally, since CMR provides better resolved and quantified data compared to echocardiography with internal checks on the data, we expect that CMR will become, for certain applications, the initial examination of choice in diagnosing congenital heart disease. Table 6.1 lists common reasons for patient referral to CMR who have no known congenital or acquired heart disease.

It is also important to point out the role of computed tomography (CT) angiography in serving as the primary diagnostic tool or as a supplementing modality to echocardiography in the diagnosis of congenital heart disease. CT angiography has excellent spatial resolution for limited intracardiac structures (e.g., coronaries and outflow tracts) as well as the capacity to assess the finest details of extracardiac structures (e.g., pulmonary arteries/veins and aorta). However, CT angiography requires exposing patients to ionizing radiation and iodinated contrast agents; therefore, this should always be used as a last resort in imaging children and females who are more radiosensitive and at higher risk for developing malignancies [1,2].

Prior to a patient entering the magnetic resonance imaging (MRI) bore, electrocardiogram (ECG) leads are placed on the anterior chest wall to enable ECG gating of the moving heart. Precise ECG gating is essential to acquiring data at a specific time during the cardiac cycle, even when performing static imaging. Certain sequences focus on data acquisition during diastole (inversion recovery dark blood, delayed enhancement, coronaries), whereas cine assesses cardiac function throughout
Table 6.1 Most common indications for CMR referral of pediatric patients to rule out (R/O) congenital or acquired heart disease.

Extracardiac structures
- R/O stenosis of tracheobronchial tree
- R/O vascular ring
- R/O systemic or pulmonary venous anomaly
- R/O coarctation of aorta
- R/O branch pulmonary artery stenosis

Intracardiac structures
- R/O coronary anomaly or stenosis
- R/O bicuspid aortic valve
- Define complex congenital heart disease

Assess chamber size and ventricular function
- R/O cardiomyopathy/myocarditis
- R/O arrhythmogenic right ventricular dysplasia
- R/O septal defect and/or left to right shunt (atrial septal defect / patent foramen ovale, patent ductus arteriosus, ventricular septal defect)
- Assess ventricular function in congenital heart disease such as single ventricle, transposition of the great arteries, tetralogy of Fallot, etc
- Assess for viability (myocardial scarring) and myocardial perfusion

Assess blood flow
- Cardiac output
- Fractional blood flow to each lung
- Qp/Qs for ASD, VSD or anomalous pulmonary venous connections
- Assess regurgitant fraction
- Quantify systemic to pulmonary arterial or venovenous collateral flow

the cardiac cycle. Reliable detection of the QRS signal can be particularly challenging in neonates and premature babies who may have heart rates that can exceed 200 beats per minute in sinus rhythm. On the other hand, patients with fast heart rates (>100 beats per min) have less respiratory-induced variation of the R-R interval (sinus arrhythmia) which allows for more consistent data acquisition. After intracardiac congenital heart surgery such as tetralogy of Fallot, some children have widening of their QRS waveform with associated T-wave enlargement (e.g., a right bundle branch block pattern in tetralogy of Fallot after repair). Also, some patients may have QRS alterations which only manifest after being placed into the MRI bore. These circumstances not infrequently result in software failure to reliably recognize the QRS waveform and can make triggering to the QRS problematic. Occasionally, enlarged T-waves will be misidentified as the QRS waveform. Triangulating the ECG leads so that a right triangle is formed between the left sternal border and diaphragm, in addition to bringing the leads closer to each other tends to lower the T-wave relative to the QRS voltage, and allows the CMR detection software to consistently differentiate between the QRS and T-waves. Furthermore, in circumstances where the radio frequency pulse alters the ECG waveforms, some MRI systems allow the users to lower the radiofrequency pulse intensity and prevent waveform distortion. With the advent of vector gating, many of these problems have been solved – most vendors now use this type of approach. The MRI scanner “learns” the vectorcardiogram outside of the bore and then when the patient gets placed at isocenter, the system can more easily detect true vector signal from noise. In addition, most vendors now also allow one to choose whether to trigger off the vectorcardiogram or any one of the limb leads, giving further flexibility to avoid this problem.

While supine, a body array coil is placed over the patient’s chest and the patient is electronically moved into the MRI bore. Small neonates often have better signal provided by a head or knee coils which can be positioned closer to their anterior chest walls. Several localizing images acquired in orthogonal planes (axial, sagittal, and coronal) are utilized for determination of the patient’s body position within the bore. Ideally, the patient’s heart should be positioned within the center of the bore for optimal signal. In order to use CMR to rule out congenital heart disease (CHD), a standard protocol is essential so that all elements are assessed and nothing is overlooked. Table 6.2 provides a complete checklist that includes all essential elements which require assessment to ensure that a patient does not have congenital heart disease.

Thoracic static axial volumetric three-dimensional imaging dataset

The exam protocol begins with acquisition of static steady state free precession (SSFP) bright
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meso/dextrocardia) and the direction of the apex (leftward versus rightward); (2) normal cardiac segments (situs solitus (normal position) of the atria, ventricular D-loop, and solitus great arteries); (3) venous and pulmonary venous connections; (4) atrial and ventricular chamber sizes and septa; (5) atrioventricular connections and ventricular-arterial alignments; (6) pulmonary blood flow distribution; (7) pulmonary to systemic blood flow ratio.

Table 6.2 Factors essential to assess to ruling out congenital heart disease.

- Position of the heart in the chest
- Trachea and bronchi size and relationship to branch pulmonary arteries (hyparterial vs. eparterial)
- Atrial, ventricular, and great artery situs
- Arch-sidesness and branching pattern
- Systemic and pulmonary venous connections
- Atrial and ventricular chamber sizes and septa
- Atrioventricular connections and ventricular-arterial alignments
- Valvular morphology and function
- Ventricular wall thickness, wall motion, and myocardial viability
- Ventricular ejection and cardiac index

Figure 6.1 Turbo spin echo dark blood image of a normal trachea (thick arrow) and bronchi. The right upper and lower lobe bronchi, and left mainstem bronchus are identified (dashed arrows).

This initial three-dimensional image dataset is very useful for clarifying many features which are essential to the assessment of congenital heart disease [3]. These images display the fundamental elements comprising the cardio-thoracic landscape: (1) position of the heart in the chest (levo/meso/dextrocardia) and the direction of the apex (leftward versus rightward); (2) normal cardiac segments (situs solitus (normal position) of the atria, ventricular D-loop, and solitus great arteries); (3) venous and atrioventricular connections; (4) ventriculoarterial alignments; (5) aortic arch anatomy; (6) tracheal anatomy (Figure 6.1) and (7) bronchoarterial relationship (eparterial vs. hyparterial). Main and branch pulmonary artery stenosis and coarctation of the aorta can be inferred on these images but off-axis imaging planes are necessary to confirm and better display these findings. Qualitative assessment of lung hypoplasia and unbalanced pulmonary blood flow can be roughly estimated by the pulmonary vascular markings. In contrast to echocardiographic windows which are frequently limited for assessment of vascular structures which course outside the heart, CMR static axial images can easily identify these structures. For example, the presence of a left superior vena cava (SVC) from its cranial origin connecting (at the junction of the jugular and subclavian veins) to a dilated coronary sinus (at the left atrioventricular groove) or the presence of an anomalous vertical or sub-diaphragmatic vein will be easily identified with
CMR. Moreover, in circumstances where the patient is a young infant and the primary question is to determine aortic arch anatomy and rule out vascular ring, a non-sedated study of diagnostic quality can be acquired using only one average [4]. This is best accomplished by swaddling the infant after a feeding thereby allowing the patient to fall asleep and remain still. Image acquisition time for the entire three-dimensional dataset can be accomplished in 20–30 seconds depending on the patient’s heart rate (Figure 6.2).

These straight axial static images can often be inconclusive for assessing smaller anatomic structures. For example, the right upper pulmonary vein connection to the left atrium may not be definitively delineated on this three-dimensional volume set. This is because the right upper pulmonary vein courses immediately behind the SVC and partial volume effect may result in blurring of their borders, giving the impression of partial anomalous pulmonary venous connection to the SVC. In these cases further imaging techniques (e.g., cine or velocity encoded mapping using off-axis imaging planes) is indicated to confirm normal right upper pulmonary vein to left atrium connection. SSFP axials is not the ideal approach to confirming that the atrial septum is intact. Since septum primum is relatively thin (<2 mm), and these slices are acquired perpendicular to the atrial septum at 3–5 mm in thickness, partial volume effect may result in the impression that there is an atrial septal defect when one does not exist. Conversely, though large atrial or ventricular septal defects are easily identified with axial imaging, small to moderate defects can easily be missed. Special sequences for determining these lesions must be performed. In addition, coronary artery origins and proximal stenosis are not reliably observed on these images. Dedicated coronary imaging sequences coupled with respiratory gating using the Navigator technique is essential to ruling out coronary anomalies (see Chapter 14 on coronary artery disease). These circumstances require more sophisticated imaging techniques to either delineate or rule out these

![Unsedated Study](image_url)

**Figure 6.2** Six-month-old presents to rule out vascular ring. The study was performed without sedation. Images (a–d) were derived through multiplanar reconstruction after being acquired in under 30 seconds as part of a three-dimensional dataset of steady state free precession static axials. Images (a) and (b) demonstrate a left aortic arch with an aberrant right subclavian artery without a retro-esophageal diverticulum (arrow). This is a normal variant. (c) and (d) demonstrate that there is no tracheal narrowing. Images (e) and (f) are high resolution turbo spin echo dark blood images. They confirm that the tracheal anatomy is normal.
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SSFP vs. HASTE
(Dental Braces Artifact)

Figure 6.3 Dental braces flow artifact obscuring the ascending aorta (arrow). The artifact is only present on SSFP (left) and not the HASTE (right) static axials.

pathologies as will be discussed briefly below and in more depth in later chapters.

In addition to SSFP, acquisition of HASTE (Half-Fourier-Acquired Single-Shot Turbo Spin Echo) axial images can be very useful. This dark blood volume set can be acquired while the patient is spontaneously breathing. The entire volume can be acquired using one average in 1–2 minutes. The advantage of this second volume set is that the images are less susceptible to flow induced artifacts. For example, mouth braces are common in adolescents and they can produce artifact which affect the appearance of the great arteries on SSFP imaging, but not on HASTE (Figure 6.3). In addition, endovascular stents in the pulmonary arteries or the aorta will not produce regional artifacts as seen with SSFP. Finally, patients with flow acceleration and turbulence due to valvar or vascular stenosis frequently have significant flow artifacts which can obscure the stenotic region (e.g., single ventricle patients after Stage I Norwood reconstruction and a systemic to pulmonary artery shunt may demonstrate no signal in the pulmonary arteries on SSFP imaging because of turbulent flow in diastole from the shunt – they are clearly seen on HASTE images).

Reviewing HASTE images can be helpful in the evaluation of extracardiac structures such as cystic masses which appear bright on SSFP, and dark on HASTE. In addition, if one of the objectives of the particular CMR study is to determine coronary artery origins, HASTE images can often visualize different parts of the course of coronary arteries. This is especially helpful when 3-point localizers are used to delineate the coronaries. With the advent of whole heart T2 prepared SSFP, this is less commonly performed. It must be noted, however, that for definitive diagnoses of coronary artery origins, in the present era, a 3D SSFP or gradient echo sequence with the Navigator technique is mandatory.

Multiplanar reconstruction

After confirming normal cardiac segmental anatomy with the 3-D volume datasets mentioned above, the next steps are the following: (1) further clarify any remaining questions regarding the intra- and extracardiac structures (valves, septa, arteries, and veins); (2) assess ventricular function; and (3) measure flows. This is accomplished using high-resolution double inversion dark blood, cine, and phase-contrast magnetic resonance (PCMR). These objectives are best addressed using off-axis in-plane and through-plane imaging slice positions. Off-axis imaging planes can be used to profile
the atrioventricular inflows and ventricular outflow tracts in addition to major systemic and pulmonary arteries and veins and their connections to the heart. The basic imaging positions required to confirm normal anatomy and function are listed in Table 6.3. To provide these imaging slice positions, we prefer to load the three-dimensional bright (or HASTE dark) blood dataset into a multi-planar reconstruction software package which enables the user to obtain any desired off-axis plane by manipulating the original dataset. The user simultaneously views the dataset in transverse, sagittal, coronal planes or any double oblique plane to demonstrate the salient points of the anatomy. For each of these planes, there are navigation bars which, when manipulated, can produce any desired off-axis plane. While navigating along one imaging plane, scout lines representing the corresponding location in space, are depicted on the other two planes. First, the user can identify the region of interest by moving through the dataset in straight-axis planes. Subsequently, the user can rotate and direct the navigation bars to arrive at the in-plane and through-plane off-axis slice positions of choice. These imaging planes depicting the salient points of the anatomy can then be saved and used as targets for much higher resolution imaging (e.g., cine, double inversion dark-blood imaging) or as a localizer to interrogate a vessel for flow (e.g., PCMR).

**Dark-blood imaging**

This form of static imaging is used sparingly because it consumes much time; 1–2 images can be obtained in a breath-hold. As the name suggests, blood signal from the cavities or vessels are absent while soft tissue is signal intense. It can be performed in a number of different ways with T1 weighting, T2 weighting, using spin echo, turbo spin echo, double or triple inversion recovery, etc. Most dark-blood imaging in children use either T1 or T2 weighted imaging with the double inversion approach. It can be used to characterize tissue as different signal intensities; for example, fat will be intensely bright on T1 weighted imaging while myocardium will be much less so. In addition, special pulses can be used to alter the signal intensity of a tissue of interest allowing for a non-invasive biopsy of the tissue based on the MRI appearance. For example, as previously mentioned, T1 dark blood imaging demonstrates fat as signal intense; a “fat saturation” pulse prior to a T1 dark blood image will destroy all spins from fat and will show an absence of signal on the image, confirming the tissue is fat. This may be useful in, for example, a mass in the atria which may be a lipoma – visualizing this mass on T1 weighted images with and without a fat saturation can confirm the diagnosis. Turbo inversion recovery magnitude images may be used to delineate edema in the tissue from, for example, a myocardial infarction or myocarditis.

Typically, in our imaging protocols, this is performed after the static contiguous images obtained by SSFP and HASTE. It is generally used when performing a study for arrhythmogenic right ventricular dysplasia (ARVD) to look for fatty infiltration (not very reliable, however, since epicardial and pericardial fat can be mistaken for fatty infiltration of the thin walled right ventricle), to view the pericardium, to image the tracheobronchial tree (in say, a vascular ring) or to characterize a tumor (both before and after gadolinium administration). It can also be used to image the cardiovascular system when stents, coils and other foreign material cause artifacts for SSFP and gradient echo imaging. Precise measurements cannot be performed within a stent, however, because of the “cage effect”. Moreover in-steam stenosis cannot be ruled out.

**Cine**

As cine SSFP can display high resolution images of the myocardium, valves, blood pool, and arteries, it is used for confirming normal cardiovascular anatomy, and assessing myocardial function. In younger children who require sedation, these images are acquired while the patient is freely breathing using multiple averages, thereby mitigating the impact of respiratory artifact. Ideally cine sequences should be “retro-gated” so that wall motion data is available for the entire cardiac cycle, without truncating the beginning or end of the R-R interval as in prospective triggering. The temporal resolution should be set to provide, in general, 20 to 30 frames per heart beat depending upon the
Table 6.3 Essential slice positions.

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PCMR, phase-contrast magnetic resonance.
heart rate. Obviously, in a patient with a heart rate of 150 beats per minute (R-R of 400 msec), 20 frames per heartbeat is more than adequate (20 msec temporal resolution) while if the heart rate is 50 beats per minute (R-R of 1200 msec), 20 frames per heartbeat is not sufficient (60 msec temporal resolution). This is because systole does not vary much as a function of heart rate; it is diastole that lengthens or shortens. A 60 msec temporal resolution for a heart rate of 50 beats per minute will not capture enough frames in systole to adequately assess the ventricle.

SSFP cine images provide excellent temporal and spatial resolution for assessment of wall motion. Regional wall motion abnormalities are readily identified throughout the cardiac cycle (Figure 6.4). If an entire ventricular volume dataset is acquired, then the ventricular volume and mass at end-diastole and end-systole can be measured, yielding the ventricular ejection fraction and mass [5–9]. Measurement of the ventricular end-diastolic volume involves contouring the area along endocardial border at end-diastole of each slice position from base to apex. This summed area

![Figure 6.4](image)

Figure 6.4 Cine imaging – images (a–f) were acquired from a steady state free precession cine for the purpose of assessing left ventricular shortening. The images on the left (a–c) demonstrate the four chamber view, and the images on the right were acquired along a slice plane perpendicular to the long axis of the left ventricle (white lines). These images capture three data points in time: (1) end-systole; (2) mid-diastole; (3) end-diastole. The images demonstrate normal longitudinal compression, radial thickening, and circumferential shortening of the left ventricle. Images (b) and (e) demonstrate normal opening of the mitral valve (dashed arrows).
Rather, we must rely on normal values acquired from adult patients for comparison to our pediatric patient data after it has been indexed for body surface area.

There are several fundamental differences between right (RV) and left ventricles (LV) which are important when considering ventricular morphology and measurement. First, the LV myocardium has relatively increased wall thickness and finely compacted trabeculations compared to the thin, coarsely trabeculated RV myocardium. Second, the anterior leaflet of the mitral valve is in fibrous continuity with the aortic valve cusps without intervening subaortic conus (muscle), whereas there is normally subpulmonary conus separating the tricuspid and pulmonary valves. Third, as the LV sinus (flow) and left ventricular outflow tract (LVOT) lie adjacent to one another, the left ventricular volume and mass can be easily measured along the same short axis plane. In contrast, RV measurements are more challenging since the RV sinus is separated from the RV infundibulum (outflow) by the intervening LVOT. In addition, RV measurements require more care as it is difficult to separate RV myocardial trabeculations from the surrounding blood pool [10] (Figure 6.6).

In addition to assessment of ventricular size, mass, and wall motion, cine imaging is excellent for identifying regional narrowing along a ventricular outflow tract or great artery. Cine can also be used to assess regurgitation of semilunar or...
Bright homogenous signal as blood moves through normal cardiovascular structures. In contrast, regions of flow acceleration may appear brighter compared to more proximal flow or may display discrete signal void at the point of anatomic narrowing, representing turbulent flow. This is a common finding in areas of significant obstruction as can occur with coarctation of the aorta or branch pulmonary artery stenosis. SSFP is less susceptible to turbulence when compared to gradient echo.

Atrioventricular valves, which is minimal in the normal heart. No turbulence should be seen across atrial or ventricular septae by cine, confirming no atrial or ventricular communication. Moreover, gradient echo imaging and to a lesser degree, SSFP, can be used to confirm normal laminar blood flow within the heart and major arteries and veins. SSFP provides a qualitative sense of blood motion direction, turbulence, and acceleration within these structures. The blood pool is assigned a bright homogenous signal as blood moves through normal cardiovascular structures. In contrast, regions of flow acceleration may appear brighter compared to more proximal flow or may display discrete signal void at the point of anatomic narrowing, representing turbulent flow. This is a common finding in areas of significant obstruction as can occur with coarctation of the aorta or branch pulmonary artery stenosis. SSFP is less susceptible to turbulence when compared to gradient echo.
Real time cine CMR (a series of continuous single shot images strung together to capture motion) and interactive real time cine CMR (being able to move the real time imaging plane interactively, similar to echocardiographic “sweeps”) provides a quick way to assess cardiovascular anatomy, function and flow. These images can be used for localization for higher resolution regular cine CMR. It is also used in the event there is too much arrhythmia so that at least a qualitative assessment of the heart anatomy and motion can be achieved.

**Phase contrast magnetic resonance**

PCMR is an essential element for ruling out congenital or acquired heart disease. PCMR is used mostly for quantifying flow across atrioventricular and semilunar valves and to a lesser degree, quantifying effective orifice area and peak velocity. In this way, PCMR can be used to quantify valvar stenosis, valvar regurgitant fraction, cardiac index, and pulmonary to systemic flow ratio ($Qp/Qs$) [11–17]. All patients undergoing CMR to rule out CHD should have flows measured in the aorta and pulmonary artery to confirm a normal $Qp/Qs$ as smaller intracardiac shunts may be missed with cine.

PCMR cardiac output flow data can be compared to cine derived cardiac output volume data [18] enabling each method to serve as an internal check on the validity of the other. For example, in the absence of mitral and aortic insufficiency, the flow measured across the aortic valve derived with PCMR should equal the ventricular stroke volume derived from cine data.

In circumstances where there is anatomic stenosis of a branch pulmonary artery, PCMR applied to both branches measures the fractional pulmonary blood flow ratio which is important data for determining if there is an indication for catheter or surgical intervention. As PCMR does not require ionizing radiation, PCMR measurements of the branch pulmonary artery flows serves as an excellent and safer substitute for radionuclide pulmonary flow studies [19]. Moreover, PCMR enables one to correlate the fractional flow data with the degree of anatomic stenosis observed with cine and magnetic resonance angiography.

Valvular morphology is another important feature to assess in all patients undergoing CMR to rule out congenital heart defects. Cine imaging displays the tissue of heart muscle and its four valves in motion. As a complement to cine imaging, PCMR is useful for identifying valve morphology; since through plane PCMR essentially “tags” flowing blood through a valve, this flowing blood outlines the leaflets edges and essentially makes a cast of the valve orifice morphology while it opens and closes. Of particular note, bicuspid aortic valve is a common clinically relevant congenital heart defect. PCMR can easily identify a bicuspid aortic valve and quantify the degree of valvar stenosis and regurgitation [20,21] (Figures 6.7 and 6.8). Other common valvar defects which can be assessed with PCMR include: Ebstein’s anomaly, dysplastic tricuspid valve, parachute/arcade/double orifice mitral valve, and acquired valvar vegetation (Figures 6.9 and 6.10).

**Gadolinium magnetic resonance angiography (MRA)**

Magnetic resonance angiography readily delineates the major pulmonary and systemic arteries and veins. Intravenous gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA) is administered at a dose of 0.2 mmol/kg. Gadolinium MRA imaging can be performed with or without ECG gating and requires approximately eight to 12 seconds to acquire an entire three-dimensional volume dataset for most applications. Bolus tracking allows for optimal timing of image acquisition (alternatively, a timing run can also be performed) for the target structure. Newer techniques such as TWIST allow for time resolved MRA in <2 seconds per 3D volume set; each volume set is a high resolution acquisition which can be made into a 3D volume rendered cast of the cardiovascular system. In the older static 3D image acquisition runs, the images should be obtained when the Gd-DTPA reaches its highest concentration in the region of interest. Additionally, if right ventricular outflow tract or pulmonary artery obstruction is suspected, image acquisition should occur prior to Gd-DTPA reaching the pulmonary veins. This will ensure that enhancement of the left ventricular outflow tract or the aorta will not obscure the regions of interest.
Multiple measurements should be used so that if the first pass is missed, image acquisition during reperfusion will be captured. The MRA three-dimensional dataset is best acquired in the coronal plane if attempting to visualize the pulmonary circulation despite the invariable need to increase pixel size in order to avoid "wrap" effect. Ideally we try to use isotropic voxels so that the imaging data can be manipulated and resliced along any off-axis plane without considerably sacrificing image resolution. Viewing the MRA raw data in the coronal plane best preserves the AAO due to turbulence from the AoV, however the remainder of the aorta is not susceptible to the significant flow artifact as seen with SSFP. In addition there is signal void (right-sided arrow) and increased signal intensity (left-sided arrow) at the juxtaligamentum region due to turbulent and increased flow velocity respectively. Finally, in-plane PCMR demonstrates increased flow velocity at the AAO and juxtaligamentum regions. The latter region has an artifact due to increased velocity which has exceeded the set velocity encoded scale limits. CMR: cardiac magnetic resonance; AoV: aortic valve; SSFP: steady state free precession; GRE: gradient echo; AAO: ascending aorta; PCMR: phase contrast magnetic resonance.

On the other hand, if aortic arch abnormalities are being ruled out (vascular ring or coarctation), then image acquisition should begin when Gd-DTPA has reached the aorta. Moreover, if aortic imaging is desired, it is ideal to delay image acquisition until Gd-DTPA has cleared the pulmonary arterial system. However, in neonates and infants who have robust blood flow through their cardiovascular system, it may not be possible to identify a point in time where gadolinium enhancement occurs specifically in the right versus left-sided structures. Moreover, any excessive delay may result in having the gadolinium miss enhancement in the region of interest. Multiple measurements should be used so that if the first pass is missed, image acquisition during reperfusion will be captured.

Figure 6.7 Normal aorta versus mild coarctation – the images in the top row displaying the findings of a normal aorta were produced using different CMR sequences. For comparison the lower row of images were produced using the same sequences in a patient with mild coarctation of the aorta. The yellow arrows point to the region of turbulence from the bicuspid AoV and the white arrows point to the juxtaligamentum region. Note the spin echo dark blood nicely defines the aortic arch contours but does not demonstrate flow physiology. The SSFP cine image shows increased signal intensity at the bicuspid AoV but is susceptible to major flow artifacts resulting in loss of signal along the remainder of the thoracic aorta. GRE cine also demonstrates signal void in the region of interest. Multiple measurements should be used so that if the first pass is missed, image acquisition during reperfusion will be captured.
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Maximum Intensity Projection (MIP), Shaded Surface Display (SSD) or Volume Rendered (VRT) image (Figure 6.11).

Coronary artery imaging and myocardial tissue characterization

Not infrequently, pediatric patients may be referred for CMR to rule out congenital or acquired coro-

Figure 6.8 Aortic valve morphology – images (a) and (b) were acquired from a patient with bicuspid aortic valve. In comparison images (c) and (d) were acquired from a patient with a normal tricuspid aortic valve. Note how the amplitude cine-like image demonstrates the valve cusps and the phase-contrast images demonstrate the differences in flow morphology. Not infrequently, the amplitude images may not be able to resolve the leaflet morphology, and in these instances the phase contrast images may be helpful in confirming or ruling out bicuspid aortic valve. Image (b) demonstrates the typical “fish-mouth” appearance of a bicuspid aortic valve. AoV: aortic valve; R, L, N: right, left and non-coronary cusps.

the three-dimensional spatial relationships between the pulmonary and systemic outflow tracts, arteries, and veins. In neonates and infants who have smaller three-dimensional volume acquisitions, it is particularly important to give serious consideration to signal-to-noise ratio even at the expense of image resolution. These data may be loaded into a multi-planar reconstruction software package to view any off-axis slice desired. Additionally, this three-dimensional dataset can be viewed as a Maximum Intensity Projection (MIP), Shaded Surface Display (SSD) or Volume Rendered (VRT) image (Figure 6.11).
Cardiac MR of congenital and acquired pediatric heart disease

The timing of least motion can be determined by reviewing cine diastolic frames for the image which best captures the heart at rest. Diaphragmatic gating allows consistent image acquisition at end-expiration. The images produced generally confirm the coronary origin from its respective sinus and display the proximal portion of the main coronary artery very well [23]. CMR can visualize coronary arteries along the entire course of the vessel (see chapter on coronary imaging).

Tissue characterization should be performed in all patients with suspected coronary artery disease, or evidence of myocardial scarring. For example, congenital anomalous coronary artery from the contralateral sinus of Valsalva may be suspected in patients with clinical symptoms or those who have had an echocardiogram suggesting the diagnosis. Patients with a history of Kawasaki disease or other forms of vasculitis may be referred to rule out acquired coronary artery stenosis or aneurysm. Moreover any patient with suspected coronary or myocardial disease may be referred to rule out myocardial fibrosis.

Coronary artery imaging can be performed in children of all ages, including neonates [22]. Ideally, the imaging data are collected during diastole at a point in time when the heart is motionless and when there is the most blood in the coronaries. The timing of least motion can be determined by reviewing cine diastolic frames for the image which best captures the heart at rest. Diaphragmatic gating allows consistent image acquisition at end-expiration. The images produced generally confirm the coronary origin from its respective sinus and display the proximal portion of the main coronary artery very well [23]. CMR can visualize coronary arteries along the entire course of the vessel (see chapter on coronary imaging).

Tissue characterization should be performed in all patients with suspected coronary artery abnormalities or diminished ventricular function. First-pass gadolinium perfusion can assess the

**Figure 6.9** Double orifice mitral valve – images (a) and (b) were acquired from a through-plane phase contrast magnetic resonance sequence which produces an amplitude cine image (a) and a phase velocity encoded image (b). The slice plane is along the left ventricular short axis and profiles the mitral (M) and tricuspid (T) valve inflows. The white, bright signal identified on the phase contrast image represents flow into and out of the plane of the page. The signal intensity is proportional to the velocity of flowing blood. Images (a) and (b) were acquired from a patient with a normal mitral valve inflow. Images (c) and (d) were acquired from a patient with a double orifice mitral valve. There are two distinct regions of mitral valve (arrows) inflow observed. The orifices are fairly equal in size. Note the signal intensity in the region of the tricuspid valve is less intense compared to the mildly stenotic mitral valve.
Cardiac ischemia is the central purpose for the CMR study. Delayed enhancement (DE) imaging identifies regions of myocardial scarring [24,25]. Approximately 5–10 minutes after the administration of Gd-DTPA, DE images are acquired. Regions of fibrosis have delayed washout kinetics of Gd-DTPA. Since there is a limit on how much Gd-DTPA a patient can receive during a single study, we do not routinely perform myocardial perfusion except in cases where myocardial ischemia is the central purpose for the CMR study.

myocardium for regional perfusion defects secondary to ischemia or fibrosis. These images are acquired at rest and during stress conditions after vasodilation with adenosine. Since there is a limit on how much Gd-DTPA a patient can receive during a single study, we do not routinely perform myocardial perfusion except in cases where myocardial ischemia is the central purpose for the CMR study.

**Pulmonary Arteries**

- **RVOT - Sagittal**
- **Transverse**
- **3-D MRA – Posterior**

Figure 6.11 Images of the right ventricular outflow tract, main, and branch pulmonary arteries. Dark-blood images (a) and (b) nicely demonstrate normal architecture without evidence of stenosis, hypoplasia, or aneurysm. There is normal myocardial signal intensity along the RVOT without evidence of fibrofatty infiltration to suggest arrhythmogenic right ventricular dysplasia. Image (c) is a volume rendered image from a three-dimensional MRA acquisition using gadolinium. This image demonstrates normal lobar branching and arborization of the branch pulmonary arteries. MRA: magnetic resonance angiography; RV, LV: right, left ventricle; RVOT: right ventricular outflow tract; M,R,L main, right, left pulmonary artery.
DTPA. These images are acquired during diastole along the short and long axis of the heart. Obviously, normal individuals should not have myocardial scarring, if an individual is suspected to have myocardial scarring from an insult or manifests arrhythmias that cannot be explained, DE is mandatory to ensure that myocardial scarring is not present and is not a nidus for arrhythmia.

Conclusion

CMR is an important diagnostic imaging modality not only to delineate the details of CHD, but important in “ruling out” or ensuring that there are no cardiovascular defects. There are multiple CMR techniques which are utilized in this assessment and all of them should be available to the imager during the exam. Some add new information and others are internal checks to add a higher level of confidence that no abnormalities exist.

References

CHAPTER 7

Abnormalities of the atria and systemic veins

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The atria and atrial septum

The atria and atrial septum are initially evaluated during routine transthoracic echocardiography (TTE). More challenging situations arise requiring the use of alternative imaging techniques to aid in diagnosis and therapy. Echocardiography may become problematic with poor or limited imaging windows due to patient body habitus, chest deformities, surgical scarring, or other acoustic obstacles. In some patients, right heart dilatation may be found, but the atrial septum not adequately visualized. Other patients may have large defects diagnosed, but with questionable rims to allow for device closure in the catheterization laboratory. Unless intervention is absolutely needed, avoiding an invasive procedure and the ionizing radiation exposure in the catheterization laboratory is preferable. Cardiac CT may not fully define intracardiac anatomy or physiology and also exposes the patient to radiation. Cardiac magnetic resonance (CMR) is an ideal non-invasive, non-radiating modality that can be utilized to further define atrial abnormalities, their physiologic effects, and associated cardiac lesions to assist in guiding clinical management.

The normal atria and atrial septum

The right atrium (RA) is located in the right posterior region of the chest, while the left atrium (LA) is more midline and posterolateral. Each atrium is composed of a free wall and septal components. The RA is distinguished by its large pyramidal appendage (major identifying feature), the limb of the fossa ovalis on the septal wall, and crista terminalis of the free wall. Thin ridges of pectinate muscles are found from the crista terminalis to the anterior aspect of the free wall and within the appendage. The inferior portion of the RA septum is the atrioventricular (AV) septum. This wall between the right atrium and left ventricle has a thin membranous portion and thicker muscular portion. The inferior vena cava (IVC) and superior vena cava (SVC) normally connect to the RA.

Features that identify the LA include its small fingerlike appendage (major identifying feature) and the connection of the pulmonary veins. There are pectinate muscles within the small appendage, but the remainder of the free wall is smooth. The ostium of the fossa ovalis is visible on the LA septum, and the septum does not contain an AV component. Also, the coronary sinus traverses the left atrium in the posterior left AV groove, draining the cardiac venous return to the ostium at the atrial septum near the anterior, inferior portion of the RA [1].

While the minute distinguishing features of the atria are easily directly visualized during surgery or autopsy, the thin detailed nature of the compo-
CHAPTER 7 Abnormalities of the atria and systemic veins

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other simple or more complex congenital heart defects. Using either bright-blood steady-state free precession (SSFP) or dark-blood turbo spin echo techniques to obtain a 3D volume data set covering the entire heart and great vessels, the atrial septum is initially localized. Combining multiple imaging methods, ASD size and margins can be measured accurately.

In the axial plane, the atrial septum is easily localized and most significant secundum defects are visible (Figure 7.1a). However, there can be a normal thinning of the septum in the region of the fossa ovalis, making the actual defect size or presence difficult to determine. With low signal intensity in some spin echo techniques, there may be signal dropout, even when no ASD is present. True defect size can also be difficult to measure with edges not easily visible. Caution should be used and further imaging undertaken to confirm the presence of the secundum ASD.

In the standard four-chamber view, multiple contiguous slices with retrospective-gated bright-blood cine SSFP or FLASH (fast low-angle shot) can reveal the location with the largest ASD dimensions in this plane (Figure 7.1b). The maximal flow across an ASD occurs in late systole or in the early-diastolic rapid filling period. The ASD jet across the septum creates a signal void, defining the width of the defect. When the shunt flow or signal void is minimal or difficult to visualize, presence of shunting can be established or enhanced with the placement of a spatial saturation slab on the LA, or inflow region (Figure 7.1c). Ensuring this slab does not cover other sources of flow into the RA (i.e., superior or inferior vena cava), a bright-blood cine

Atrial septal defects

Secundum ASD

Defects in the atrial septum may be detected from early infancy through late adulthood. The most common type of atrial septal defect (ASD) is a secundum ASD, the result of a deficiency in the septum primum. CMR can be used to determine the presence, dimensions and margins of a secundum ASD, as well as the degree of shunting and effects on right heart volume and cardiac function. CMR also allows detection of associated lesions, such as anomalous pulmonary veins or

Figure 7.1 Secundum ASD (arrowheads) (a) Dark-blood HASTE axial (b) Bright-blood SSFP 4-chamber with light gray flow across the defect. (c) Presaturation band across LA tags flows into RA through ASD, defining width of the defect; 4-Chamber bright-blood FLASH cine image.
is repeated and saturated dark blood will appear in the RA only if shunting from left to right is present. If shunting is right to left, bright blood will appear in the “dark” LA. Similarly, a saturation slab can be applied to the RA if there is a question of right to left shunting [2,3]. In addition, in-plane phase encoded velocity mapping can “label” blood shunting across the defect when the direction of the velocity is encoded properly (see below); as always, care needs to be taken with regard to thickness because of partial volume effects. Views can include off-axis sagittal and 4-chamber views to visualize the flow in these defects.

Since secundum ASDs are not perfectly circular, the maximal diameter may occur in a plane other than the 4-chamber. Thus, bright-blood cine and phase contrast imaging in multiple planes with multiple contiguous slices are used to determine the shape and maximal diameter. Multiple planes are also required to measure ASD rims when deciding if a large defect is likely to be adequately closed with a device in the catheterization laboratory. Further bright-blood cines of the ASD can be obtained in the standard short axis view at the atrial level through the defect, perpendicular to the 4-chamber view, and maximal diameter determined in this plane. With the normally thin atrial septum, an “en face” view for sizing can be difficult to interpret with standard bright-blood or dark-blood techniques. The thinning of the atrial septum at the defect edges could be mistaken for part of the defect. Phase contrast imaging has been employed for the purpose of ASD sizing as described by Beerbaum et al. [4]. The plane of the ASD is localized in the axial and sagittal images, and the resulting off-axis coronal image shows the defect. The velocity encoding can be set at 50–70 cm/sec. These images show the overall shape and size of the ASD (Figure 7.2). Measurements of the ASD and the surrounding rims to adjacent structures can be performed in both the magnitude and phase images. Using the coronal image and prior axial images, planning scans through the ASD to the aorta, SVC, and IVC allows for more rim measurements. Using this method in 65 children, Beerbaum showed that CMR can determine ASD size and rims and can detect associated venous anomalies when TTE results are not conclusive [4]. This allows the patient to be appropriately sent to the catheterization laboratory or the operating suite for ASD closure. Multiple studies and reports have compared CMR methods for ASD sizing to TTE, TEE, catheterization, and surgical findings [2,4–9]. While the simple dark-blood spin echo technique alone is not adequate, the combination of bright-blood and phase contrast imaging methods described above provide accurate measurements of ASD size and rims. Durongpisitkul et al. determined that an adequate posterior rim is the most predictive factor for successful transcatheter closure of a secundum ASD and showed that CMR, rather than TTE or TEE can best visualize this [6].

Phase contrast, velocity-encoded imaging is also applied to determine the degree of cardiac shunting in patients with an ASD. Pulmonary and aortic flow results can be obtained with usual through-plane measurements of the main pulmonary artery and proximal ascending aorta (Figure 7.3). Velocity encoding is normally set at 150 cm/sec. The ratio of pulmonary to systemic flow (Qp/Qs) is then calculated. This information may be useful for deciding whether a defect should be closed, based on the degree of shunting. Obtaining the shunt fraction in this manner eliminates the need for oxygen measurements, which are performed invasively in the catheterization laboratory. Utilizing CMR for flow measurements has been proven reliable and accurate when applied to atrial or ventricular level shunts [10–12]. Through-plane measurement within the plane of the ASD, however, is not necessarily an accurate method to quantify Qp/Qs, as it may overestimate the shunt compared with that obtained by pulmonary and aortic flows [11].

Secundum ASDs can cause significant right ventricular volume overload. With CMR, the full extent of the RA and RV dilatation can be evaluated. Diastolic flattening of the ventricular septum with bright-blood cine imaging in the four-chamber and short axis planes will be apparent (Figure 7.4). The degree of right chamber dilatation can be quantified with standard ventricular volume analysis in the short axis orientation. The RV end-diastolic volume will be higher than normal. Comparison of the right and left ventricular stroke volumes will also be discrepant due to the shunt volume.

With right heart dilatation, the pulmonary and tricuspid valves may also dilate or demonstrate
"cryptogenic" strokes are often referred for echo imaging in search of a PFO. Using contrast echocardiography, the patient may be asked to perform a Valsalva maneuver to ascertain the presence of a PFO with right to left shunting. When transthoracic echocardiography is not adequate, TEE may be performed. In adults, TEE has been used as the gold standard for diagnosis of a PFO. TEE is semi-invasive and allows for imaging in only a single plane when contrast is administered. While children cannot easily cooperate with the Valsalva

regurgitation. The pulmonary artery and its proximal branches may also enlarge. These findings can all be displayed with routine CMR imaging.

**Patent foramen ovale (PFO)**

A PFO exists when the flap of the foramen ovale fails to fully close. A PFO with a small amount of left to right shunting is usually of no clinical significance. However, if the PFO allows for right to left shunting, this may put a patient at risk for a stroke or ischemic event. Thus, patients with

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**Figure 7.2 Secundum ASD.** (a) Axial and (b) Sagittal planes are used to obtain (c), (d) off-axis coronal images in plane of ASD (arrowhead) for phase contrast image (c) Magnitude image, difficult to visualize defect. (d) Phase image with VENC of 60 cm/sec can be used to measure ASD size (arrowhead).
In a similar study of 75 patients, CMR had a significant false negative rate [14]. MRI is advantageous due to its non-invasive nature and ability to visualize associated anomalies, but it may not fully exclude the possibility of a PFO. Regardless, diagnosing a PFO in a child may not frequently require the use of anything beyond transthoracic echocardiography.

Figure 7.3 Flow curves from phase contrast images for aorta and pulmonary artery in patient with secundum ASD. Ao: aorta; PA: pulmonary artery.

In cooperative adults, recent studies comparing CMR and TEE have had variable results. Dynamic contrast-enhanced MRI has been used, showing excellent feasibility in a series of 15 patients, for diagnosing both PFO and atrial septal aneurysm, visually or with signal time curves in the LA [13].
Abnormalities of the atria and systemic veins

Blood flow from both the RUPV and LA to drain easily into the RA. Using a 3D volume data set, planes are manipulated to identify pulmonary venous drainage not initially visualized and bright-blood cines are performed (Figure 7.5). 3D gadolinium data sets will also identify anomalous pulmonary venous drainage. In addition, as with the secundum ASD, the hemodynamic significance of the shunting can be determined using the flow and volume analysis techniques described above (Figure 7.3).

In a recent study of children comparing CMR with surgical findings, CMR accurately demonstrated sinus venosus defects and anomalous pulmonary venous drainage. CMR identified additional important anomalies and was superior to echocardiography (TTE and TEE) in diagnosing these defects [15].

Sinus venosus ASD
Sinus venosus ASDs account for less than 10% of all ASDs. The most common type of this defect occurs when there is deficient development of the wall between the right SVC and right upper pulmonary vein (RUPV), which results in a significant amount of left to right shunting and right heart dilatation. The defect is located in the posterior atrial septum, behind the fossa ovalis. More rare sinus venosus defects are located inferiorly towards the IVC with anomalous drainage of the right lower pulmonary veins. The right middle pulmonary vein may also drain anomalously. CMR is utilized when a sinus venosus ASD is suspected or when the pulmonary venous drainage pattern needs to be determined in preparation for surgical repair. Similar to TEE, CMR may require sedation or anesthesia in infants and young children, but allows for a non-invasive exam, better field of view and multiplanar viewing capability. One of the key advantages of CMR in this lesion is to identify all pulmonary veins pre-operatively; it is especially important that the surgeon knows what veins he might have to baffle and whether or not a Warden procedure is needed.

With standard axial bright-blood or dark-blood images covering the heart and great vessels, a sinus venosus ASD is immediately identified. Anomalous pulmonary venous drainage may often also be easily seen. As the SVC approaches the area of the RA and RUPV, the posterior SVC wall is absent and there is not a complete wall of atrial septal tissue dividing the RA and LA. The wall separating the SVC and RUPV is also deficient and this situation allows blood flow from both the RUPV and LA to drain easily into the RA. Using a 3D volume data set, planes are manipulated to identify pulmonary venous drainage not initially visualized and bright-blood cines are performed (Figure 7.5). 3D gadolinium data sets will also identify anomalous pulmonary venous drainage. In addition, as with the secundum ASD, the hemodynamic significance of the shunting can be determined using the flow and volume analysis techniques described above (Figure 7.3).

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Atrial septal aneurysm
An atrial septal aneurysm is a bowing of the tissue of the fossa ovalis, which may or may not be associated with an ASD. It is usually formed by redundant atrial tissue of the septum primum, which may move back and forth during the cardiac cycle or appear as a stable bulge into the lower pressure atrium. It can be mistaken for an atrial tumor and is thought to be the origin of emboli in some patients. With CMR, an atrial septal aneurysm can be seen, eliminating the diagnosis of a tumor and determining the presence or absence of associated shunting [13,14,16] (Figure 7.6).

The closed secundum ASD
Most ASD devices used to close defects in the catheterization laboratory allow the patient to undergo
occluder devices with impact on surrounding structures and protrusion into cardiac chambers. Using turbo spin echo for morphology and dynamic imaging with 2D cine FLASH, thorough evaluation of all but the smaller coronary sinus entrance to the RA was obtained [17].

MRI for other clinical cardiac or non-cardiac indications. For example, an Amplatzer septal occluder creates only local artifact and the remainder of the cardiac structure and function is easily evaluated (Figure 7.7). A recent pediatric study of 26 patients used CMR to evaluate positioning of large ASD

Figure 7.5 Sinus venosus ASD at the junction of the SVC/RUPV at the RA. Blood flows from the RUPV into the RA, along with flow from the LA. (a) Axial and (b) off-axis sagittal bright-blood SSFP cine images. ASD: atrial septal defect; LA: left atrium; RA: right atrium; RUPV: right upper pulmonary vein; SVC: superior vena cava.

Figure 7.6 Atrial septal aneurysms (arrowheads) (a) Patient with secundum ASD; FLASH 4-chamber cine image and (b) Patient with corrected transposition of the great arteries, steady state free precession (SSFP) 4-chamber cine image.
Abnormalities of the atria and systemic veins

Common atrium
Often associated with heterotaxy syndromes, the common atrium can be demonstrated by CMR. There is a lack of atrial septal tissue. These patients are often evaluated by CMR to clarify other associated complex cardiac anomalies as well as measuring Qp/Qs.

Other atrial findings
Cor triatriatum sinister may be seen with CMR. Resulting from an abnormality in pulmonary venous development, it is a membrane within the LA which septates it into a proximal chamber receiving the pulmonary venous return and a distal chamber with the left atrial appendage and the mitral valve. The membrane may be fairly complete and obstructive, have one or more perforations, or be quite incomplete without any significant obstruction. There may also be an associated secundum ASD in the proximal or distal chamber. When left heart obstruction is suspected, but the etiology not clearly defined, CMR is recommended for diagnosis, avoiding the need for cardiac catheterization [18]. CMR can identify the membrane and associated anomalies (Figure 7.9). This especially includes turbulence at the membrane level.

Primum ASD
The primum ASD is located inferiorly on the atrial septum, near the AV valves. It is part of the spectrum of atrioventricular septal defects, which exhibit abnormal structure and function of the atrioventricular (AV) valves, as well as inlet region ventricular septal defects (see chapter on the ventricle and pericardium) (Figure 7.8). A primum ASD is easily seen by CMR, which may be ordered in these patients to evaluate other problems, particularly whether unbalanced ventricles are adequate size for complete surgical repair. It may be difficult to separate atrial level from ventricular level shunting and there may also be flow through the inferior AV septum, from the LV to the RA. Still, pulmonary to systemic flow ratio can be determined with the phase contrast velocity encoding technique across the pulmonary artery and aorta. Associated defects can be delineated.

![Figure 7.7 Bright-blood SSFP images in patient after Amplatzer septal occluder (arrows) placement. (a) 4-chamber cine; (b) coronal; and (c) sagittal views.](image)

![Figure 7.8 Patient with atrioventricular septal defect. Large primum ASD (arrowhead) and large inlet VSD (arrow).](image)

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Cor triatriatum dexter may be seen as a membrane in the RA when the right valve of the sinus venosus fails to fully regress into the eustachian and thebesian valves. The remaining tissue is called a Chiari network. Usually, the septation is only minor, but if more substantial, it can cause obstruction of the IVC or tricuspid valve. If extensive, it can prolapse into the RV and cause right ventricular outflow obstruction. CMR is a
oblique sagittal planes can be selected for further imaging of the coronary sinus and left atrium, using both dark-blood techniques and bright-blood cine techniques [20]. Qp/Qs is assessed using phase contrast CMR.

CMR intervention
ASD closure has been successfully performed in animals using real-time CMR-guided interventional procedures and equipment. Before it is practical to test in humans, there are still hurdles to overcome related to imaging artifacts from devices used and the thin quality of the atrial septum [21]. As CMR techniques improve, with better spatial resolution and techniques to minimize artifact, CMR-guided interventions may become more appropriate. Radiation exposure to the patient and both operators (TEE cardiologist and the interventionalist) could be eliminated. It has the potential to provide the interventionalist with an accurate 3D representation of the atrial septum, not fully possible with catheterization or standard TEE. The small risk of esophageal rupture associated with TEE could also be avoided. Likewise, additional sensitive and non-invasive technique that can diagnose this rare cause of right heart obstruction [19]. More often, however, this remnant of tissue may be an insignificant and incidental finding in a patient referred for cardiac imaging for other indications.

The coronary sinus is clearly identified in its normal course through the LA, opening into the RA. A dilated coronary sinus may be associated with a left SVC (see below). A defect in its path through the LA is called an unroofed coronary sinus, and will result in shunting across the atrial septum. It may contribute to systemic oxygen desaturation and may put a patient at risk for brain abscess or emboli. An unroofed coronary sinus is a rare occurrence and is often associated with a persistent left SVC connecting to the LA, or other more complex congenital heart defects. It can be identified with CMR, but is usually an incidental finding. Unless there is a high suspicion for an unroofed coronary sinus, it may not be discovered during routine echocardiography or cardiac catheterization. Using CMR, once the location of the coronary sinus is identified in the axial plane, oblique sagittal planes can be selected for further imaging of the coronary sinus and left atrium, using both dark-blood techniques and bright-blood cine techniques [20]. Qp/Qs is assessed using phase contrast CMR.

**Figure 7.9** Cor triatriatum sinister and abnormal pulmonary veins. (a, b, c) are bright-blood SSFP cine images. (a) Axial plane demonstrates the cor triatriatum membrane (arrow) within the LA. (b) 4-Chamber view shows secundum ASD in the distal LA chamber. (c) Off-axis coronal plane demonstrates path of left pulmonary veins across midline, draining into the LA. Arrow points to the cor triatriatum membrane, with a perforation opening into the distal LA chamber below. (d) 3D coronal maximum intensity projection (MIP) in anterior view with arrow pointing in the direction of flow of pulmonary venous return. (e) 3D shaded surface display in posterior view; all pulmonary veins drain to the LA on the right.
The systemic veins

The SVC and IVC normally connect to the RA from above and below, bringing all of the systemic venous return back to the heart to eventually travel to the pulmonary arteries for oxygenation. With standard CMR imaging protocols, this anatomy is easily visualized [22]. There are normal variants of the systemic veins that ultimately may not change the basic physiology, but which may have an impact on surgical or catheter-based interventions, making knowledge of their presence critical. Examples include a persistent left SVC draining to a dilated coronary sinus, azygos continuation of an interrupted IVC, or a retroaortic innominate vein. Other variations will change the physiology and are thus important to delineate. These variations include anomalous connection of a persistent left SVC to the LA, anomalous pulmonary vein connections to the SVC, innominate vein, or IVC, and other more complex cases often associated with heterotaxy syndromes.

Although CMR may not be needed to diagnose every case of a congenital systemic venous anomaly, it is extensively used in evaluating venous anatomy and plays a definitive role. Due to variable imaging windows, transthoracic echocardiography may not always define systemic venous anomalies. Transesophageal echocardiography can be even more limiting for the evaluation of extracardiac structures. Cardiac catheterization may be able to characterize these lesions well, but it is invasive and exposes the patient to ionizing radiation. Cardiac CT also uses ionizing radiation. CMR, which is non-invasive and non-radiating, with its wide field of view and multiplanar capability, is the ideal modality to evaluate systemic venous abnormalities. It is able to clearly establish the origin, course and termination of an anomalous vessel, where echo may have more difficulty or demonstrate a less clear picture. Standard dark-blood and bright-blood imaging for morphology will reveal these anomalies in patients referred for CMR for any reason. Once identified, further imaging in any chosen plane can clearly demonstrate the abnormality in question with single-phase images, multi-phase cine, imaging with gadolinium administration, and creation of 3D reconstructions. As well, CMR velocity encoded phase contrast imaging can be used to determine magnitude and direction of flow in anomalous vessels. Time resolved gadolinium sequences can track the flow of blood and can reveal shunting at various levels (e.g., with a left SVC connected to the LA without a bridging vein, injection of gadolinium in the left arm will “light up” the LA first, not the RA).

In 2002, Greil et al. reported the ability of gadolinium-enhanced 3D magnetic resonance angiography (MRA) to evaluate systemic venous anomalies in 61 patients [23]. While standard CMR is accurate, it may take several minutes and various imaging planes for thorough coverage of the desired anatomy. Using the 3D MRA sequence, high-quality diagnostic images were obtained in an average of 29 seconds. MRA diagnoses were compared with other cardiac imaging modalities and surgical or autopsy specimens, and all venous anomalies were accurately diagnosed. When compared with other cardiac imaging studies available, MRA found previously unsuspected anomalies in 28% of patients and provided additional clinically important information in another 46% [23]. With more recent, high performing gradients, multiple sequential acquisitions can be obtained, each <2 seconds during gadolinium contrast administration (time resolved gadolinium mentioned above). The set of images acquired at the most appropriate time interval can be used for reconstruction and diagnosis.

Left SVC

The most common anomaly of the systemic veins is a persistent left SVC draining into the RA via the coronary sinus. In the embryo, the anterior cardinal vein on the left fails to regress. A persistent left SVC is often initially suspected when images of a dilated coronary sinus are obtained by echo. Its course from extracardiac to intracardiac is easily traced in the axial set of CMR images. Bright-blood cine of its longitudinal plane can be obtained. There is most often a right SVC and an absent or very small bridging innominate vein, with other variations easily diagnosed by CMR (Figure 7.10).
Figure 7.10 Persistent left SVC. (a) 3D MR angiography of bilateral SVCs with larger left SVC. (b) Axial dark-blood HASTE. (c) and (d) Bright-blood cine images demonstrating the dilated coronary sinus (CS). (e) Volume rendered image of the posterior surface of the heart with dilated CS. LSVC: left superior vena cava; RSVC: right superior vena cava.
CHAPTER 7 Abnormalities of the atria and systemic veins

Miscellaneous systemic venous abnormalities

Retroaortic innominate vein

The finding of a retroaortic innominate (left brachiocephalic) vein is usually incidental by cardiac catheterization, CT or CMR. Its course can be followed in coronal or axial dark or bright-blood images through the superior half of the chest. A more specific plane through the path of the vessel can also be obtained. Rather than the usual oblique passage downward and anterior to the aortic arch and its branches, the anomalous vein traverses beneath the aortic arch, posterior to the ascending aorta. It can occur in up to 1.7% of patients with congenital heart defects, with the most common association being tetralogy of Fallot [25] (Figure 7.12). Most importantly, it should be differentiated from a persistent left SVC, partial anomalous left pulmonary venous connection, or a vertical vein in the case of anomalous pulmonary venous return. This can be done by using CMR images to carefully trace the entire path of the vessel towards its insertion into the SVC.

Azygos continuation of the IVC

The most common anomaly of the IVC is absence of the hepatic segment, which occurs when the suprarenal part of the subcardinal vein fails to develop early in the embryo. Inferior venous return then drains through the azygos and hemiazygos venous system, and the hepatic veins drain directly into the RA via the suprahepatic segment of the IVC.

Again, this is easily visualized when there is no vessel coursing from below superiorly through the liver to the right atrium. In the axial plane, the IVC and aorta are seen as “double barrels” anterior to the spine, with the azygos to the right of the aorta. MRI nicely demonstrates a dilated azygos vein draining to the SVC (Figure 7.11). Circumstances requiring awareness of this variation include surgical staging for the single ventricle patient and planning access for cardiac catheterization/interventions.

Knowledge of the left SVC is important for surgical cannulation for bypass and for surgeries such as the bi-directional Glenn or hemi-Fontan procedure. Otherwise, it may be clinically insignificant.

A persistent LSVC may drain directly to the LA or may be associated with an unroofed coronary sinus, which drains into the LA. It can be an explanation for lower oxygen saturation in an otherwise stable patient. It is frequently associated with additional congenital heart defects. Its presence and entire length can be confirmed with CMR [24], especially with time resolved imaging using contrast and injection in a left arm vein demonstrating the LA “lighting up.” Phase contrast imaging of a cross-section through the left SVC can be used to verify flow amount and direction towards the heart [22].

Heterotaxy syndromes

Numerous systemic venous anomalies are associated with heterotaxy syndromes. The IVC and/or the SVC may connect to either the right or left-sided atrium. There may be azygos or hemiazygos continuation of the IVC to a right or left-sided SVC. The veins may enter the atrium in abnormal positions. There are a multitude of possibilities, with and without definite embryologic explanations. CMR can delineate details of these various complex connections, and associated intracardiac congenital heart disease, in preparation for further therapy.
Anomalous pulmonary venous connections to systemic veins
CMR of anomalous pulmonary veins are addressed in detail elsewhere in this text. It is important to know that anomalous pulmonary veins often connect to the systemic veins, causing minor or major changes in vessel size, in addition to significant hemodynamic abnormalities. Examples include partial anomalous pulmonary venous return to the SVC or innominate veins, scimitar syndrome, total anomalous pulmonary venous return to a systemic vein, and numerous other alterations, for which CMR can provide a definitive anatomic and physiologic diagnosis [26,27].

Summary
CMR is the ideal non-invasive, non-radiating modality to clarify defects involving the atria and systemic veins, especially in the event of a difficult or non-diagnostic echocardiogram. Diagnoses missed by other modalities are easily discovered, and suspected anomalies are confirmed. CMR provides valuable physiologic data in addition to anatomic detail and can provide accurate assessment of Qp/Qs, for example, that other non-invasive imaging modalities can’t. Information acquired through use of CMR techniques allows appropriate guidance of clinical decisions and therapies. As techniques continue to advance with higher spatial and temporal resolution and faster imaging capabilities, utilization for delineating atrial and systemic venous abnormalities will continue to increase. This may also allow for safe and reliable CMR-guided interventional procedures, decreasing risks for the patient, physician and staff.

References
Abnormalities of the atria and systemic veins


There are many times when echocardiography is insufficient as a non-invasive technique to evaluate the ventricles and pericardium in neonates, children and adolescents. Echocardiographic imaging may be limited by suboptimal acoustic windows or by interference from adjacent air-filled structures. Perhaps more importantly, cardiac magnetic resonance (CMR) has unique advantages over echocardiography in accurately and reproducibly quantifying global and segmental ventricular anatomy, morphology and function [1,2]. CMR can be used to understand the complex 3-dimensional geometry and function of the right and left ventricles, their inter-relation through the ventricular septum, and their orientation within the thorax. CMR enables assessment of cardiac chamber dilation or hypoplasia, either due to congenital abnormalities of cardiac development, as a result of shunting or valvar lesions, or following surgical intervention. Regional and global ventricular wall thickness, either as a primary abnormality or as a consequence of obstruction to ventricular outflow, can be quantified by CMR. CMR also facilitates serial assessment of changes in ventricular geometry and/or function following surgical or medical interventions. Regional wall motion and strain under conditions of both rest and stress can be determined by CMR techniques. CMR also can be used to investigate myocardial characteristics in ways that are not currently feasible by 2-D echocardiography; for example, one can perform rest or stress perfusion and/or viability imaging following the injection of a gadolinium chelate contrast agent to assess for evidence of myocardial ischemia/infarct, fibrosis, inflammation, or abnormal myocyte infiltration.

**CMR techniques for assessment of ventricular and pericardial morphology and function**

There are several CMR techniques that are used to assess ventricular and pericardial morphology and function in neonates and children, including static steady state free precession (SSFP), ECG-gated cine, viability and real-time cine imaging in any anatomic orientation [3].

CMR examination performed to investigate ventricular morphology and function would typically include a set of axial static images from the level of the aortic arch to the diaphragm, either performed with breath-holding or respiratory gating via a navigator technique. This can be done with static bright-blood imaging or by using a T1-weighted spin echo (“black-blood”) technique. T1-weighted spin-echo imaging, particularly an ECG-gated, breath-hold, double inversion recovery pulse sequence, is an excellent technique with which to begin CMR delineation of ventricular and pericardial anatomy as it takes advantage of the high contrast between low-signal intensity “black blood” (rapid blood flow having been nulled by the inversion pulse), intermediate-intensity myocar-
Abnormalities of the ventricles and pericardium

Many congenital and acquired cardiac anomalies lead to abnormal loading conditions on the right ventricle (RV); there could be abnormal volume load (higher or lower than normal), pressure load, or a combination of both. The right ventricle may also be intrinsically smaller or larger than normal. In addition, correction or palliation of congenital heart defects can result in changes to right ventricular loading. One of CMR’s greatest strengths in pediatric cardiology is its ability to quantify and serially assess the magnitude and etiology of abnormal right ventricular loading non-invasively and without the need for ionizing radiation.

**Right ventricular volume overload**

Children and adults are often referred for CMR evaluation when their right ventricle appears dilated by conventional 2-D transthoracic echocardiography. CMR enables one not only to quantify RV size, but also to evaluate potential causes for RV dilation, such as atrial septal defects, partial or total anomalous pulmonary venous drainage, tricuspid or pulmonary regurgitation, pulmonary hypertension, arrhythmogenic right ventricular dysplasia/cardiomyopathy or from post-operative sequelae such as tetralogy of Fallot repair with a transannular patch and pulmonary insufficiency. During CMR examination in subjects with RV volume overload, one would also assess for evidence of right ventricular hypertension by examining RV mass and ventricular septal position; other CMR techniques, such as dynamic perfusion MRI [4], or pulmonary artery PC flow mapping [5], can also be included to obtain indirect assessment of pulmonary pressure and resistance.

An indication for CMR might be in a patient diagnosed by transthoracic echocardiography to have RV dilation with an atrial septal defect, but where one cannot exclude anomalous pulmonary venous drainage or in a patient where isolated anomalous pulmonary venous return is suspected in the absence of an ASD. In both of these cases,
CMR will help determine the need for and most suitable type of intervention (e.g., surgical vs. transcatheater ASD closure) [6]. During CMR examination of a patient with right heart dilation, one would also assess for the presence of right ventricular hypertrophy, ventricular septal orientation during systole and diastole, and at least qualitatively assess for the presence of right ventricular segmental wall motion abnormalities. The degree of left to right “atrial-level” shunting, most often a consequence of an ASD and/or partial anomalous pulmonary venous return, can be quantified by calculating the ratio of pulmonary to aortic blood flow (via PC flow imaging) and by calculating right to left ventricular stroke volumes from cine SSFP short-axis imaging. Note that this latter method of pulmonary to systemic flow ratio (Qp:Qs) calculation is only be valid in the absence of other sources of right or left ventricular volume overload, such as atrioventricular valve or semilunar valve regurgitation, or cardiac shunting at a different level. When RV volume overload is due to shunting at more than one level, or a combination of shunting plus valvar regurgitation, right vs. left ventricular stroke volume measurement by cine SSFP imaging and through-plane PC flow measurement across the atrioventricular valves, semilunar valves, and of any identified shunt can be compared to measure the proportion of right ventricular volume overload due to atrial shunting vs. that due to other levels of shunting (such as anomalous pulmonary venous return) or valvar regurgitation [7].

Figure 8.1a through 8.1c are 2-chamber, 4-chamber, and short axis cine SSFP images obtained during CMR performed to assess pulmonary venous return after transthoracic echocardiography demonstrated a large sinus venosus atrial septal defect in a teenager who presented with palpitations. Note the significantly dilated, not hypertrophied, right ventricle. The large sinus venosus atrial septal defect is seen with partial anomalous pulmonary venous return of the right upper and middle pulmonary veins in the region of the defect; there is an additional small secundum ASD, not shown. Qp:Qs calculation by RV vs. left ventricular (LV) volumetric analysis and by MPA vs. aorta PC flow imaging revealed a left to right shunt of 2.5:1. As an incidental finding, the patient was also noted to have partial absence of the pericardium with leftward displacement of the heart.

**RV volume overload due to valvar regurgitation**

A CMR examination protocol similar to that described above can be performed when the etiology of right ventricular dilation is valvar regurgitation, either due to an abnormality of the tricuspid valve such as Ebstein malformation, or of the pulmonary valve, most typically following transcatheter or surgical pulmonary valvuloplasty.

Ebstein malformation is a congenital cardiac abnormality characterized by failure of delamination of the tricuspid valve from the right ventricular endocardium, leading to dysplasia and inferior
displacement of the tricuspid valve. CMR has been used to diagnose the lesion, to further characterize the degree of inferior tricuspid valve displacement, the magnitude of atrialized vs. "true" right ventricular dilation and/or dysfunction, to assess for the presence and hemodynamic significance of associated lesions, such as atrial-level shunting or right ventricular outflow tract obstruction and to characterize 3-dimensional function. Ebstein's anomaly has also been associated with segmental LV dysfunction and non-compaction, which can be easily assessed by CMR but may go unrecognized on standard transthoracic echocardiographic imaging. CMR is especially useful in evaluating right ventricular volumes, function, and the degree of tricuspid regurgitation following surgical repair of Ebstein's anomaly.

CMR assessment of ventricular anomalies associated with Ebstein malformation (Figure 8.2) is best accomplished by a combination of T1-weighted spin echo imaging and cine SSFP imaging to assess the extent of abnormal tricuspid leaflet motion caused by leaflet attachments to the RV free wall and/or ventricular septum (tricuspid valve morphology), abnormal ventricular septal orientation, as well as to calculate RV and LV volumes and function [8]. Figure 8.2a,b demonstrates typical inferior-apical displacement of the hinge point of the tricuspid valve and effective tricuspid orifice, dilation of the "atrialized" right ventricle, abnormal tricuspid attachments to the ventricular septum and RV free wall and abnormal ventricular septal position. There are often associated intracardiac lesions, such as pulmonary stenosis or atresia; Figure 8.2c,d illustrates CMR performed on an infant with combined Ebstein's anomaly and pulmonary atresia who previously underwent a Blalock-Taussig shunt. There may be abnormalities of left ventricular morphology and function that are related in part to paradoxical motion of the ventricular septum, although left ventricular non-compaction has also been reported. PC flow imaging of the ascending aorta and MPA is obtained to measure right and left ventricular output and to assess for intracardiac shunting; accurate PC flow assessment of tricuspid inflow and regurgitation may be technically challenging due to the asymmetric shape of the effective tricuspid orifice in Ebstein's anomaly, and the magnitude of TR may be more reliably assessed by subtracting pulmonary outflow measured by PC flow imaging from total RV volume measured by cine SSFP imaging.

Ventricular anomalies typically seen in subjects with isolated pulmonary regurgitation following transcatheter valvuloplasty or surgical valvotomy include RV dilation with increased right ventricular end-diastolic and end-systolic volumes and diastolic flattening of the ventricular septum. RV systolic and/or diastolic function may be diminished in cases of long-standing severe pulmonary regurgitation, or if there is a combination of significant tricuspid and pulmonary regurgitation. There may be aneurysmal right ventricular outflow tract (RVOT) patch dilation in subjects whose RVOT surgical repair included patch augmentation that can contribute to RV dysfunction which can be visualized by cine CMR in the off-axis sagittal view. Although most patients with isolated pulmonary regurgitation have a dilated right ventricle, there is a subset of patients in whom the right ventricle was originally hypoplastic and despite longstanding pulmonary regurgitation following relief of right ventricular outflow tract obstruction, the right ventricle does not dilate commensurately. These subjects have abnormal right ventricular compliance and diastolic dysfunction that can be assessed by CMR.

**Anomalies with elevated right ventricular pressure load**

**Right ventricular outflow tract obstruction**

Right ventricular outflow tract obstruction may be isolated (such as subvalvar, valvar, supra-valvar or branch pulmonary artery stenosis) or associated with other lesions, such as a ventricular septal defect (VSD) and ventricular septal malalignment in tetralogy of Fallot. In each case, the specific type of lesion, degree of obstruction, and presence of associated defects will influence right ventricular geometry, mass and function; through ventricular-ventricular coupling, right ventricular outflow tract obstruction may also lead to anomalies of left ventricular anatomy and function, all of which can be accurately assessed by CMR.

Children with tetralogy of Fallot and other forms of right ventricular outflow tract obstruction
morphology and function can make the diagnosis and describe ventricular sepal defect(s) location and size, the degree and location of any mid-RV cavity or subvalvar pulmonary stenosis, and assessing the degree of compensatory or unexpected

Figure 8.2 (a) 4-chamber breath-hold SSFP image of a young adult with native Ebstein anomaly, demonstrating significant inferior displacement of the tricuspid hinge point (*) with abnormal attachments of the anterior tricuspid leaflet to the RV free wall (arrow). There is a large "atrialized" portion of the right ventricle (RV) with abnormal position of the ventricular septum in this region. (b) Short-axis cine SSFP image in the same patient, demonstrating the inferior displacement of the effective tricuspid orifice (*). (a) and (b) are courtesy of Dr. Tal Geva, Children's Hospital-Boston. (c) Free-breathing 4-chamber T1-weighted double inversion dark blood image of a six month-old infant with Ebstein malformation and pulmonary atresia, after Blalock-Taussig shunt. Note the inferior displacement of the septal tricuspid leaflet. On 4-chamber cine SSFP imaging (d), there is dephasing artifact from tricuspid regurgitation originating within the right ventricular body (*) as well as right ventricular hypertrophy and posterior bowing of the ventricular septum caused by supra-systemic right ventricular pressure (the right ventricular outflow tract was not opened in this case). RA: right atrium.

are often referred for pre-operative CMR in lieu of invasive cardiac catheterization in order to evaluate the branch pulmonary arteries when adequate assessment is not feasible by 2-D echocardiography. CMR imaging of right and left ventricular
ventricular hypertrophy. A combination of T1-weighted fast spin echo, cine SSFP imaging in four-chamber, short axis, right and left ventricular outflow tract planes, and PC flow measurement in the ascending aorta, MPA, and right and left branch pulmonary arteries is usually performed in addition to a gadolinium-enhanced MRA. As pre-operative assessment is typically performed in infants and young children unable to breath-hold, these sequences may be obtained via free breathing with multiple excitations or with respiratory navigation, or using suspended respiration under general anesthesia. A similar CMR imaging protocol is often performed in cases where a palliative pulmonary artery band has been previously placed prior to definitive surgical intervention in order to non-invasively assess branch PA anatomy; during CMR, ventricular morphology, function, degree of intracardiac shunting, and thoracic vascular anatomy can also be evaluated.

**Post-operative assessment following tetralogy of Fallot repair**

CMR has become the standard imaging modality to assess right (and left) ventricular volumes, mass, function, and the degree of right ventricular outflow tract obstruction and/or pulmonary regurgitation following surgical repair of tetralogy of Fallot [9,10], and to determine suitability for, and prognosis following, pulmonary valve replacement.

CMR ventricular evaluation of a patient after tetralogy of Fallot repair includes ECG-gated cine SSFP imaging in long axis, 4-chamber, short axis views of the right and left ventricles, and oblique axis imaging through the right and left ventricular outflow tracts, as well as measurement of net and regurgitant pulmonary and aortic flow via PC flow imaging [11]. Some have suggested that effective RV stroke volume (equal to the total RV stroke volume measured by cine SSFP imaging minus pulmonary regurgitant volume measured by phase-contrast flow imaging) is a more accurate means of assessing right ventricular function [12], but this is very controversial. Myocardial delayed enhancement (MDE) imaging can be included to assess the extent of ventricular fibrosis, particularly in the RVOT region following trans-annular patch/ventriculotomy [13]; the degree of MDE has been correlated with multiple parameters of right ventricular function. Myocardial tissue tagging can also be obtained to assess regional systolic and diastolic ventricular function and strain.

A number of recent studies have “defined” an upper limit of CMR-determined indexed right ventricular volume above which recovery of ventricular parameters, will not occur in patients with pulmonary regurgitation. For example, Oosterhof et al. reported that normalization of RV volumes occurred in those subjects with pre-operative RV end-diastolic volume indexed to BSA of less than 160 cc/m² [14]; although specificity was high, sensitivity was significantly lower. Another finding was that although RV ejection fraction itself did not improve, left ventricular stroke volume and diastolic volume increased, emphasizing the importance of ventricular–ventricular interaction. Right ventricular diastolic function has also been demonstrated by CMR to be impaired in children and adults following tetralogy of Fallot repair, and may impact the clinical course [15]. Figure 8.3 illustrates typical CMR findings in two patients following tetralogy of Fallot repair. In both cases, the right ventricle is severely dilated as a consequence of long-standing, severe, pulmonary insufficiency. There is aneurysmal dilation of the right ventricular outflow tract without significant right ventricular hypertrophy or residual right ventricular outflow tract obstruction.

**Post-operative right ventricular abnormalities following intra-atrial baffle for d-transposition of the great arteries**

Prior to the past two decades, children born with d-transposition of the great arteries (aorta arising rightward from the RV, pulmonary artery arising posteriorly from the LV) underwent an intra-atrial baffle procedure rather than the arterial switch operation that is more commonly performed today. Two types of intra-atrial baffles (Senning and Mustard procedures) were devised to direct systemic venous blood into the left atrium, across the mitral valve into the left ventricle and out the pulmonary artery, while pulmonary venous return is directed to the right atrium, across the tricuspid valve, into the right ventricle and out the aorta.
Figure 8.3 Breath-hold cine SSFP images in 4-chamber (a) and short axis (b) views after tetralogy of Fallot repair. There is severe right heart dilation caused by severe pulmonary regurgitation, without significant right ventricular outflow tract obstruction or right ventricular hypertrophy. (c) Demonstrates delayed enhancement along the right ventricular outflow tract region of this patient (arrow). (d) Demonstrates a right ventricular outflow tract cine SSFP view on a different subject after tetralogy of Fallot repair, revealing the limits of the patch reconstruction in the right ventricular outflow tract (RVOT) and mildly thickened pulmonary valve leaflets with a dephasing jet into the main pulmonary artery. There is severe pulmonary regurgitation (not shown) and a severely dilated right ventricle. LV: left ventricle; RA: right atrium; RV: right ventricle.
Abnormalities of the ventricles and pericardium

The Mustard and Senning procedures differ in the surgical details, but both result in the same postoperative physiology where the right ventricle serves as the systemic ventricle and the tricuspid valve as the systemic AV valve while the mitral valve and left ventricle supply the pulmonary circulation.

Atrial switch procedures for d-TGA lead to abnormalities in ventricular geometry and function; in particular, there is often systemic RV dysfunction and tricuspid regurgitation. Other abnormalities can also occur following atrial switch, including dynamic left ventricular outflow tract obstruction (sub-pulmonary stenosis), obstruction to either systemic or pulmonary venous return, intra-atrial baffle leaks, and (particularly atrial) arrhythmias such as sinus node dysfunction; these anatomic abnormalities can themselves lead to changes in ventricular geometry and function based on abnormal pressure and volume loads on both the right and left ventricles.

CMR has emerged as the non-invasive imaging procedure of choice to assess anatomy and function following atrial switch procedure including adults. Typical CMR evaluation includes assessment of right and left ventricular volumes, ejection fraction and mass via ECG-gated cine SSFP imaging, as well as evaluation of associated lesions, such as the degree of tricuspid regurgitation (by ECG-gated velocity-encoded cine PC MRI sequences and RV volumetric analysis), presence of associated lesions such as intra-atrial baffle obstruction, or dynamic sub-pulmonary stenosis/ left ventricular outflow tract obstruction caused by systemic RV pressure leading to posterior displacement of the ventricular septum. Delayed-enhancement imaging can be performed to look for regional hyperenhancement; the presence of RV delayed enhancement after atrial switch has been associated with increased risk of RV dysfunction and arrhythmia [16]; pharmacologic stress imaging, typically following dobutamine infusion, can also be performed.

Figure 8.4 illustrates ventricular geometry typical of a patient following atrial switch procedure with RV dilation and physiologic hypertrophy as well as posterior systolic bowing of the ventricular septum, while the left (pulmonary) ventricle is characteristically thin-walled.

**Right ventricular pressure overload: pulmonary hypertension**

Pulmonary hypertension is a common diagnosis leading to CMR referral; pulmonary hypertension may be idiopathic (so-called primary pulmonary hypertension), or it may be secondary to connective tissue diseases, pulmonary disorders, left-sided heart disease, or hematologic diseases such as sickle cell disease. Pulmonary hypertension not only results in abnormal right ventricular volumes and function that can be quantified by CMR, but can also lead to distortion of the left ventricular cavity through ventricular–ventricular interaction, thereby causing changes in global left ventricular geometry and function. A further advantage of CMR in evaluating subjects with pulmonary hypertension is that CMR facilitates evaluation of not only the heart, but can also evaluate the lungs and pulmonary circulation from both a structural and functional perspective.
Abnormalities in right and left ventricular volumes and systolic function in pulmonary hypertension patients can be quantified by acquiring a stack of ECG-gated cine steady state free precession images in the short axis plane encompassing the cardiac mass from base to apex. Although pulmonary vasodilator treatment has not consistently been associated with improvement in CMR derived parameters of right ventricular systolic function [17], right ventricular dilation and impairment in left ventricular filling have been associated with disease prognosis in idiopathic pulmonary hypertension [18].

CMR techniques have also been used to demonstrate impaired right ventricular diastolic function that can acutely improve following pulmonary vasodilator therapy in patients with pulmonary hypertension [19].

Recently, Marcus and colleagues [20] used CMR myocardial tissue tagging and strain analysis to demonstrate delayed left ventricular to right ventricular myocardial peak shortening velocity in pulmonary hypertension patients. They determined that this delay was not related to a delay in mechanical activation or to electrical conduction, but was related to an increase in right ventricular wall tension and longer duration of right ventricular shortening. This inter-ventricular delay was associated with posterior ventricular septal bowing, decreased left ventricular filling, and decreased left ventricular stroke volume [20]. Myocardial delayed enhancement imaging performed in patients with pulmonary hypertension has demonstrated punctate “insertion point” hyperenhancement at the superior and inferior junctions of the ventricular septum and in the right and left ventricular free walls in subjects with pulmonary hypertension; extension of the hyperenhancement into the ventricular septum has been correlated with more severe pulmonary hypertension and inversely correlated with right ventricular ejection fraction [21]. The physiological significance of this delayed enhancement in pulmonary hypertension patients remains unknown. Figure 8.5 is from the study by Marcus et al., demonstrating right ventricular hypertrophy and dilation with posterior bowing of the ventricular septum and delayed left ventricular contraction revealed through myocardial tissue tagging.

**Abnormalities of left ventricular structure**

**LV volume load abnormalities**

Many congenital and acquired anatomic cardiac and extracardiac abnormalities can result in left ventricular dilation with preserved left ventricular systolic function, each of which can be evaluated by CMR. These may include ventricular septal defects, valvar regurgitation (mitral or aortic), aorta to pulmonary artery shunts (such as patent ductus arteriosus, aorto-pulmonary window, or aorta to pulmonary collateral arteries), large fistulas or arterio-venous malformations, and anemia. A CMR examination performed to assess LV dilation would have a protocol similar to that described above for assessment of RV dilation, often with additional oblique spin-echo and/or cine SSFP imaging in orientation planes chosen during the CMR examination to more completely characterize ventricular and/or valvar anatomy.

**LV volume overload: ventricular septal defects (VSD)**

CMR can characterize intracardiac anatomy in children with ventricular septal defects, with or without associated intracardiac lesions especially VSD location and size or the geometric relation of the VSD to the outflow tracts vs. ventricles. CMR can also be used to quantify the degree and direction of intracardiac shunting by measuring RV vs. LV stroke volumes using cine SSFP imaging, corroborated with measurements of aortic and pulmonary flow obtained by PC flow mapping. CMR can be helpful in the assessment of associated cardiac lesions, such as valvar regurgitation, right ventricular outflow tract obstruction including “double chamber” RV (abnormal mid-cavity RV muscle bundles), or left ventricular outflow tract obstruction. Additional levels of intra- or extracardiac shunting can be evaluated, such as an ASD or PDA. Indirect CMR assessment of the degree of pulmonary hypertension can be made by evaluation of ventricular septal orientation and the degree of right ventricular hypertrophy; an anatomically-large VSD seen in the absence of left ventricular volume overload or right ventricular outflow tract obstruction, or low calculated Qp:QS,
Figure 8.5 MRI Cine and Tagged Images. Three-chamber images (top panels), short-axis images (middle panels), and short-axis tagged images (bottom panels), at the time of aortic valve closure at trigger delay of 252 ms (left) and the time of peak right ventricular (RV) shortening at 341 ms (right). The 3-chamber images show that maximal leftward septal bowing occurs at 341 ms, well after aortic valve closure. In the tagged image at 341 ms, the distance of the tagging lines in the RV free wall show further shortening (thick white arrows), whereas the tagging lines in the left ventricular (LV) free wall show relaxation. MRI: magnetic resonance imaging; ms: millisecond (Reprinted with permission from [20]).
would suggest pulmonary hypertension or possible Eisenmenger syndrome. CMR can directly measure pulmonary hypertension by obtaining maximum velocity of a tricuspid regurgitant jet with PC mapping, similar to echocardiography. VSDs seen in conjunction with more complex congenital heart disease can be further delineated by CMR, including assessment of the relation of the VSD to ventricular inflow and/or outflow anomalies.

Figure 8.6 displays a small to moderate perimembranous VSD in a young adolescent; CMR demonstrated a very mildly dilated left ventricle with Qp:Qs of approximately 1.5:1. There is no evidence elevated right heart pressure.

CMR can also define valvar abnormalities that are consequences of VSDs, such as sinus of Valsalva aneurysms; ruptured sinus of Valsalva aneurysms can present as sudden ventricular volume overload together with typical aortic abnormalities demonstrated by CMR. For example, Figure 8.7 illustrates a teenager with DiGeorge syndrome, shortness of breath, and a new murmur; a ruptured sinus of Valsalva aneurysm through a previously undetected VSD was demonstrated. There was right and left ventricular dilation and a calculated Qp:Qs of 2:1. Surgical inspection confirmed this diagnosis.

**Left ventricular volume overload due to mitral and/or aortic regurgitation**

CMR has been demonstrated quantify mitral and aortic regurgitation through a combination of LV volumetric analysis and PC flow mapping. Direct quantification of valvar regurgitation itself by PC flow mapping will not be described in detail in this

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**Figure 8.6** (a) 4-chamber cine SSFP image of a restrictive perimembranous ventricular septal defect in a young adolescent (arrow). The left ventricle is very mildly dilated; the right ventricle is not hypertrophied. (b) Oblique sagittal cine SSFP view demonstrating continuity between the VSD, left ventricular outflow tract, and tricuspid inflow (arrow). (c) Short axis cine SSFP systolic image demonstrating dephasing artifact from the accelerated VSD flow jet from the left ventricle (LV) into the right ventricle (RV) (arrow). RA: right atrium.

**Figure 8.7** (a) Short axis cine SSFP image at the level of the aorta (Ao), left atrium (LA) and right atrium (RA) demonstrating the distorted aortic sinus with a “windsoc” created by a sinus of Valsalva aneurysm. There is a small turbulent jet into the RA and right ventricle (RV) (dephasing artifact) that represents flow through the ruptured aneurysm. (b) 4-chamber cine SSFP image demonstrating the jet into the right atrium (arrow). (c) Diastolic short axis cine SSFP image demonstrating left ventricle (LV) and RV dilation. Ventricular systolic function was preserved.
chapter (for a review, see [22]) but is described elsewhere in this textbook; by using the techniques described above, CMR is ideal for serially assessing ventricular systolic and diastolic functional responses to valvar regurgitation as well as assessing the regurgitation itself.

**Left ventricular outflow tract obstruction: subaortic stenosis, aortic stenosis, aortic coarctation**

CMR is usually performed on infants and children with left ventricular outflow tract obstruction in order to assess ascending aorta and/or aortic arch anatomy; assessment of anomalies in ventricular morphology, mass, and function can also be determined. CMR is used to serially assess, with excellent accuracy and reproducibility, changes in left ventricular function or mass in order to determine need for intervention.

Ventricular abnormalities that are seen in subjects with left ventricular outflow tract obstruction include increased left ventricular mass and wall thickness, which is generally symmetric and commensurate with the level of obstruction; when increased afterload is uncompensated, left ventricular dysfunction can also be present. There can also be diastolic dysfunction that can lead to elevation in left atrial pressure, pulmonary venous hypertension with elevation in right ventricular pressure and right ventricular hypertrophy.

A CMR protocol performed to assess the ventricles in children with left ventricular outflow tract obstruction would include the same T1-weighted and cine SSFP 2-chamber, 4-chamber and complete short axis stack through the ventricular chambers, with additional cine SSFP acquisitions through the LVOT and aortic root as indicated. PC flow mapping across mitral inflow, aortic and pulmonary outflow, would usually also be obtained. Right and left ventricular mass, volumes, ejection fraction, and diastolic function parameters could thereby be assessed. Regional wall motion and diastolic function can be evaluated by myocardial tissue tagging sequences as well. CMR evaluation of adults with compensated aortic stenosis has demonstrated “supra-normal” LV systolic function but abnormal strain with reverse remodeling and normalization of left ventricular parameters following aortic valve replacement [23]. Delayed enhancement imaging may be performed; hyper-enhancement has been demonstrated along the endocardial surface of infants and children with severe valvar aortic stenosis [24]. The hyper-enhanced left ventricular myocardium appears to represent endocardial fibroelastosis (see section below).

**Left ventricular combined volume and pressure load: mixed aortic valve disease**

Many subjects have a combination of mixed aortic valve disease, with both aortic regurgitation and insufficiency. CMR is often used to serially assess for changes in ventricular volumes and/or hypertrophy that may indicate progressive valvar abnormalities which might warrant surgical intervention. A similar imaging protocol to that described for left ventricular outflow tract obstructive lesions would be used (Figure 8.8).

Figure 8.8a,b demonstrates breath-hold, cine SSFP CMR imaging in short axis and oblique left ventricular outflow tract views in an adolescent with a bicuspid aortic valve and a combination of aortic stenosis (moderate) and regurgitation (mild to moderate). Note the concentric left ventricular hypertrophy and the dilated aortic sinus. Figure 8.8c,d demonstrate CMR images from a different young adolescent with a bicuspid aortic valve, but whose predominant lesion is moderate to severe aortic regurgitation with only minimal aortic stenosis. The left ventricle is dilated but not significantly hypertrophied. There is dephasing artifact in the left ventricular outflow tract from the aortic regurgitant jet, as well as a dilated aortic sinus.

**CMR in children with unrepaired AV canal defect**

In general, these are often small infants in whom the morphology of AV valve attachments can be obtained using standard CMR techniques by paying close attention to technical details such as signal to noise, matrix size, bandwidth, echo train length, etc. CMR can quantify the degree of atrial and/or ventricular shunting (again, typically through a combination of cine SSFP evaluation of right vs. left ventricular stroke volume, together with PC flow imaging through the planes of the ascending aorta and main pulmonary artery),
assess for coexistent lesions such as anomalous venous drainage, additional shunts, or unrecognized branch pulmonary artery stenosis. There are also occasions when CMR may be beneficial to assist surgical planning of more complex AV canal defects or in those in whom a prior palliative procedure has been performed. For example, Figure 8.8 demonstrates CMR imaging performed in a 3-year-old with Down syndrome, a complete AV canal defect, and a relatively small RV; the patient had previously undergone PA band placement. The youngster was being evaluated for definitive surgical repair, but right ventricular size could not be adequately evaluated by 2-D echocardiography.

Figure 8.8 (a) Short axis cine SSFP imaging in an adolescent with a bicuspid aortic valve, moderate aortic stenosis and mild to moderate aortic regurgitation, demonstrating concentric left ventricular (LV) hypertrophy. (b) Left ventricular outflow tract (LVOT) cine SSFP view in same subject demonstrating aortic (Ao) sinus dilation as well as the left ventricular hypertrophy (LVH). (c) Short axis cine SSFP view in a different young adolescent with a bicuspid aortic valve, moderate to severe aortic regurgitation and left ventricular dilation. Note the dephasing artifact from the aortic regurgitant jets in the LVOT view (d), again with a dilated aortic sinus.
Figure 8.10 a,b illustrates free-breathing, non-sedated, CMR images in a young girl with severe aortic and pulmonary regurgitation after truncus arteriosus repair, resulting in significant right and left ventricular dilation. Transthoracic echocardiography could not adequately assess the degree of ventricular dilation, valvar regurgitation, or ventricular volumes due to the coexistence of right and left sided volume overload. CMR demonstrated severe aortic and pulmonary regurgitation, with both left and right ventricular end-diastolic volumes that had increased in size (indexed to body surface area) compared with prior CMR examinations. Figure 8.10 c–e illustrates CMR imaging performed on a young adolescent who had undergone truncus arteriosus repair in infancy, followed by prosthetic aortic valve replacement for severe aortic regurgitation. Note the biventricular hypertrophy due to combined aortic stenosis, severe stenosis of the RV-PA conduit, as well as significant bilateral branch pulmonary arteries. Hyperenhancement is demonstrated in the right ventricular outflow tract region and ventricular septum following gadolinium injection, as seen in the short axis delayed enhancement images. This patient presented with exercise-induced chest pain and syncope.

to decide if her right ventricular size would preclude two-ventricle repair.

**Combined RV and LV volume and/or pressure overload**

There is a subset of children after surgical repair of congenital heart defects in whom CMR is especially useful; subjects who have combined right and left semilunar valve regurgitation, (e.g. after truncus arteriosus repair or Ross procedure). In these lesions, there may be coexistent right and/or left ventricular outflow tract obstruction. Through the assessment of RV vs. LV stroke volume via cine SSFP imaging together with quantification of the degree of pulmonary and/or aortic regurgitation via PC velocity mapping, one can more accurately assess ventricular morphology, function, and anatomy in order to make educated decisions regarding clinical management and potential timing for further surgical intervention. In adolescents and young adults after Ross procedure, CMR has demonstrated abnormal left ventricular systolic function related to aortic root dilation, decreased aortic distensibility, and greater aortic regurgitation [25] (Figure 8.10).
Figure 8.10  (a) 4-chamber and (b) short axis free-breathing cine SSFP imaging in a young girl s/p repair of truncus arteriosus. Note the significant right and left ventricular (LV) dilation due to severe pulmonary and aortic regurgitation, respectively. (c) 4-chamber breath-hold cine SSFP imaging in an adolescent s/p truncus arteriosus repair and subsequent prosthetic aortic valve replacement. There is severe right and left ventricular hypertrophy due to combined RV-PA conduit stenosis, bilateral branch pulmonary artery stenosis, and valvar aortic stenosis. Short-axis myocardial delayed enhancement demonstrates hyperenhancement in the RVOT (d) and the ventricular septum (e), respectively (arrows).
Anatomic abnormalities associated with significant RV or LV hypoplasia: single ventricle physiology

CMR greatly facilitates evaluation of anatomy and physiology in subjects with significant RV or LV hypoplasia. Although dealt with in another chapter, CMR can be used to assess ventricular anatomy, morphology and function in infants and children throughout the stages of palliative surgical correction [26].

Abnormalities of cardiac situs, ventricular position or ventricular looping

Because CMR enables 3-D tomographic imaging in any unique plane, abnormalities of cardiac situs, ventricular position, and/or ventricular looping and their co-association with other intracardiac or extracardiac lesions are ideally investigated by CMR [1]. CMR is useful in demonstrating the relation between adjacent thoracic structures, and, together with abdominal imaging, can be used to rule out abnormalities of situs, that is, heterotaxy syndrome, with three-dimensional tomographic imaging made available by CMR techniques such as ECG-triggered T1-weighted spin echo and SSFP imaging. Heterotaxy and other complex congenital heart disease are discussed extensively in Chapter 15. Intrinsic morphologic differences between the right and left ventricle can be visualized by CMR in order to assess atrioventricular and ventricular-arteries concordance or discordance. For example, one can assess right ventricular morphologic characteristics such as a tripartite shape with inflow, body, and muscular outflow regions, septal tricuspid valve attachments, the presence of a moderator band or a more trabeculated appearance of the endocardium compared with the smooth-walled left ventricle. In this manner, abnormalities of ventricular situs, looping or position can be investigated.

CMR assessment of cardiac situs, ventricular looping, or ventricular positional abnormalities is often best begun through a series of axial ECG-gated spin echo or rapid SSFP images from the level of the outflow tracts to the diaphragm. In this way, one can systematically determine viscerocoeal situs and any abnormalities of systemic venous return, ventricular looping, and the relation of the outflow tracts to each other. Associated lesions, such as ASDs and VSDs, can be delineated, as can abnormalities of the atrioventricular and semi-lunar valves; associated cardiac anomalies are quite common in all situs abnormalities, with the exception of mirror-image dextrocardia with situs inversus. Abdominal situs and the tracheobronchial anatomy, key issues in heterotaxy syndromes, are easily delineated by one test – CMR.

Ventricular displacement due to extracardiac anomalies

CMR is often requested to evaluate ventricular anatomy, size, and function in instances where extracardiac anomalies may lead to displacement and/or deformation of the heart within the thorax. In these cases, it may be quite difficult using echocardiographic views to accurately assess the right and left ventricles. Examples may be thoracic or mediastinal masses, lung hypoplasia, diaphragmatic hernia, or when there are significant pectus chest wall abnormalities.

Situs inversus totalis with dextrocardia

Abnormalities of abdominal situs may occur as isolated defects, or can be seen in associated with significant structural heart disease. Situs inversus totalis with dextrocardia (i.e., mirror-image situs inversus) is often incidentally diagnosed by chest X-ray; situs inversus totalis is also demonstrated in approximately 50% of subjects with primary ciliary dyskinesia (Kartagener’s syndrome). CMR can usually confirm this diagnosis in children and assess for associated structural heart disease, which may be seen in some subjects. Using CMR myocardial tissue tagging, intrinsic differences in myocardial fiber architecture have been demonstrated in subjects with “isolated” situs inversus, with non-uniform systolic left ventricular torsion in situs inversus totalis, rather than torsion being strictly “mirror image” of that seen in the normal left ventricular. This abnormality of torsion, resulting from abnormal myocardial fiber architecture, may impact on long-term ventricular function in subjects with this rare defect [27].
**Congenitally corrected transposition of the great arteries**

Viscero-atrial situs solitus, L-ventricular looping and L-transposition of the great arteries is also known as “congenitally corrected” transposition since the physiological pattern of blood flow from the systemic veins through the heart to the lungs and body is preserved; this is the second most common type of transposition of the great arteries (Figure 8.11) [11]. Congenitally corrected transposition of the great arteries can occur as an isolated

**Figure 8.11** (a) Axial cine SSFP imaging at the level of the great arteries in a patient with L-looped transposition of the great arteries (L-TGA), demonstrating the aorta (Ao) leftward to the pulmonary artery (PA). (b) Axial cine SSFP imaging more inferiorly demonstrating a left-sided, morphologic right ventricle (RV) with coarse trabeculations and apical displacement of the tricuspid annulus (arrow). (c) Axial and (d)-(e) coronal cine SSFP imaging in a young adult with abdominal and atrial situs inversus, “congenitally-corrected” transposition (right-sided morphologic left ventricle (LV) and left-sided, morphologic right ventricle), VSD and pulmonary stenosis, who had undergone morphologic LV apical to PA conduit placement (*) and VSD closure. Note there is dextrocardia with ventricular hypertrophy due to obstructive thrombus in the LV apex to PA conduit. In this figure, labeling of the atria and ventricles are by morphology and not by anatomic location. RA: right atrium; LA: left atrium.
defect, or more commonly can be associated with other intracardiac abnormalities such as ventricular septal defects, Ebstein malformation of the (systemic) tricuspid valve, and/or sub-valvar and valvar pulmonary stenosis. Prognosis typically depends on systemic right ventricular function, degree of tricuspid regurgitation, and associated lesions [11].

Some subjects can have abnormal twist of the heart about the long axis during early embryologic development, resulting in crossing of the systemic and pulmonary flows at the mitral and tricuspid valve levels (so-called “criss-cross heart”); these patients typically have a superior-inferior relation between the ventricles with a horizontal position of the ventricular septum. This condition is virtually always associated with other congenital heart lesions, and usually these patients must undergo single-ventricle palliative repair due to their abnormal cardiac geometry. This abnormality is often quite difficult to conceptualize by 2-D transthoracic echocardiography, and CMR provides increased understanding of the 3-D relation of the atria, AV valves, ventricles, and outflow tracts in order to decide upon surgical options [28]. Some children with heterotaxy syndrome also must undergo staged single-ventricle palliative surgeries; CMR enables non-invasive serial assessment of ventricular morphology, size, and function through stages palliation without the need for ionizing radiation.

Figure 8.12 displays CMR images in two patients with heterotaxy syndrome. Figure 8.12a demonstrates a youngster with dextrocardia, right atrial isomerism, and complete AV canal defect with a common atria and single ventricle. Bilateral right atrial appendages are seen (*). Figure 8.12b through 8.12d demonstrate CMR images from a teenager with mesocardia (note the position of the heart relative to the spine), left atrial isomerism, double outlet right ventricle, VSD, and pulmonary steno-
Figure 8.12 (a) Axial cine SSFP free-breathing image of a three-year old with dextrocardia, right atrial isomerism, AV canal defect with common atrium and single ventricle. Note the bilateral right atrial appendages (*). (b–d) Breath-hold cine SSFP imaging in a different youngster with mesocardia, left atrial isomerism, double outlet right ventricle, VSD, and pulmonary stenosis, after VSD patch closure, RV-PA conduit and repair of total anomalous pulmonary venous return. There is RV-PA conduit stenosis (mild to moderate) and insufficiency (severe), resulting in right ventricular dilation and hypertrophy. There is systolic and diastolic flattening of the ventricular septum. The IVC crosses to the left of the aorta in the abdomen (white arrow). Ferro-magnetic artifact obscures the right ventricular outflow tract. (d) Oblique coronal cine SSFP image through the left ventricular (LV) outflow tract, demonstrating the angled VSD patch directing flow from the LV to the aorta (Ao) (black arrow).
CHAPTER 8 Abnormalities of the ventricles and pericardium

Abnormalities of the ventricles and pericardium

Consequence of the LV aneurysm, and assess for myocardial viability and scar formation [30].

Ventricular aneurysms in pediatric patients can also occur as the result of trauma, infection, surgical myocardial damage, or known coronary artery abnormality. Figure 8.13a,b demonstrates a case of blunt upper abdominal trauma that led to inferoseptal and apical LV aneurysm formation in a 3-year-old girl. Note that the myocardium is thinned with delayed hyperenhancement seen in the aneurysm that extends into the ventricular septum. (c) Short axis free-breathing cine SSFP image demonstrating a large right ventricular (RV) outflow tract pseudoaneurysm (*) due to dehiscence of the RV-PA conduit in an infant following tetralogy of Fallot repair. Note the dilated and hypertrophied right ventricle. LV: left ventricle.

Ventricular aneurysms and other regional abnormalities of ventricular structure and function

Isolated ventricular aneurysms

Congenital ventricular aneurysms are rare abnormalities whose etiology and even prognosis are unclear. Congenital ventricular aneurysms (most commonly left ventricular) may present with prenatal or post-natal ventricular arrhythmias, congestive heart failure, spontaneous aneurysm rupture, and sudden cardiac death. On the other extreme, ventricular aneurysms may simply be noted as an incidental finding performed for another indication. CMR offers a number of advantages over standard echocardiographic techniques in the evaluation of ventricular aneurysms, including enhanced ability to precisely localize the aneurysm within a particular three-dimensional segment of the ventricle (i.e., septal, apical, or free wall), quantitatively assess abnormalities of regional and global ventricular contraction as a consequence of the LV aneurysm, and assess for myocardial viability and scar formation [30].

Ventricular aneurysms in pediatric patients can also occur as the result of trauma, infection, surgical myocardial damage, or known coronary artery abnormality. Figure 8.13a,b demonstrates a case of blunt upper abdominal trauma that led to inferoseptal and apical LV aneurysm formation in a 3-year-old girl. Note that the myocardium is thinned with delayed hyperenhancement seen in the aneurysm. Pseudoaneurysms of the ventricle can also occur, when there is actual rupture of the ventricular wall that is contained by the pericardium and/or scar tissue; this may occur following myocardial infarction or, more commonly in children, following open-heart surgery. Figure 8.13c demonstrates a large pseudoaneurysm of the right ventricular outflow tract following tetralogy of Fallot repair in a patient who had significant residual bilateral branch pulmonary artery stenosis and a supra-systemic right ventricle, with dehiscence of the RV-PA conduit connection.

CMR assessment of post-operative segmental regional wall abnormalities

As more and more children undergo successful “definitive” or palliative surgical repair of congenital heart lesions, many are left with residual abnor-
malities of segmental ventricular function. Such abnormalities may be present prior to or immediately following surgery, or they may develop over time. Examples include right ventricular outflow tract aneurysms in children following tetralogy of Fallot or double outlet right ventricle repair, wall motion abnormalities near VSD patches, and segmental wall motion abnormalities due to coronary ischemia, such as in cases of anomalous coronary artery origin or following coronary re-implantation as a part of the arterial switch or Ross procedures. Segmental anomalies of ventricular function in children can be evaluated by a combination of cine CMR, ECG-gated cine gradient echo imaging rest and stress imaging, myocardial tissue tagging, first pass perfusion imaging and myocardial delayed enhancement imaging [31,32]. CMR has much better spatial resolution when compared with nuclear imaging and therefore may be better at diagnosing smaller perfusion defects, although temporal resolution is limited, particularly in children with high heart rates [31]. With the advent of better hardware and software, this problem has mostly been alleviated. Abnormalities in segmental ventricular function and the presence of infarcted myocardium have been demonstrated to influence clinical status, so their diagnosis by CMR is important for clinical decision-making algorithms; CMR can also assess post-operative improvement in global and segmental ventricular systolic function.

**CMR assessment of cardiomyopathies**

CMR has become the non-invasive procedure of choice to quantify and serially assess ventricular function and dimensions in adults with various forms of cardiomyopathy, including dilated, hypertrophic, restrictive, infiltrative cardiomyopathies, LVs with non-compaction, and arrhythmogenic right ventricular cardiomyopathies [33,34]; CMRs applicability in children with cardiomyopathy has also been increasingly appreciated.

**Dilated cardiomyopathy**

Dilated cardiomyopathy is characterized by impaired ventricular systolic function in the presence of increased ventricular end-diastolic and end-systolic volumes, whereas ventricular wall thickness is usually normal in dilated cardiomyopathy. Dilated cardiomyopathy in infants and children can be idiopathic or may be secondary to a metabolic or other genetic syndrome (such as muscular dystrophy), myocardial ischemia, myocarditis, or cardiotoxic chemotherapy. Dilated cardiomyopathy can also occur in pediatric patients following open-heart surgical repair of congenital heart defects.

Standard ECG-gated cine SSFP imaging can be used to quantify ventricular volumes, function and mass in children with dilated cardiomyopathy, and can be used to serially assess improvement or deterioration in ventricular function. Ventricular dilatation and dysfunction may be limited to the LV, or it may also include the RV; indeed, higher right ventricular ejection fraction appears to be an independent predictor of survival in subjects with idiopathic dilated cardiomyopathy [35]. One caveat to remember when one uses CMR to determine normal vs. abnormal ventricular function in children is that there are known to be age-related norms for LV systolic function in children as assessed by fractional shortening measured by 2-D echocardiography, with normal values for LV fractional shortening that are higher in infants and younger children. One would therefore expect that CMR-derived ventricular ejection fraction would also be age-dependent. Currently, however, comparative CMR-derived ventricular volumetric and functional measurements in normal young children have not been established. Serial measurements in the same patient, however, can easily be done.

The presence of myocardial delayed enhancement has been shown to be important in the differentiation of primary dilated cardiomyopathy from cardiomyopathy secondary to ischemic heart disease, and in determining prognosis of dilated cardiomyopathy, particularly in adult populations. Although the majority of subjects with dilated cardiomyopathy have not been demonstrated to have segmental hyperenhancement, likely due to the diffuse nature of myocardial fibrosis, there is a characteristic pattern when segmental myocardial delayed enhancement is seen. Non-ischemic myo-
cardial delayed enhancement typically involves the mid-epicardial portion of the myocardial wall in a linear fashion, whereas ischemic-etiologic MDE is subendocardial, may have transmural extension, and is located in a region corresponding to the perfusion territory of an epicardial coronary vessel [33]. In a study by Assomull and colleagues of patients with dilated cardiomyopathy, the presence of mid-wall myocardial delayed enhancement, seen in 30% of their subjects, was a predictor of the combined end point of all-cause mortality and cardiovascular hospitalization [41].

**Hypertrophic cardiomyopathy**

Hypertrophic cardiomyopathy is defined as an increase in ventricular wall thickness in the absence of, or out of proportion to, abnormal ventricular afterload. There are several different forms of hypertrophic cardiomyopathy, including symmetric and asymmetric (typically restricted to the ventricular septum or apex) and obstructive vs. non-obstructive. Echocardiographic characterization of ventricular mass and wall thickness is often problematic due to the non-uniformity of hypertrophy within the ventricular chamber, whereas CMR can more accurately assess 3-D ventricular wall thickness and mass (relative to body surface area) [40,41]. CMR can be used to assess the extent of LV outflow tract obstruction in cases of asymmetric septal hypertrophy, as well as to serially assess changes in ventricular mass and function following therapeutic interventions (such as surgical or medical treatments, including LVOT resection or septal ablation). Furthermore, CMR findings in hypertrophic cardiomyopathy may have prognostic implications; higher indexed LV mass has been associated with a poorer prognosis in adults with hypertrophic cardiomyopathy [41], and the presence of myocardial delayed enhancement has been demonstrated to correlate with the likelihood of ventricular dysrhythmias, perhaps since myocardial scars or fibrosis may represent potential foci for ventricular arrhythmias [42]. When present, delayed enhancement is typically localized to hypertrophied regions and to the junctions of the ventricular septum and RV free wall. Subjects with hypertrophic cardiomyopathy often have impairment in diastolic function, and may present with atrial enlargement due to elevated end-diastolic pressure. Figure 8.14 illustrates CMR images in a teenager with hypertrophic cardiomyopathy; the subject presented with increasing exercise intolerance and venous congestion, and was noted to have evidence of restrictive physiology in addition to the significant ventricular hypertrophy. Note the heterogeneous thickening of the left ventricular myocardium, compared with the more uniform left ventricular hypertrophy seen due to left ventricular outflow tract obstruction.

**Myocarditis**

CMR has been demonstrated to have several advantages over other imaging techniques in the diagnosis and evaluation of myocarditis. ECG-gated cine SSFP imaging can be used to accurately quantify 3-D ventricular volumes and function and serially assess temporal changes in these parameters. More importantly, CMR can be used as a diagnostic tool to differentiate acute myocarditis from dilated cardiomyopathy in subjects presenting with decreased ventricular systolic function of unknown etiology. For example, the presence of myocardial delayed enhancement (MDE) has been demonstrated to correlate with endomyocardial biopsy-proven evidence of myocarditis. Furthermore, the pattern of MDE within the myocardium has been shown to be associated with the particular viral etiology (i.e. subepicardial region of the LV lateral wall in parvovirus-B19 mediated myocarditis, and mid-ventricular septal MDE with human herpesvirus 6-associated myocarditis) [37]. This is in distinction to subendocardial hyperenhancement more typically seen with ischemic heart disease [38]. Although hyperenhancement has been demonstrated in subjects with myocarditis, there often remains some uncertainty as to whether the hyperenhancement is a direct result of inflammation, edema, or infarction/fibrosis. For this reason, recent CMR studies have suggested that T2-weighted imaging sequences should be added to CMR studies performed for evaluation of myocarditis vs. cardiomyopathy as an additional method to determine the extent of inflammation and/or edema [39].
Some adolescents and young adults are referred for CMR to help discriminate physiologic left ventricular hypertrophy due to “athlete’s heart” from pathological forms of ventricular hypertrophy such as hypertrophic cardiomyopathy. Endurance-trained athletes have been described as having increased ventricular volume load, whereas strength-trained athletes have increased ventricular pressure load, both of which may lead to increased left ventricular mass. In a small study, a CMR-derived measure that has been suggested to differentiate physiologic (e.g., “athlete’s heart”) vs. pathologic ventricular hypertrophy is a left ventricular diastolic wall thickness to volume ratio less than 0.15 mm $\times$ m$^2$/ml [43]; others have suggested measurement of CMR-derived ventricular mass and volume at baseline and after a several-month period of athletic deconditioning, after which time there should be a decrease in ventricular mass in non-pathologic cases.

**Non-compaction cardiomyopathy**

Non-compaction cardiomyopathy is another abnormality of the myocardium, characterized by a primitive, or “spongiform” appearance to the ventricular wall(s) with prominent trabeculations and deep inter-trabecular recesses, often in association with decreased ventricular systolic function. This form of idiopathic cardiomyopathy is thought to originate from suspension of normal myocardial development early in gestation. It can

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**Figure 8.14** (a) 4-chamber axial T1-weighted image in a teenager with hypertrophic cardiomyopathy. The patient also demonstrated findings of restrictive physiology, with significant bi-atrial enlargement and venous congestion. (b) 2-chamber cine SSFP systolic image in the same subject, demonstrating relatively thin left ventricle (LV) apex and more marked septal hypertrophy. (c) Diastolic short axis cine SSFP image demonstrating heterogeneous ventricular hypertrophy. LA: left atrium; RA: right atrium.
be diffuse through the RV and LV, or isolated to portions of the (generally left) ventricle. CMR has a number of advantages in evaluation of this form of cardiomyopathy over echocardiography, especially in the evaluation of apical LV non-compaction (perhaps the most common segmentally-affected region) and in myocardial tissue characterization. A recent, although small, study reported CMR-based diagnostic criteria for non-compaction cardiomyopathy, defining it as a ratio of non-compacted to compacted myocardial tissue greater than 2.3 at end-diastole [44]; studies have also demonstrated delayed hyperenhancement within the abnormal trabeculations.

**Restrictive and/or infiltrative cardiomyopathy**

Most cases of restrictive cardiomyopathy in pediatric populations are idiopathic. Restrictive cardiac physiology (as assessed by CMR) can also be seen in association with other cardiac or systemic diseases, such as endomyocardial fibro-elastosis, glycogen storage diseases, or radiation fibrosis; lymphoma, sarcoid, and amyloid are quite rare in pediatric populations but are more common etiologies for restrictive cardiomyopathy in adults.

Typical CMR findings in a subject with restrictive cardiomyopathy may include dilation of the atria and systemic veins with preservation of ventricular systolic function (ejection fraction) and low- or low-normal end-diastolic and end-systolic volumes; a pericardial effusion is also not uncommon. Diastolic flow patterns through the mitral and/or tricuspid valves obtained via PC flow mapping are usually abnormal. Finally, there may be elevation in pulmonary vascular resistance (as a result of increased left ventricular end-diastolic pressure) that can be indirectly assessed by CMR, or may be directly assessed using the maximum velocity of the tricuspid regurgitant jet via PC flow mapping.

CMR is frequently used to differentiate restrictive cardiomyopathy from constrictive pericarditis, as constrictive pericarditis can be successfully treated by pericardial stripping, whereas the prognosis for restrictive cardiomyopathy is quite poor and in general requires cardiac transplantation. This is one example where real-time cine CMR imaging can facilitate recognition of ventricular–ventricular interaction, as described below under constrictive pericarditis [45].

**Arrhythmogenic right ventricular dysplasia (cardiomyopathy)**

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD) is currently understood to be a disorder of desmosomal function, characterized by fatty and/or fibrous tissue replacement localized mainly to the right ventricle. Patients typically present with ARVD in the second to fifth decades of life, usually with palpitations, syncope, or sudden cardiac death [46]. Features of ARVD include segmental RV wall thinning and/or dysfunction, RV aneurysm formation, progressive RV enlargement, RV global dysfunction, ventricular arrhythmias, and sudden cardiac death; LV involvement has also been demonstrated. Although there is currently no definite diagnosis for ARVD, clinical task force criteria have been established to predict the likelihood of ARVD based on the presence of major and minor criteria in adults [46]. In addition to ECG abnormalities and a positive family history, major criteria include severe global or segmental RV dilation, decreased RV ejection fraction, RV aneurysm formation, progressive RV enlargement, RV global dysfunction, ventricular arrhythmias, and sudden cardiac death; LV involvement has also been demonstrated. Although there is currently no definite diagnosis for ARVD, clinical task force criteria have been established to predict the likelihood of ARVD based on the presence of major and minor criteria in adults [46]. In addition to ECG abnormalities and a positive family history, major criteria include severe global or segmental RV dilation, decreased RV ejection fraction, RV fatty infiltration, or RV aneurysm, whereas minor criteria include localized and less severe RV dysfunction. CMR is therefore an ideal non-invasive technique with which to evaluate subjects suspected to have ARVD, and evaluation for evidence of ARVD is a common indication for CMR in pediatric and young adult populations.

CMR studies to assess for ARVD are targeted at abnormalities of right ventricular myocardium and function. RV fatty infiltration and wall thinning is assessed through thin-slice T1-weighted spin echo imaging in axial and short axis views (with fatty infiltration appearing as a bright signal compared with darker, "normal", myocardium); fat-suppression by using a fat saturation pulse by CMR has been shown to be beneficial in question-able cases. Following this, ECG-gated cine SSFP in axial and short axis views through the entire right ventricle is performed, this can determine any segmental regions of abnormal right ventricular wall motion or lack of right ventricular wall thickening, together with quantitative assessment of global right ventricular volumes and ejection fraction which is used for right ventricular dilation and
systolic dysfunction. Left ventricular function is also assessed. Right atrial and ventricular enlargement as well as right ventricular outflow tract ectasia, also CMR signs of ARVD, can be assessed with cine. Finally, myocardial hyperenhancement is performed, as investigators have determined that the presence of right ventricular delayed enhancement correlates with histopathologically confirmed ARVD and has been demonstrated to predict inducible VT on programmed electrical stimulation in adults [47]. See figure 8.15 as an example of ARVD by CMR.

When interpreting the results of CMR performed to assess for evidence of ARVD, one should keep in mind that minor RV wall motion abnormalities are common in normal adult populations. The high sensitivity and low specificity of CMR to detect ARVD may be a result of task force criteria for ARVD diagnosis being less sensitive to mild, early forms of ARVD, as explained by Masci et al. [33].

CMR criteria for ARVD in pediatric patients are even less clear cut. Fogel et al. [48] employed adult CMR-derived diagnostic criteria for ARVD and determined that application of the adult ARVD criteria would yield diagnosis of ARVD in few children, even in those with a history suspicious for this diagnosis. They surmised that the paucity of positive CMR findings in at-risk children might have been due to ARVD typically manifesting at an older age, and suggested that serial CMR evaluations of these children might be indicated every two to three years [48].

Figure 8.15 demonstrates CMR imaging in a teenager with a family history of ARVD, who on echocardiogram was noted to have a dilated right ventricle. There are multiple small aneurysms demonstrated within the right ventricular free wall, together with right ventricular dilation, global dysfunction, and segmental dyskinesis; the anterior right ventricular free wall appears bright on T1-weighted imaging (Figure 8.15c), but dark on the corresponding fat-suppressed image (Figure 8.15d). This patient also had a small to moderate, apical muscular ventricular septal defect, seen in Figure 8.15b.

**Endomyocardial fibro-elastosis**

Endomyocardial fibro-elastosis (EFE) is a relatively rare abnormality of the ventricular endocardium characterized by replacement of viable endocardium by fibrotic tissue. Today, this is most commonly seen in some patients with Shone’s complex or hypoplastic left heart syndrome, although historically this can also be a primary etiology of cardiomyopathy. Tworetsky et al. recently described the applicability of CMR imaging for surgical planning in four infants with marginal left ventricular size following fetal cardiac intervention for aortic valve stenosis and post-natal EFE resection [24]; they found that endocardial fibro-elastosis, confirmed on pathologic examination, could be diagnosed through myocardial delayed enhancement CMR imaging, but not using standard spin-echo or gradient-recalled cine SSFP techniques, and that CMR delineation of EFE could subsequently be used to plan surgical EFE resection in the hopes of improving LV growth and ventricular function (Figure 8.16).

**Cardiomyopathies as consequences of other systemic disorders**

Cardiomyopathies that result from systemic disorders are rare in the pediatric population, including diseases such as sarcoid and amyloid-induced cardiomyopathy. Abnormal ventricular systolic function has been demonstrated in younger subjects by CMR in patients with a variety of different hemoglobinopathies, including sickle cell anemia and beta thalassemia, which have as a common denominator of iron overload due to chronic transfusion therapy.

CMR has been used to assess myocardial iron by the relaxation time constant T2* (1/R2*) of ventricular myocardium. Patients with thalassemia major who are found to have lower values of T2* (or high relaxation rates R2*) have been demonstrated to be at greater risk of systolic ventricular dysfunction, arrhythmias, and of need for medications than those who have normal values of T2* [49]. There are, however, a several technical factors that must be considered when one uses CMR for T2* analysis, as outlined recently by Wood et al. [50].

**Abnormalities of the pericardium**

CMR is the ideal imaging modality to evaluate abnormalities of the pericardium; for a recent
CHAPTER 8 Abnormalities of the ventricles and pericardium

Figure 8.15 (a) Axial and (b) short axis cine SSFP images in a young adolescent with a family history of ARVD. Several small aneurysms are seen along the RV free wall (arrows), along with RV dilation and abnormal septal motion. A small muscular VSD is demonstrated in (b) as well (*). (c) Axial T1-weighted double inversion dark blood imaging demonstrates increased signal intensity along the RV free wall, which appears dark on fat-suppressed imaging (d); this is consistent with fatty infiltration. LV: left ventricle; RV: right ventricle.

review, see Ordovás et al. (2008) [34]. CMR allows evaluation of the entire pericardial space and its relation to particular cardiac and thoracic structures without the radiation inherent in chest computed tomography (CT); it also enables assessment of physiologic compromise in cardiac function that can result from pericardial abnormalities, such as pericardial tamponade or diastolic dysfunction. CMR offers advantages over echocardiography such as larger field of view, better soft tissue contrast, and more precise delineation of the location and tissue characterization of pericardial abnormalities.

Pericardial effusion

Normal individuals typically have a small amount of fluid in the pericardial space, but CMR can delineate the location and size of pathological and/or clinically significant pericardial effusions. Common causes of a pericardial effusion in children include infection (particularly viral, less likely bacterial or tuberculous), heart failure, renal failure, or as a consequence of injury to the pericardial sac (such as following open-heart surgery). Characterization of the location, degree, and type of pericardial effusion are performed by
Constrictive pericarditis

Constrictive pericarditis is unusual in the pediatric population; it should be considered in children with clinical evidence restrictive cardiomyopathy, as both conditions may present with similar clinical findings. Restrictive cardiomyopathy has no “cure” short of cardiac transplantation whereas, constrictive pericarditis is curable through surgical pericardiectomy. For adults, CMR is considered by many to be the “gold standard” technique to diagnose constrictive pericarditis when greater than 4 mm thickening of the pericardium is seen in the context of supportive clinical findings [34]. In children, increased pericardial thickness of less than 4 mm can be hemodynamically important. CMR criteria for pericardial thickening in children have not been established. Even in the absence of marked pericardial thickening, CMR can be of assistance in diagnosis of pediatric constrictive pericarditis through the demonstration of a mildly thickened but non-uniform-appearing pericardium, inferior vena caval distension, bi-atrial dilatation, relatively “underfilled” appearance of the ventricles, and abnormal ventricular septal motion [51].

CMR can also be used to assess for evidence of tamponade physiology caused by the presence of a pericardial effusion; one can identify right ventricular free wall collapse in early diastole, and right atrial free wall collapse in late diastole, by gradient echo-recalled cine or SSFP.

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in adults. This diagnosis is typically suspected when there is an abnormal chest X-ray in an otherwise well child. CMR characteristics of pericardial constriction include accentuated early diastolic septal motion posteriorly toward the left ventricle just after the onset of inspiration (“dancing septum”), whereas there is no significant respiratory variation in septal orientation seen in restrictive cardiomyopathy [33,45].

**Partial or complete absence of the pericardium**

This very rare defect is typically an isolated abnormality; it is most often contemplated as a diagnosis when a chest X-ray demonstrates an abnormal cardiac silhouette in an otherwise asymptomatic...
individual. Typical chest X-ray findings could include extreme levocardia, prominent main pulmonary artery segment, and interposition of lung tissue between the aorta and main pulmonary artery and a “box shaped” heart. Absence of the pericardium can also be seen in conjunction with other congenital heart defects prior to any intervention. The clinical importance in isolated partial absence of the pericardium is a small risk of herniation of the adjacent cardiac structure through the pericardial rent, which can lead to a compromise in coronary perfusion, chest pain, arrhythmias, myocardial ischemia, and even sudden cardiac death. T1 weighted images visualize the pericardium as gray in between the bright fat of the pericardium and epicardium. Fat suppressed gradient echo imaging nulls fat from the pericardium and epicardium and the pericardium itself is bright. CMR findings parallel the chest X-ray abnormalities described above [34,53]. CMR can also demonstrate, by cine, structures such as the left atrial appendage, jutting out from the pericardial sac and appearing enlarged (Figure 8.18).

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CHAPTER 9

MRI in conotruncal anomalies
(except tetralogy of Fallot)

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Introduction
Conotruncal anomalies are a group of congenital heart defects involving the outflow tract and large vessels with the same root cause – a defect in conotruncal formation in utero. Tetralogy of Fallot, transposition of the great arteries, double outlet right ventricle, congenitally corrected transposition of the great arteries, truncus arteriosus, interrupted aortic arch and aortopulmonary window are considered part of this group of malformations. All of these lesions have become amenable to surgical repair and/or catheter interventions. This involves staged procedures in some lesions. Residual problems are common and may require re-intervention. Therefore, detailed information on anatomy and function of the heart and large vessels is required, not only at initial diagnosis but also during long-term follow-up. Considering the combination of intra- and extracardiac (large vessel) abnormalities, the often complex spatial relationships between the anatomical structures and the important hemodynamic consequences, both before and after (surgical) correction, MRI has become an important diagnostic tool both pre- and post-operatively [1,2]. In this chapter the role of MRI in the evaluation of the different conotruncal anomalies (except tetralogy of Fallot) will be discussed. Because of its frequency in both referrals to CMR as well as epidemiologically, tetralogy of Fallot has a separate chapter (see Chapter 10).

Transposition of the great arteries
Transposition of the great arteries is one of the most common types of cyanotic congenital heart disease, comprising approximately 3% of congenital heart malformations in newborns [3]. In isolated (D-) transposition of the great arteries, atrial situs is normal, as is the atrioventricular connection. Both ventricles are normal and well developed. The aorta arises from the right ventricle, the pulmonary artery from the left ventricle. This may result in severe hypoxemia shortly after birth.

Common associated lesions include a ventricular septal defect (approximate 15–25% of cases), left ventricular outflow stenosis (approximately 5–15% of cases), and aortic arch anomalies (12%) [4,5]. MRI can depict the abnormal position of the great arteries, with the aorta anterior to the pulmonary artery and arising form the right ventricle [6–8] (Figure 9.1). Treatment strategies have included two types of procedure in the last decades. From the late 1950s to the end of the 1970s–early 1980s, atrial redirection was the procedure of choice. Two techniques were used, the Mustard and Senning procedures [9,10]. Both used the principle of redirecting the blood at atrial level in such a way that pulmonary venous blood drained into the right ventricle (RV) (and subsequently into the aorta), systemic venous blood into the left ventricle (LV) (and pulmonary artery), thereby
Techniques of assessment of biventricular function and valve flow have been discussed elsewhere in this textbook. More than half of the patients demonstrate signs of diminished systemic ventricular function at 25 years after the atrial redirection procedure [12]. The exact causes of this decline remain subject of debate. They may include tricuspid valve incompetence, demand ischemia from excessive hypertrophy, impaired coronary perfusion and limited preload reserve [11,13,14]. Assessment of late gadolinium enhancement (LGE), according to methods common in ischemic heart disease, has revealed LGE in considerable percentages in post-atrial correction patients. In a small number of patients, LGE correlated with age, lower RV ejection fraction, RV wall stress, oxygen uptake and arrhythmias and clinical deterioration with follow-up [15]. MRI combined with physical or pharmacological stress has been used to demonstrate diminished functional reserve in these patients, but the prognostic role of stress MRI in this situations has not been established [16–19].

Evaluation of the venous pathways (baffles) can be performed with transthoracic and transesophageal echocardiography; however, MRI is an attractive complement [20,21] and is routinely used to image the 3D anatomy and function, especially in patients with poor windows. 3D contrast MRA can

restoring the normal circulatory pattern (Figures 9.2–9.4). In the Senning operation, atrial tissue was used for this rerouting; in the Mustard procedure pericardium or artificial tissues. In this situation the right ventricle remains the ventricle supporting the systemic circulation. Right ventricular failure, obstruction of the atrial pathways (“baffles”) and supraventricular arrhythmias are common problems after the atrial redirection procedures [4,11].

Post-operative evaluation is a common indication for MRI after Mustard and Senning procedures [1]. Evaluation commonly includes evaluation of large vessel anatomy, atrial baffle anatomy and function, biventricular function, atrioventricular valve flow, and semilunar valve flow.

**(Figure 9.1)** Axial SSFP image of transposition of the great arteries. Ao: aorta; PA: pulmonary artery.

**(Figure 9.2)** Axial SSFP image of transposition of the great arteries after Mustard operation. Ao: aorta; LV: left ventricle; Arrow: superior cava vein, baffle to LV.

**(Figure 9.3)** SSFP image of transposition of the great arteries after Mustard operation. Modified 4-chamber view. RV: right ventricle; LV: left ventricle; Arrow: pulmonary venous baffle to RV.
problems are related to the role of the pulmonary valve and root in the systemic circulation, resulting in aortic root dilatation and aortic valve regurgitation. Other problems occurring during late follow-up after the arterial switch operation include stenosis of re-implanted coronaries, impaired ventricular function, as well as distortion of the pulmonary valve and root. 

Currently, combined standard balanced SSFP, phase contrast and 3D MRA sequences are commonly applied for these purposes (see Figures 9.1–9.6).

After the introduction of the arterial switch operation in the late 1970s, this operation has become the preferred technique for most patients with transposition of the great arteries [4]. Long-term results are excellent [23]. Major residual
monary arteries and right ventricular outflow tract obstruction. MRI is the most important diagnostic tool for assessment of large vessel anatomy and function after the arterial switch [24]. This requires combined use of black-blood and SSFP techniques for anatomical evaluation, cine SSFP for biventricular function as well as phase contrast measurements for quantification of suspected stenosis or regurgitation. Often the evaluation is combined with 3D contrast enhanced studies (see Figures 9.7 and 9.8). Although technically challenging, MR coronary angiography has been demonstrated in post-ASO patients [25] and should be part of routine evaluation. Late gadolinium enhancement has been used to detect abnormalities in myocardial viability that may have functional consequences [25]. Recently, RV diastolic function abnormalities have been reported as potential markers of early disease at long-term follow-up after ASO [26,27]. MRI may have an important role in detection of RV abnormalities in these patients.

In transposition of the great arteries with subpulmonary stenosis and ventricular septal defect (VSD), the Rastelli procedure may be considered. In this procedure the LV is rerouted to the aorta by means of an intracardiac tunnel through the VSD (see Figure 9.9). The right ventricle is connected to the pulmonary artery by placement of a conduit. Evaluation of conduit stenosis, residual VSD and left ventricular outflow tract obstruction are the most common indications for MRI after the

Figure 9.7 Axial SSFP image of transposition of the great arteries after arterial switch operation. Note abnormal position of left pulmonary artery branch anterior to aorta after “Lecompte” maneuver, and compression of left pulmonary artery branch by posterior aorta. Ao: aorta; PA: pulmonary artery.

Figure 9.8 Volume rendered 3D reconstruction of transposition of the great arteries after arterial switch operation. Note abnormal position of left pulmonary artery branch anterior to aorta after “Lecompte” maneuver. Ao: aorta; PA: pulmonary artery.

Figure 9.9 SSFP oblique image of transposition with VSD after Rastelli procedure. The LV has been tunneled to the aorta. Ao: aorta; LV: left ventricle; Star: ventricular septal defect (VSD).
Rastelli procedure. This includes assessment of conduit anatomy with black-blood and steady state free precession (SSFP) techniques, evaluation of function (maximum flow velocity, regurgitation fraction) with phase contrast sequences, and assessment of anatomy of the conduit and pulmonary arteries with contrast angiography. Other complications of the Rastelli procedure may include residual VSD and LV outflow obstruction [28].

**Double outlet right ventricle**

Double outlet right ventricle is a rare congenital heart malformation, characterized by a ventriculo-arterial connection in which both great vessels arise predominantly from the right ventricle. A VSD is nearly always present, and provides the outlet for the LV. The LV can be of normal size, but may also be hypoplastic. There may be abnormalities of the mitral valve. The relationship between the VSD and the large vessels and the subarterial anatomy may vary considerably. The VSD in DORV may be located in direct relationship to one of the great vessels (“committed”), or may have no relationship with one of the semilunar valves (“non-committed”). If an outlet septum is absent, the VSD may be related to both great vessels directly (“doubly committed” VSD). The relationship of the VSD and the large vessels has guided the most common classification system of DORV, which recognizes four subcategories: (1) subaortic VSD with or without pulmonary stenosis; (2) subpulmonic VSD; (3) doubly committed VSD; (4) non-committed VSD [29]. This variation is of direct consequence to surgical management strategies [30]. Therefore, detailed anatomical work-up assessing ventricular size, AV valve size and morphology, type, size and location of the VSD, and spatial relationships between the VSD and semilunar valve areas is mandatory before surgery. This is aimed at making a correct choice for either biventricular repair, in cases where the VSD can be closed to connect the LV to the aorta, or a Fontan (univentricular) type of repair in which details of anatomy are unfavorable for biventricular repair. MRI has replaced invasive cine-angiography in many cases, particularly if large vessel anatomy needs to be imaged [31–33] and to help decide one versus two ventricle repair. Axial, coronal, sagittal and selected angulated views to image the ventricular septum “en face” have been used for evaluation of DORV [32–34], with agreement with findings at surgery (Figure 9.10). 3D isotropic MR imaging may be highly useful for evaluation of this type of complex anatomy [8]. CMR is extremely useful in evaluating biventricular function and size in this type of lesion along with evaluating mitral valve morphology.

Biventricular repairs include closure of the VSD in case of subaortic VSD without additional malformations, VSD closure and arterial switch in cases of subpulmonic VSD, and more complex operations creating intraventricular rerouting [30,35,36]. In approximately 5% of cases, RV-PA homografts have been used [30]. Residual problems after biventricular repair include residual VSDs, semilunar valve stenosis and/or regurgitation, ventricular outflow tract stenosis, homograft stenosis and large vessel problems.

Post-operative evaluation of patients treated for DORV therefore includes systematic evaluation of intracardiac and large vessel anatomy and biventricular, valve and conduit function. Currently, combined black-blood, standard balanced SSFP, phase contrast and 3D magnetic resonance angiography (MRA) sequences are commonly applied for this purpose. This is illustrated in Figures 9.10–9.13.

**Congenitally corrected transposition of the great arteries**

Congenitally corrected transposition of the great arteries is a rare heart disease, accounting for approximately 0.5–1% of all cases of congenital heart abnormalities. The lesion is characterized by ventriculoarterial as well as atrioventricular discordance. The right atrium is connected to the left ventricle, which is connected to the pulmonary artery. The left atrium is connected to the right ventricle that connects to the aorta (Figures 9.14 and 9.15). The circulatory pattern that arises from this configuration is normal, that is the systemic and pulmonary circulation are arranged in series. In the large majority of cases, additional anatomical abnormalities are present, such as a ventricular septal defect (in 70% of cases), abnormal tricuspid valve (90%), including Ebstein’s anomaly and pulmonary stenosis (40%), either valvular, subvalvu-
Figure 9.10 Spin echo images of patient with double outlet right ventricle. Axial image orientation from caudal to cranial (a–f). Courtesy of R. Beekman, MD, Department of Pediatric Cardiology, Leiden University Medical Center, Leiden, The Netherlands. Ao: aorta; LA: left atrium; OS: opening; PA: pulmonary artery; *: represents a ventricular septal defect.
lar or supravalvular [4,37]. Congenitally corrected transposition is also referred to as L (levo)-transposition of the great arteries (L-TGA). L-TGA is found in association with dextrocardia in approximately 25% of cases (Figure 9.15). CMR has been recognized as a wonderful imaging tool to diagnose
and additional procedures in case of biventricular repair, or a Fontan procedure in case of unbalanced ventricles [41]. Alternatively a double switch procedure may be performed, consisting of a combination of an atrial rerouting procedure (Senning or Mustard operation) and arterial switch or Rastelli operation [42,43].

In post-operative patients with congenitally corrected TGA, the goal of CMR in general is to evaluate residual anatomical and functional abnormalities. A routine study would include assessment of intracardiac and large vessel anatomy, AV valve, aortic and pulmonary flow and RV function.

**Truncus arteriosus**

Truncus arteriosus (TA) is a rare congenital cardiovascular malformation caused by a failure in conotruncal septation of the embryonic truncus arteriosus and conus [44]. TA represents about 2–4% of all heart malformation [45]. The clinical course of these patients is variable, but without surgical intervention in early infancy or childhood, only fewer than 25% of children with TA will survive beyond the first year of life [45,46]. The classic presentation is a cyanotic patient with progressive tachypnea and cyanosis accelerating to congestive heart failure [44,47].

In TA, a single arterial trunk arises from the heart giving origin to the coronary arteries, the pulmonary arteries and the systemic arterial circulation [48]. In virtually all patients with TA, there is a large ventricular septal defect (VSD) and a single semilunar valve [47,48].

Several classification schemes have been proposed previously, of which those of Collett and Edwards, and Van Praagh are most widely used [47–49]. These classifications schemes are invariably based on the position of the main pulmonary artery segment and branch pulmonary arteries. Furthermore, the Van Praagh classification distinguishes type A and type B truncus arteriosus with type B being TA without a VSD [50]. The latter probably represents a large aortopulmonary window or aortic atresia [48,49]. Type 1A and 2A represent approximately 92% of all TA cases [45].

- **Type 1A:** proximal single arterial trunk with a distal main pulmonary artery segment.
patients have truncal valve regurgitation in various degrees ranging from mild to severe [51].

Numerous associated cardiovascular anomalies have been described of which variant coronary artery anatomy is surgically relevant [48]. This anomaly, common in type 4A, frequently involves a right coronary origin from the left anterior descending coronary artery. The origin of the left coronary artery is often rather distal at the posterior margin of the common trunk close to the branch pulmonary arteries and the ostium may be narrowed [45,46,48]. Other cardiovascular associations are less frequent and include right aortic arch, persistent left superior vena cava, aberrant origin of the left subclavian, patent foramen ovale, partial and complete atroventricular canal defects, mitral and tricuspid malformations, double-inlet or hypoplastic left ventricle, left pulmonary artery sling and anomalous pulmonary

- **Type 2A**: left and right branch pulmonary arteries originate from the posterior margin of the trunk (i.e., no main pulmonary segment is present).
- **Type 3A**: the origin of one pulmonary branch artery is truncal and the contralateral is ductal or aortic of origin (or collaterals).
- **Type 4A**: large ductus arteriosus supplying the aorta descending with a coexisting hypoplastic or interrupted aortic arch (usually type B) supplying the innominate arteries.

The annulus of the single semilunar valve is larger than a normal semilunar valve [51]. This truncal valve contains 2, 3, 4 or more leaflets which may be thickened or nodular [48,51]; most are tricuspid (70%) or quadracuspid (23%) [52]. The semilunar valve usually overrides the interventricular septum with dominance above the right ventricle (42%), but may occasionally override the left ventricle (16%) [47,48]. About 28% of TA

![Figure 9.16](image-url) (a) Frames from 3D gadolinium enhanced MRA of truncus arteriosus, interrupted aortic arch (type A), single RV. AO: aorta; LPA: left pulmonary artery; PDA: patent ductus arteriosus; RA: right atrium; RPA: right pulmonary artery; RV: right ventricle. Courtesy of E. Valsangiacomo, MD, Dept. of Pediatric Cardiology, Children’s Hospital, University of Zurich, Zurich, Switzerland. (b) Volume rendered 3D reconstruction of truncus arteriosus and interrupted aortic arch (type A). View from posterior. Ao: aorta; PA: pulmonary artery; PDA: patent ductus arteriosus.
venous connections [48,53]. Truncus arteriosus occurs relatively frequently in the setting of DiGeorge syndrome.

Although other imaging modalities are utilized prior to CMR for TA [47,54], it can provide additional as well as complementary diagnostic information in complex cases or in delayed diagnosis [46]. The pioneering work by Mansfield and co-workers in the early 1980s in diagnostic images in a type 2A truncus arteriosus patient, using an echo planar imaging technique with 3 mm in-plane resolution and 35 msec temporal resolution [55–57]. Ever since, however, only a few reports have been published on this subject owing to the rarity of this lesion. With recent developments, however, in new pulse sequences, parallel imaging techniques, multichannel coil designs and possibly also blood pool contrast media, a complete anatomical description may be obtainable with most clinical imagers, and should include the type of truncus arteriosus (including origin of pulmonary trunk, branches and collaterals), functional abnormalities of the truncal valve (regurgitation, stenosis, morphology of the valve, number of cusps), alignment of the truncal valve with respect to the ventricular septum, brachiocephalic vessels, pulmonary veins, and aorta, associated cardiac anomalies, mediastinal structures (hypoplasia or absence of thymus) (Figure 9.16). Hemitruncus [58] should not be confused with TA.

Surgical correction is completed in the neonatal period in order to avoid the severe morbidity and progressive pulmonary vascular obstructive disease [45]. Mortality is associated with concurrent interrupted aortic arch and the moderate to severe pre-repair truncal regurgitation [45].

The surgical one-stage approach of truncus arteriosus type A1 and A2 is, firstly, separation of the right and left branch pulmonary arteries from the arterial trunk and association of those branch pulmonary arteries with the RV. Most commonly the right ventricular outflow tract (RVOT) is reconstructed with a conduit [45,48]. Initially the RVOT was reconstructed with porcine valve in a synthetic tube, but more recently cryopreserved aortic and pulmonary valved allografts are more frequently used [45,51]. A direct anastomosis between RV and pulmonary arteries is rarely viable [45]. Reoperations of the anastomosis are almost inevitable because of conduit stenoses and regurgitation in the homograft. The size of the conduit is related to the rate of conduit stenosis during follow-up, with earlier failure of smaller sized conduits [45,51]. Also, aneurysms may develop at the location of surgical incisures such as adjacent to the homograft, but also after ventriculotomy [59,60]. MRI is useful to evaluate the anatomical and spatial relations between such pseudoaneurysm and its surrounding structures [59]. Secondly, the aortic root defect is closed primarily or, in larger defects, with a patch (pericardial or artificial) [45]. With regard to the truncal valve, no repair is performed in mild cases of regurgitation [51]. In moderate to severe regurgitation or stenosis, the preferred technique is valvuloplasty. Previously, homograft replacements and mechanical valve replacements were employed, but these resulted in disappointing outcomes [61]. After valvuloplasty, most truncal valves have mild residual regurgitation [51]. Both patients without truncal valve surgery and patients with repair or replacement should be monitored for progressive valve dysfunction [45]. Finally, the VSD is closed, usually by a patch applied through a transventricular approach or right infundibulotomy [45,51]. In a minority of patients a residual VSD remains [45].

One-stage repair is also the method of choice in infants with TA and associated anomalies. In type A4 truncus arteriosus, that is, with aortic arch hypoplasia or interruption, the aortic arch is reconstructed with direct anastomosis between the aortic arch and descending aorta [62]. A direct anastomosis, however, might potentially result in tension on the anastomosis and compression of the bronchovascular structures, and patch augmentation may be required or, alternatively, a Lecompte maneuver may be performed [62]. There is only a small risk of recurrent or residual anatomic and hemodynamic complications such as obstruction or pseudoaneurysm formation.

Cardiovascular MR has a greater role in the follow-up of patients with truncus arteriosus after surgical repairs than prior to repair. The issues that are encountered and which can be detected and quantified with imaging include [63] obstructive and aneurysmatic changes in conduits and anastomosis, changes in function of homograft and
reconstructed valves (regurgitation and stenosis), anatomical and functional imaging of both ventricles, vessel imaging (aorta, brachiocephalic arteries and pulmonary branches), residual intracardiac shunts, imaging of the associated cardiovascular malformations and reconstructions.

The MR pulse sequences required to complete the anatomic and functional assessment are described elsewhere and should at least encompass cine imaging with balanced steady-state free precession pulse sequences for functional and volumetric imaging of both ventricles and assessment of cardiovascular anatomy and valvular jets, phase contrast imaging for assessment of valve function, and 3D MR angiography (MRA) for evaluation of vessels and shunts. The older time-resolved MRA (e.g., TRICKS) in larger patients has limited value because of reduced in-plane resolution, that is, conventional MRA provides superior image quality [64,65]. However, with newer sequences with higher spatial and temporal resolution (e.g., TWIST), time resolved CMR can play a very useful role. It has recently been shown that in patients with RVOT reconstruction and patch closures of ventricular defects, these anatomical structures demonstrate delayed enhancement after administration of intravenous gadolinium [66]. This was attributed to fibrous tissue which may be relevant in the development of conduit stenosis. In case of repair of aortic arch hypoplasia or interruption, fast spin echo black-blood inversion recovery imaging enables the visualization of mediastinal and in particular the bronchovascular structures.

**Interrupted aortic arch**

Interrupted aortic arch (IAA) describes a point of atresia or loss of luminal continuity between the ascending and descending thoracic aorta, with some having remnant fibrous tissue between both segments [67,68]. Although all patients have an open ductus arteriosus [69], patients become invariably critically ill with closure of the ductus, regardless associated cardiovascular anomalies [67] and, untreated, patients will die within 4–10 days, following closure of the ductus arteriosus [69,70]. Only a few cases have been documented in adults, all without concomitant anomalies [71–74]. The universally used classification system by Celoria and Patton is based on the side of interruption with regard to the brachiocephalic arteries [75]:

- **Type A**: interruption distal to the left subclavian artery (25–30%).
- **Type B**: interruption between the left common carotid artery and left subclavian artery (70%).
- **Type C**: between left and right common carotid arteries (1–4%) [67,69].

Isolated IAA is rare and >98% of patients have associated cardiac anomalies, most commonly a VSD (>85%) [69,70,72]. Over half of VSDs are perimembranous with or without a posterior deviation of the infundibular septum [69]. Therefore, obstruction of the LVOT is commonly present with subaortic stenosis in 43% and valvular disease in 13% [69]. The aortic valve may be bicuspid [67]. Other frequent associations are truncus arteriosus and an aberrant right subclavian artery. Less commonly, more complex cardiovascular malformations can be found, like transposition of the great arteries, double outlet right ventricle, hypoplastic left ventricle, aberrant systemic venous drainage, pulmonary sling and aortopulmonary window [67,69,70,76]. A non-cardiac finding is DiGeorge syndrome in >25% [68,69].

Neonates with IAA present within several days after birth with tachypnea, cyanosis and progressive congestive heart failure. In the presence of a large ductus arteriosus, echocardiography [69] may have difficulties in delineating the distal aspect of the aortic arch, and moreover, echocardiography may not identify the aberrant brachiocephalic arteries [73]. Cardiovascular CMR is an established and comprehensive technique for evaluating the aortic arch in both adults and infants (e.g., coarctation [64,72,77,78]). Particularly, 3D contrast enhanced MRA can provide diagnostic information on the aortic arch [79], as adjunct to more conventional anatomical imaging of the mediastinum using spin echo pulse sequences [73] (Figure 9.16). 3D MRA is more reliable and effective than echocardiography in making correct diagnosis [71] and has the potential to replace cardiac catheterization [80]. Optimal synchronization of the contrast bolus and image acquisition in infants is, however, challenging (but not impossible) as circulation times are short and because of com-
plicating abnormalities such as truncus arteriosus, intra- and extracardiac shunting and collaterals. Time-resolved MRA (TRICKS) is not hampered in this respect and may clearly and dynamically demonstrate vascular opacification. For surgical planning, the following structures should be evaluated [73]: the anatomy of the ascending and descending aorta, the distance between the proximal and distal segments of the aortic arch interruption, branch pattern of the great arteries, the presence of a patent ductus arteriosus, the left ventricular outflow tract, and associated structural abnormalities.

In 1975, a first successful end-to-end anastomosis of IAA type B and direct VSD closure was accomplished by Trusler [69,70]. Currently, one-stage approach is preferred in IAA with complete primary repair and closure of the VSD [70]. Aortic arch repair is usually with direct end-to-end anastomosis, and in fewer cases with interposition grafting or patch augmentation [69]. For interposition grafting, one can use the left common carotid artery sacrificing the common carotid artery (“arterial swing down”) [70] or the left subclavian artery [62]. Repair of truncus arteriosus with interrupted aortic arch has been described previously and is generally done with a conduit; an additional patent PDA is ligated and arterial switch with Lecompte maneuver may be performed in patients with additional Taussig–Bing and double outlet right ventricle [70]. However, the results of one-stage repair are not necessarily better than multi-staged repairs [70]. In staged repairs, an initial palliative operation is performed in which the continuity of the aortic arch is reestablished and the distal pulmonary vasculature is protected by pulmonary artery banding. In a later stage, the cardiovascular anatomy is restored completely with pulmonary debanding and VSD closure [68].

The clinical focus is moving from early operative survival to long-term outcomes; more recent patient cohorts have a 5-year survival of 93% [69] with freedom of operation in >60% [69,70,81]. Reinterventions are now dominated by stenosis of the left ventricular outflow tract, whereas improvements in surgical methods minimize the rate of stenosis of the repaired interrupted aortic arch [81]. The incidence of secondary obstruction is amongst others related to potential growth of the aortic annulus and the position of the VSD closure. Several procedures to prevent or redress stenosis have been described, of which the myectomy or myotomy has the worst outcomes due to injuries to the aortic valve [69,82]. Another simple approach is placing the VSD patch on the left site of the conal septum rather than on the right, pushing the septum to the right ventricle and enabling growth of the subaortic area [70,83]. In more severe cases of LVOT obstruction, a Damus–Kaye–Stansel procedure may be performed in which the VSD is baffled to both great arteries and an anastomosis of the end of the pulmonary artery to the side of the ascending aorta is made. The incidence of residual luminal narrowing or restenosis of a previously interrupted aorta is low (up to 100% 5-year freedom of reintervention) [81], and is generally treated with balloon dilatation or reoperation and in only few cases with stent placement or placement of additional conduits [84]. As mentioned before, compression on bronchi and pulmonary vessels may be found both after direct end-to-end anastomosis and graft interpositions [70,81]. Aneurysmal changes at sites of operation are rare [84].

MR imaging in interrupted aortic arch should be tailored to detect hemodynamically significant restenosis of the aortic arch and left outflow tract, and their effects on ventricular function [63]. The aorta and great arteries are preferably imaged with 3D MRA. 3D MRA with isotropic voxels, either with thin slabs or after (additional) interpolation, is advantageous as it provides high quality images that enable multiplanar reformattting in any direction (MPR) and generation of maximum intensity projections (MIP) or volume rendered images. Bronchovascular structures are, again, best imaged with fast spin echo black-blood inversion recovery imaging [73]. In addition, spin echo based pulse sequences are optimal whenever imaging anatomical structures with metallic materials in situ, such as aortic valve prosthesis or stents at the site of previous interruption. Magnetic resonant signal loss around valves and (in-)stent is profound in gradient echo based pulse sequences (including balanced steady-state free precession) due to spin dephasing and should therefore be omitted [85].
Aortopulmonary window

In aortopulmonary window, also aortopulmonary fenestration [86], there is a communication between the ascending aorta and the pulmonary trunk or its branches. In contrast to truncus arteriosus, two separate well-defined semilunar valves are present [87]. Aortopulmonary window is reported in 0.1–0.15% of all congenital cardiac anomalies [87,88]. More than one-third of untreated patients will die in the first year of life due to congestive heart failure and progressive pulmonary vascular hypertension [89].

Aortopulmonary window has historically been classified based on the location of the defect [87,90]:
- **Type I**: proximal defect located directly above the semilunar valves.
- **Type II**: distal defect involving the pulmonary bifurcation at the level of the right pulmonary artery.
- **Type III**: total absence of the aortopulmonary septum resulting in a combined form of type I and II defects.

In the majority the defect is located exclusively between the aorta and main pulmonary trunk [88]. An intermediate type, in which the superior and posterior rims of the aortopulmonary septum are more developed, was more recently added to the classification [91]. This type is most suited for percutaneous transcatheter device closure [87].

The size of the defect varies from a few millimeters to several centimeters [89] and the degree of shunting depends on the size of the defect and the pulmonary vascular resistance. In large left-to-right shunting, volume overload results in enlargement of the left atrium and ventricle [86]. Most commonly, aortopulmonary window is not an isolated defect and morbidity and mortality are related to associated cardiac malformations or pulmonary hypertension. These complicating lesions include interrupted aortic arch (20%), VSD (15%), tetralogy of Fallot (20%), double outlet right ventricle, aortic coarctation and a left vena cava superior [87,88]. The more distal the aortopulmonary communication, the more likely aortic arch malformations are present [92].

The goal of correction is to obliterate the defect between the aorta and pulmonary arteries and to reduce the risk of stenosis in their trajectories. To do so, the defect can be approached through the window itself, through an aortotomy or through a pulmonary artery incision; the window is closed directly (ligation) or with a pericardial or synthetic patch [88]. Preferably, associated anomalies are corrected concomitantly. In small intermediate defects without further cardiovascular anomalies, transcatheter closure is performed [87]. Within 10 years after the initial repair of an aortopulmonary window, reintervention is required in approximately one-third of patients. The transpulmonary approach in particular may cause tapering of the pulmonary trunk or the proximal right or left pulmonary artery [88]. In fewer cases the aortic arch becomes stenotic and the aortic valve stenotic or insufficient.

Neonates with an isolated aortopulmonary window present similar to truncus arteriosus with tachypnea, cyanosis and congestive heart failure [86]. Echocardiography and angiography [89] has been used in the past for diagnosis. In the literature, MRI has been used for initial diagnosis only a few cases which is not to say that it should not be used routinely instead of angiography and as an adjunct to echocardiography. In the medical literature, CMR is more dedicated to patient follow-up. The radiologic signs largely depend on hemodynamics in infants and on residual shunting and stenosis post-operatively [89,92]: location and, size of the window (initial diagnosis), increase in atrial and ventricular size (initial diagnosis), dilatation of the pulmonary artery (initial diagnosis), increased lung vascularity (initial diagnosis), associated structural abnormalities, ventricular function, right ventricular hypertrophy, valve stenosis and insufficiency.

References

PART II Cardiac MR of congenital and acquired pediatric heart disease

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Cardiac MR of congenital and acquired pediatric heart disease

Introduction

Tetralogy of Fallot is the most common type of cyanotic congenital heart disease, accounting for approximately 5.5% of all patients with congenital heart disease [1]. Anterior displacement of the outflow septum is the primary defect, causing a ventricular septal defect, overriding of the aorta, a spectrum of right ventricular outflow tract (RVOT) lesions, ranging from RVOT narrowing to pulmonary atresia, and secondary right ventricular (RV) hypertrophy [2]. In 1955 intra-cardiac repair of tetralogy of Fallot was introduced [3], aiming at closure of the ventricular septal defect and relief of the RVOT obstruction. This surgical approach has been refined and is now performed at the age of 3 to 11 months without prior placement of palliative shunts [4] with excellent long-term survival [5]. Relief of the RVOT obstruction can be achieved by resection of muscular tissue, RVOT patch placement or transannular patch placement. In cases of extreme tetralogy of Fallot with pulmonary atresia, conduit placement is necessary to restore communication between the RV and pulmonary arteries.

Pre-operative assessment of patients with tetralogy of Fallot utilize gadolinium-enhanced 3D magnetic resonance angiography (MRA) to delineate all sources of pulmonary blood supply, especially with tetralogy of Fallot with pulmonary atresia prior to surgical correction [6]. Cardiac magnetic resonance imaging (CMR) can be utilized to delineate the degree of RVOT obstruction as well as the ventricular septal defect.

During follow-up after tetralogy of Fallot repair, residual morphological and functional cardiac abnormalities are common, despite improved pre-, peri-, and post-operative management [7]. Timely detection and monitoring of residual abnormalities is essential and requires accurate and preferably non-invasive imaging methods. CMR is ideally suited to assess abnormalities of morphology and function after correction for tetralogy of Fallot, because this technique is not hampered by anatomical limitations and has the ability to assess 3D anatomy, function, physiology and tissue characterization. CMR can provide quantitative data on the amount of valvular insufficiency and on dimensions and function of the left ventricle (LV) and RV, which has proven to be essential in the follow-up of corrected tetralogy of Fallot patients and in decision-making for pulmonary valve replacement (PVR) [8].

In this chapter we discuss the application of the different CMR techniques to evaluate cardiac morphology and function after correction of tetralogy of Fallot. In addition to a standard CMR examination, in a patient with corrected tetralogy of Fallot, special interest is focused on the morphology of the RVOT, function of the pulmonary valve, morphology of the pulmonary arteries and assessment of the severity of stenoses and finally on the dimensions and systolic as well as diastolic function of the RV and LV.
Dyskinesia of the RVOT occurs in more than 50% of corrected tetralogy of Fallot patients [9]. Akinesia is defined as less than 10% thickening of the RV muscle diameter during systole [9]. Dyskinesia (aneurysm) is defined as outward movement during systole of part of the ventricular wall or its reconstructed outflow tract [9]. After tetralogy of Fallot correction, patients with RVOT aneurysms or akinesia have higher RV end-diastolic and systolic volumes (both indexed for body surface area [BSA]) and lower RV ejection fraction. However, no significant correlation between the incidence of RVOT aneurysm or akinesia and type of RVOT reconstruction (RVOT patch, transannular patch or no patch) has been observed [9]. The finding that RVOT aneurysm formation is not directly related to patch placement suggests that scarring and fibrosis in the RVOT at the site of surgery may contribute to the occurrence of RVOT aneurysm formation after tetralogy of Fallot correction. This hypothesis is supported by the significant correlation between RVOT fibrosis and RVOT dilatation observed after tetralogy of Fallot repair, using delayed enhancement CMR [11].

The recent introduction of percutaneous pulmonary valve replacement (PPVR) to restore pulmonary valve function [12] necessitates detailed evaluation of RVOT morphology after correction.

Right ventricular outflow tract

The outflow tract of the RV is, opposite to the LV, surrounded by muscular tissue. In tetralogy of Fallot patients with a stenosis of the RVOT, relief of the stenosis can be achieved by resection of muscular tissue, although RVOT patch placement is sometimes necessary. If the stenosis is situated at the valvular level, most often a transannular patch is needed, compromising valvular function. RV volumes (e.g., end-diastolic volume) and the amount of pulmonary regurgitation (PR) are significantly increased in patients with an RVOT patch or transannular patch repair as compared to without the use of a patch [9].

The importance of evaluating the RVOT is stressed by the independent association between RVOT aneurysm/dyskinesia or akinesia and RV dilatation as assessed with CMR (Figure 10.1) in corrected tetralogy of Fallot patients [9] and the recent evidence that prolonged QRS duration in corrected tetralogy of Fallot mainly reflects abnormalities of the RVOT rather than the RV itself [10].

CMR evaluation of the RVOT is usually performed by planning a cine in the oblique sagittal plane through the RVOT [9,11] and/or using the short-axis cine images of the RV [9]. Akinesia or dyskinesia of the RVOT occurs in more than 50% of corrected tetralogy of Fallot patients [9]. Akinesia is defined as less than 10% thickening of the RV muscle diameter during systole [9]. Dyskinesia (aneurysm) is defined as outward movement during systole of part of the ventricular wall or its reconstructed outflow tract [9]. After tetralogy of Fallot correction, patients with RVOT aneurysms or akinesia have higher RV end-diastolic and systolic volumes (both indexed for body surface area [BSA]) and lower RV ejection fraction. However, no significant correlation between the incidence of RVOT aneurysm or akinesia and type of RVOT reconstruction (RVOT patch, transannular patch or no patch) has been observed [9]. The finding that RVOT aneurysm formation is not directly related to patch placement suggests that scarring and fibrosis in the RVOT at the site of surgery may contribute to the occurrence of RVOT aneurysm formation after tetralogy of Fallot correction. This hypothesis is supported by the significant correlation between RVOT fibrosis and RVOT dilatation observed after tetralogy of Fallot repair, using delayed enhancement CMR [11].

The recent introduction of percutaneous pulmonary valve replacement (PPVR) to restore pulmonary valve function [12] necessitates detailed evaluation of RVOT morphology after correction.

Figure 10.1 End-diastolic (a) and end-systolic (b) gradient echo MR images at midventricular level in a tetralogy of Fallot patient showing dilatation of the right ventricle (RV) with akinesia of the RV free wall; no motion or wall thickening was observed during the cardiac cycle. Tetralogy of Fallot correction in the past was followed by pulmonary valve replacement and tricuspid valve reconstruction. In this patient, there was no pulmonary regurgitation or tricuspid regurgitation. RV end-diastolic volume was 284 ml, RV ejection fraction was 25%. Left ventricle (LV) end-diastolic volume was 145 ml, ejection fraction was 49%, which is subnormal and likely caused by interventricular dependence of ventricular function. Note in these axial images the chest deformity with ventral out-pouching of the left hemithorax due to the RV dilatation.
of tetralogy of Fallot [13]. The selection of patients for PPVR depends on the presence of an appropriate location for valve-containing stent implantation within the RVOT [13]. Contrast enhanced MRA is well-suited for 3D reconstruction of the RVOT lumen (Figure 10.2b) [13]. Based on the reconstructions of 83 consecutive patients with RVOT dysfunction, five common types of RVOT morphologies could be identified covering 98% of RVOT shapes. Type I, a pyramidal shape, is most commonly observed, especially after tetralogy of Fallot correction, and has been associated with transannular patch repair and higher pulmonary regurgitant fraction. This type I RVOT morphology is unsuitable for percutaneous valve replacement using the currently available device, because of lack of a suitable stent fixation point and therefore risk of stent migration [13]. Current potential tetralogy of Fallot candidates for PPVR are those with a history of surgical pulmonary valve replacement (PVR) but without transannular patch repair, who suffer from degenerative disease of the bioprosthesis [15].

**Pulmonary valve function**

Pulmonary valve function is one of the key issues during follow-up after tetralogy of Fallot repair and influences clinical outcome [7]. Longstanding volume or pressure overload of the RV has been associated with biventricular systolic as well as diastolic dysfunction at rest [16,17], and at exercise [18], ventricular arrhythmias [7], impaired exercise capacity [19], and sudden death [7]. Quantification of PR volume is limited with conventional imaging techniques, such as echocardiography and nuclear imaging. CMR can be used to quantify PR by two different approaches. PR can be assessed by the difference between LV and RV stroke volume, obtained by cine CMR. Net output of the LV and RV should be equal and in patients with pulmonary valve incompetence, the regurgitant volume can be calculated by subtracting LV stroke volume from RV stroke volume [20]. In the presence of tricuspid, mitral and/or aortic valve regurgitation or the presence of an intracardiac shunt such as a (residual) ventricular septal defect, this approach cannot be used. Therefore, direct measurement of PR using phase-contrast CMR in the pulmonary artery is the preferred technique to quantify PR and this approach has proven accurate (Figure 10.3) [20]. To assess PR, a multiphase phase-contrast MRI is planned perpendicular to the main pulmonary artery immediately distal to the level of the pulmonary valve. The optimal posi-

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**Figure 10.2** (a) Black-blood turbo spin-echo MR image of a tetralogy of Fallot patient showing the proximal pulmonary arteries in axial view. There is a pulmonary artery (RPA) and normally sized proximal left pulmonary artery (LPA). Note the dilated ascending aorta (*) that has a 4.2 cm diameter size, and note the right descending thoracic aorta (arrow). In patients with tetralogy of Fallot, the ascending aorta is prone to dilatation [14]. (b) Gadolinium-chelate enhanced MR angiography 3D reconstruction image showing the pulmonary arteries in oblique craniocoronal view. In the past, pulmonary trunk reconstruction was performed by (aorta) homograft (H) interposition between the right ventricle and pulmonary confluence. There is a normal size of the pulmonary trunk homograft (26 mm). Note the small sized right pulmonary artery (RPA) that measures 9 mm and the normally sized left pulmonary artery (LPA) that measures 21 mm. Right ventricle (RV), right atrium (RA).
Figure 10.3 MRI gradient echo white-blood images (a, b) in 9-year-old male with corrected tetralogy of Fallot. Patient has pulmonary regurgitation, a normal right pulmonary artery (RPA) (a) and proximal left pulmonary artery (LPA) stenosis (b). Thirty-six-phase flow mapping graph shows flow curves for the pulmonary trunk (PT), and separately for the RPA and LPA (c). Separate measures were made for each artery, collected in this single graph. Flow curves represent forward flow above the x-axis and regurgitant flow below. Pulmonary regurgitation measured over the PT was 43%. Note the large contribution to the total pulmonary flow for the RPA whereas the contribution of the LPA is only small in this patient with proximal LPA stenosis. Turbulence at the LPA stenosis resulted in impaired image quality that caused the summation of the LPA and RPA to be slightly less than the total PT flow.
tion of the scanning plane is based on axial, sagittal, and coronal scout images, and sometimes additional oblique scout images. Flow volume and velocity across the pulmonary valve can be calculated after semiautomatic drawing of contours around the pulmonary valve throughout the cardiac cycle.

**Main and branch pulmonary arteries**

Evaluation of vascular morphology after tetralogy of Fallot correction focuses mainly on evaluation of the pulmonary arteries. During follow-up, abnormalities of the main and peripheral pulmonary arteries are common and stenosis can have a substantial impact on lung perfusion and exercise capacity [21]. Furthermore, the amount of PR is influenced by geometric differences of branch pulmonary arteries, unequal pulmonary vasculature and pulmonary perfusion abnormalities [22]. Black-blood imaging is useful to evaluate the pulmonary arteries (Figure 10.2a), conduits and the aorta, especially when turbulence or metal-related artifacts are present [23] and can be used to visualize stenoses of the pulmonary arteries [24,25]. Gadolinium-enhanced MRA is an attractive technique to visualize the pulmonary arteries in an angiographic format that provides 3D reconstructions (Figure 10.2b) [6]. This is preferably performed with sedation in patients under 7 years old and with breath-holding in older individuals [26]. MRA has a high sensitivity (94%) and accuracy (97%) for detection of stenosis or hypoplasia of the pulmonary arteries compared with conventional angiography [27]. Furthermore, additional aortopulmonary collateral arteries can be diagnosed with MRA, not observed using conventional angiography [27].

Recently, the individual contributions of the left pulmonary artery (LPA) and right pulmonary artery (RPA) to PR in corrected tetralogy of Fallot patients have been investigated by phase-contrast MRI by planning imaging planes perpendicular to the LPA or RPA [22]. The average contribution of the LPA to the total regurgitant volume was 54%, whereas its average contribution to total forward flow volume was 44%. These findings suggest that in patients in whom there is a significant unilateral contribution to total PR, localized intervention may be used to reduce regurgitation [22]. Other groups have validated branch pulmonary artery regurgitant fractions as well [23].

Assessment of the functional significance of branch pulmonary artery stenosis is important because asymmetry in lung perfusion frequently occur in tetralogy of Fallot patients due to peripheral pulmonary stenoses [29]. Asymmetry of lung perfusion influences exercise capacity and is a predictor of outcome [29]. An example of flow measurements through the main pulmonary artery and the individual contributions of the LPA and RPA to pulmonary perfusion in a patient with proximal LPA stenosis is shown in Figure 10.3.

Radionuclide scanning has been used in the past to determine the perfusion distribution throughout the lungs [30] but this causes a significant radiation burden. Phase-contrast MRI measurement of the LPA and RPA flow are now standard of care to reveal differences in left and right lung perfusion in children with suspected branch pulmonary artery stenosis [30]. For adequate measurement of lung perfusion for each lung, the total forward flow volume for each lung should be measured in the presence of pulmonary valve incompetence to correct for the individual contribution to PR of the LPA and RPA [30].

**Systolic function**

Regular evaluation of biventricular systolic function after tetralogy of Fallot repair is essential during follow-up, because RV and LV dysfunction are common. RV as well as LV systolic dysfunction predict major adverse clinical outcomes late after tetralogy of Fallot repair [31]. Volume overload due to pulmonary valve regurgitation is common after tetralogy of Fallot repair, especially when a transannular patch is used, causing RV dilatation and decreased ejection fraction. Besides increased RV volumes and decreased RV stroke volume, decreased LV ejection fraction has also been shown to be an independent risk factor for major adverse clinical outcomes [31].

To evaluate systolic RV function, echocardiography is not adequately suited because of its lack of reliable quantification of RV end-diastolic volume and end-systolic volume [32]. Therefore, CMR is regarded as the gold standard for volume
quantification and evaluation of ventricular stroke volume and ejection fraction in children with congenital heart defects [33]. Cine images obtained in the transverse plane, along the short-axis of the LV, or using a modified RV short axis orientation [34] are most frequently used for volumetric quantification with low interobserver and intraobserver variability in children [16]. Using post-processing programs and by delineating the endocardial ventricular border end-diastolic and end-systolic volume, stroke volume and ejection fraction can be calculated (Figure 10.4). In the presence of pulmo-

Figure 10.4 Gradient echo images with endocardial left ventricular (LV) and right ventricular (RV) contours drawn at end-diastolic (a) and end-systolic phase (c) in 15-year-old female patient with tetralogy of Fallot after repair. Contour drawing is performed at all slice-levels covering the ventricles (usually 8–10 slices). Then, the end-diastolic (b) and end-systolic volumes (d) and ejection fraction can be calculated. RV end-diastolic volume was 226 ml, ejection fraction was 56%. RV ejection fraction corrected for pulmonary regurgitation (47%) was 32% (see text). LV end-diastolic volume was 128 ml, LV ejection fraction was 57%. This was done in the transverse plane although it is more common to do this in short axis.
nary valve regurgitation, stroke volume and ejection fraction of the RV is largely determined by the volume of regurgitant flow [35]. To correct the RV ejection fraction for the amount of regurgitation after tetralogy of Fallot repair, the “net” flow across the pulmonary valve, as obtained by flow velocity measurements, can be divided by the RV end-diastolic volume [35]. This “corrected” RV ejection fraction provides a tool to assess the improvement of RV ejection fraction after PVR to restore pulmonary valve competence in corrected tetralogy of Fallot patients [35] although the use of the corrected RV ejection fraction and its physiologic/functional meaning is controversial.

Timing PVR is one of the key questions during the follow-up. Recent studies have shown that RV end-diastolic and systolic volume rather than the amount of regurgitant flow through the pulmonary valve seems to be the most critical factor in the decision of replacing the pulmonary valve [8,36–38]. After PVR, remodeling of the RV occurs promptly in most patients (Figure 10.5), evidenced by decrease in RV end-diastolic and end-systolic volume [35,36] and decrease of RV mass [36]. Reduction of RV volumes after PVR has been observed independently of surgical reduction of the RVOT by resection of aneurysmal tissue [8]. In children with repaired tetralogy of Fallot, normalization of RV volume and mass has been observed within 6 months after PVR, if performed in patients with RV end-diastolic volume index exceeding 150 ml/m² body surface area. However, no normalization occurred if PVR had been performed in patients with RV end-diastolic index >200 ml/m² [36]. In adults after Fallot repair, normalization of RV volumes can be achieved after PVR in patients with RV end-diastolic volume indexed for BSA of <160 ml/m² or RV end-systolic volume of <82 ml/m² [8]. Other authors have reported cut-off values for PVR of

![Figure 10.5](image-url)
RV end-diastolic volume indexed for BSA of <170 ml/m² or RV end-systolic volume of <85 ml/m² [37]. The general similarities in cut-off values between both adult studies underline the robustness of the CMR technique to aid in planning PVR [8,35,36].

Besides RV dysfunction, LV dysfunction at rest and at exercise can be observed after tetralogy of Fallot repair (Figure 10.1) [18,39–41]. Evaluation of LV function should be part of any imaging study in this patient population [17]. Global LV function can be evaluated using cine techniques and by planning a stack of slices from the apex to the base of the heart in the short axis direction. From a practical point of view, both LV and RV function can also be derived qualitatively from a single data set of axial slices. Cine-loops in axial orientation are of additional value in evaluating morphology and function, for example, of the pulmonary arteries.

The observed correlation between the amount of PR and dilatation of RV and LV dysfunction [17,41] stresses the detrimental role of PR on biventricular function. Altered systolic movement of the intraventricular septum due to chronic volume overload of the RV, patch placement to close the VSD and septal fibrosis may be responsible for adverse ventricular-ventricular interaction as evidenced by the correlation between PR and LV dysfunction [17] and the correlation between increased RV volumes and LV dysfunction [9]. Other causes for LV dysfunction have been proposed, such as episodes of acute hypoxia and operative procedures [17]. This is supported by the findings of LV scar tissue at the sites of surgical manipulation (VSD), although this found in a minority of patients. Additionally, in 24% of the corrected tetralogy of Fallot patients, delayed enhancement was observed in the RV trabeculations including the moderator band, which may reflect ischemic insults [42].

Diastolic function
Diastolic ventricular filling is a complicated process that depends on atrial- and ventricular pressure, myocardial compliance, myocardial relaxation, atrioventricular valve function and other factors [43]. The importance of evaluating RV diastolic function in tetralogy of Fallot patients is stressed by the observation that a restricted diastolic filling pattern of the RV predicts slow post-operative recovery [44] but superior exercise performance [45]. Furthermore, diastolic dysfunction may precede later systolic dysfunction during follow-up [43]. Diastolic function of a ventricle can be assessed by the flow characteristics through the atrio-ventricular valve as obtained by echo-Doppler echocardiography [46] or phase-contrast MRI [47]. A “restrictive” filling pattern is also evidenced by antegrade end-diastolic flow in the main pulmonary artery by CMR phase contrast mapping.

In the presence of PR, filling of the RV occurs through the tricuspid valve as well as through the incompetent pulmonary valve and therefore RV filling can not be assessed adequately by evaluation of the tricuspid valve flow pattern alone. Phase-contrast CMR of both the tricuspid valve and the pulmonary valve (Figure 10.6) allow the construction of RV time–volume curves in the presence of PR [16]. This RV time–volume curve can be used to demonstrate impaired relaxation and restriction to filling as signs of abnormal RV diastolic function [16]. More recently, this approach has been used during dobutamine stress [48]. Using stress CMR imaging, diastolic dysfunction was revealed in patients after tetralogy of Fallot correction with normal diastolic RV filling at rest [48].

Conclusion
CMR is currently widely used for follow-up of patients after tetralogy of Fallot correction. Quantification of PR and assessment of biventricular morphology and function, as well as assessment of pulmonary artery morphology are unique features of the different CMR techniques. Application of CMR during follow-up can identify risks factors for major adverse clinical events [7,31]. Furthermore, CMR to assess RV dimensions proved to be important in the decision-making whether or not to replace the pulmonary valve after tetralogy of Fallot correction and provides cut-off values for PVR in children [36] and adults [8,37]. Finally, CMR is essential in the detailed evaluation of RVOT morphology, which is essential to select patients suitable for PPVR [13].
Cardiac MR of congenital and acquired pediatric heart disease

Figure 10.6 9-year old male with corrected tetralogy of Fallot. The patient has pulmonary regurgitation (PR). (a) Right ventricular and left ventricular inflow curves measured across the mitral valve (MV) and tricuspid valve (TV), respectively, throughout the cardiac cycle. Note the normal E/A ratio of early (E) to late atrial kick contribution to filling (A) that is larger than 1 for flow across the MV. Across the TV, an E/A ratio <1 is observed, suggesting impaired relaxation. However, this patient has PR that renders the TV inflow curve unreliable. (b) To correct RV inflow for PR, the PR volume should be added to the TV volume. After correction, it can be observed that the E/A ratio is larger than 1. No evidence of impaired relaxation or restriction found after correction. Same patient as in Figure 10.3.

References

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Aortic arch anomalies

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CHAPTER 11

Introduction – imaging of arch anomalies

History of imaging arch anomalies

Barium esophagography

Aortic arch anomalies have been recognized pathologically since the early 18th century. However clinical confirmation of a specific lesion during life did not occur until the early 20th century when Kommerell first discovered and identified what has turned out to be a very rare vascular ring by barium esophagography. Although plain chest roentgenograms often reveal arch sidedness and, in the face of appropriate symptoms, may suggest a vascular ring, they are insufficient for definitive diagnoses of arch anomalies.

Angiography

From the 1960s to the 1980s, angiography became the diagnostic tool of choice for these anomalies, especially vascular rings; although many radiologists still contended that they could make all necessary diagnostic decisions with barium esophagography. Although plain chest roentgenograms often reveal arch sidedness and, in the face of appropriate symptoms, may suggest a vascular ring, they are insufficient for definitive diagnoses of arch anomalies.

Echocardiography

With the advent of transthoracic two-dimensional echocardiography in the late 1970s to early 1980s [1–3], the focus shifted to this modality for identification of arch branching sequences and, indirectly, arch sidedness and arch abnormalities. The limitations of this tool center on difficulty in imaging the air-filled trachea, which is central to the identification of aortic arch sidedness, in general, and vascular rings, in particular. This problem is obviated in the fetus where the trachea and lungs do not scatter ultrasound waves and where the ductus arteriosus, often a component of a vascular ring, is always patent.

Computed tomography (CT)

The use of spiral CT scans has been described in the characterization of arch anomalies and vascular rings since 1995 [4]. The introduction of the multiphase CT in 1998 greatly improved the speed and resolution of CT scans [5] and prompted some to advocate their routine use in the evaluation of arch anomalies [6]. However, despite progress in reducing radiation, these exams still expose a particularly vulnerable population to doses of ionizing radiation that may significantly increase their long-term risk of malignancy.

Magnetic resonance imaging (MRI)

Beginning in the mid-1980s magnetic resonance imaging (MRI) entered the scene. Initially the limited availability of MRI and protracted examinations left echocardiography and computed tomography as the mainstays of arch anomaly diagnoses. However, in the mid to late-1990s, MRI supplanted echocardiography as the imaging tool of choice for arch anomalies with its ability to view arch sidedness and all branches directly while also...
visualizing airway compression and yet avoiding ionizing radiation [7–9].

**MRI – specific sequences**

Most arch anomalies may be diagnosed quite easily by cardiac MRI using cardiac gated, static steady-state free procession (SSFP) contiguous axial slices. However, it can occasionally be challenging to definitively define a diverticulum of Kommerell on axial imaging, especially in infants. Therefore, it is often useful to perform coronal imaging to better define a diverticulum. At our institution, we generally perform off-axis coronal images, in which the initial SSFP axial stack is used to better align with the plane of the descending aorta and course of the aberrant subclavian artery utilizing multiplanar reconstruction.

If a vascular ring is diagnosed, high-resolution imaging of the trachea is desirable to quantify the current degree of tracheal narrowing at the level of the ring. Cardiac gated, high resolution double inversion dark blood sequences with a 2–4 mm slice thickness are useful for this purpose. Orthogonal views aligned with the trachea using multiplanar reconstructions from the axial stack are then performed. Similar images can occasionally be useful for confirming the presence or absence of a diverticulum or aortic dimple, since they provide excellent contrast between the vessel lumen and vascular wall.

Cine imaging is rarely required in the diagnosis of an arch anomaly. It can be useful for visualizing a connection from the pulmonary artery to an isolated subclavian when a ductus arteriosus is still patent. It is also useful for visualizing turbulence if arch obstruction or narrowing is suspected from static imaging.

Similarly, phase contrast magnetic resonance (PC-MR) velocity mapping is generally not required for the diagnosis of most arch anomalies. However, there are two instances in which it can be quite helpful as ancillary data. In the setting of an isolated subclavian artery (i.e., subclavian artery arising exclusively from the ductus arteriosus) with ductal closure, in which the subclavian artery is supplied by retrograde flow from the vertebral artery, it is helpful to verify the direction of flow in the vertebral artery since this is the substrate for subclavian steal syndrome. Using an axial slice position at the level of the vertebral artery, just superior to its insertion into the subclavian artery, a through-plane PC-MR sequence can verify that the vertebral flow on the side of the isolated subclavian is in the opposite direction of the carotid arteries and opposite the contralateral vertebral artery. The other situation where PC-MR can be helpful is in quantifying the flow in both distal aortic arches in double aortic arch. Because of variation in arch caliber and obliquity of the two arches relative to each other, it may be difficult to determine which is the smaller of the two and therefore the one which should be surgically divided. Quantitative flow measurements in the two distal arches permits determination of which is carrying the lesser blood flow, thus the one to be divided.

Special mention should be given to the use of cardiac MRI to diagnose arch anomalies in the neonate. In the past, the long scan times of MRI and the need for general anesthesia has justifiably made clinicians reluctant to utilize MRI for arch diagnosis in the newborn period. However, recent improvements in acquisition times and image quality have made both of these concerns obsolete. In the vast majority of cases, the imaging required to accurately diagnose an arch anomaly can be performed in 5–10 minutes in the hands of an experienced cardiac MRI clinician. A 40–45 slice SSFP axial stack (3 mm slices, 10–20% overlap) (2–3 minutes), a 20-slice off-axis SSFP coronal stack (1–2 minutes), and two double inversion dark blood sequences of the airway (1–2 minutes) should be all that is required to diagnose the vast majority of arch anomalies in infants. The short scan time eliminates the necessity of sedation in the majority of otherwise healthy infants, who can simply be fed and then swaddled for the duration of the scan. With rare exception, this technique has been sufficient in our experience when the only clinical question is the aortic arch anatomy, having ruled out other cardiac disease by echocardiography. While cardiac CT is certainly even faster and arguably less labor-intensive, most would agree that it is desirable to avoid ionizing radiation when possible. However, when one compares “apples to apples,” CT is not that much faster than MRI; a contrast enhanced magnetic resonance angiogram
(MRA) can take as little as 6 seconds whereas a CT may take just a few seconds less. Both can make definitive 3-dimensional images of the aortic arch anomaly.

**Definitions**

**Arch sidedness**

Aortic arch sidedness is determined by the bronchus over which the aorta passes. This is independent of the sidedness of the ascending aorta. There is a rule, designed primarily for use with echocardiography and angiography, that the first arch vessel gives rise to a carotid artery opposite the side of the arch, that is, first arch vessel giving rise to a left carotid artery predicts a right aortic arch. While it is true that the branching pattern of the brachiocephalic arteries usually predicts arch sidedness, it must be remembered that this is an indirect method that has several exceptions. Two rare exceptions are that in the case of a retroesophageal innominate artery or an isolated innominate artery, the first arch vessel is ipsilateral to the arch. In double aortic arch regardless of which carotid artery is thought to be “first,” there is an ipsilateral arch in addition to a contralateral arch. But the most common and therefore important exception to the above rule is that in cases where the first two arch vessels are both carotid arteries, it may be difficult to decide which is first. However, with MRI, since vessels and major airways are easily visualized, rules for inference of arch sidedness are irrelevant. One rule that does hold true always is that retroesophageal vessels and isolated vessels (i.e., subclavian and/or carotid arteries arising exclusively from a ductus arteriosus) are always opposite the side of the aortic arch. This rule is not generally of utility with echocardiography.

**Anatomical classification**

Arch anomalies can be categorized as to arch sidedness – right, left, double; branching abnormalities – including retroesophageal subclavian or rarely innominate artery, isolated subclavian or rarely innominate artery; persistent embryonic fifth aortic arch; interrupted aortic arch; anomalous origin of a branch pulmonary artery – from the ascending aorta or from the contralateral branch (so-called pulmonary artery sling).

**Clinical classification**

Patients with aortic arch anomalies fall into several groups clinically: vascular rings, non-ring airway compression abnormalities, non-compressive branching abnormalities, ductal dependent abnormalities, abnormalities of branch pulmonary artery origin.

**Vascular rings**

Vascular rings are arch anomalies in which both the trachea and esophagus are surrounded by vascular tissue, but some portions of the ring may be atretic. This last point raises the question of how the diagnosis of vascular ring can be made by present-day imaging modalities, all of which rely upon flowing blood to permit visualization. The answer is that the presence of any of three features, all of which begin with the letter “D,” always indicates the existence of a vascular ring when they occur opposite the side of the aortic arch: an aortic diverticulum, a dimple, or the descending aorta. A diverticulum is a large outpouching from the descending aorta from which a smaller vessel – the distal subclavian artery arises. A dimple is a small, blindly ending outpouching from the descending aorta. The descending aorta opposite the side of the arch means that the aortic arch crosses behind the esophagus in the upper thorax where it abruptly turns inferiorly.

**Double aortic arch**

The most common vascular ring is the group of anomalies in which there are both right and left aortic arches. While most cases have patency of both arches, some have atresia of the left arch. Whether patent or atretic all double aortic arches are examples of vascular rings as defined above. While all are rings, not all cause significant tracheal compression.

When considering surgical division of a vascular ring, it must be kept in mind that a ligamentum arteriosum (or rarely a patent ductus arteriosus) may be present in addition to the arches (particularly when there is no major intracardiac anomaly, such as a large ventricular septal defect). Thus division of an aortic arch could leave a vascular ring completed by a ligamentum (or ductus).
With equal arches

While not the most common form of double aortic arch, this type almost always has significant tracheal compression. Symptoms consisting of stridor, cough, or apnea often occur in infancy. On axial MRI the four arch vessels arise separately and symmetrically from the two arches. The left arch passes anterior and to the left of the trachea, the right arch passes to the right and posterior. Coronal imaging shows the arches on either side of the trachea, and a more posterior slice shows the distal limbs of the two arches join to form the descending aorta. (Figure 11.1).

With dominant right arch

Both patent

Most double aortic arches have a dominant right arch (Figure 11.2). This is often best appreciated in a coronal view, either at the level of the trachea where both arches are seen in cross section, or more posteriorly, where the two arches are seen to join and form the descending aorta. Because there is variation in the location of the smallest part of the left arch, three-dimensional surface displays make it easier to quickly locate this. Alternatively one can measure flow in the two distal arches using phase-encoded velocity mapping to determine which arch carries less blood and therefore is the better one to divide.

Atretic left arch

The extreme form of double aortic arch with a dominant right arch is double aortic arch with atretic left arch (Figure 11.3). There are two variations: atresia between the left carotid and left subclavian arteries or atresia distal to the left subclavian artery. Depending on the sidedness of the descending aorta, the appearance of the former may be identical to that seen in the vascular ring right aortic arch with diverticulum of Kommerell or the anomalous subclavian artery form of right aortic arch with left descending aorta; the latter may look

![Figure 11.1 Double aortic arch with equal sized arches.](image)

(a) Axial imaging demonstrating symmetrical origins of right (RCA) and left (LCA) carotid and right (RSCA) and left (LSCA) subclavian arteries. Note dilated esophagus (E). LInV: left innominate vein, RJV: right jugular vein, T: trachea. (b) More inferior axial image demonstrating two relatively equal proximal left (LAoA) and right (RAoA) arches surrounding narrowed T. SVC: superior vena cava. (c) More inferior axial image showing two nearly equal distal LAoA and RAoA. AAO: ascending aorta, Az: azygos vein. (d) Farther inferior axial image showing fusion of RAoA and LAoA posteriorly. (e) to form the descending aorta (DAo). MPA: main pulmonary artery. (f) Coronal image demonstrating relatively equal sized RAoA and LAoA on either side of the T. LPA: left pulmonary artery. (g) Posterior-cranial view of 3-dimensional surface display showing equal sized right arch (closed arrowheads) and left arch (open arrowheads) forming a complete ring of vessels through which the trachea and esophagus (not shown) pass.

Note: Image orientation arrows in all the figures use the following abbreviations: A: anterior; I: Inferior; L: left; P: posterior; R: right; S: superior.
Figure 11.1 Continued
similar to the vascular ring right aortic arch with retroesophageal left ductus (ligamentum), but the location of the dimple on the descending aorta may help to distinguish the two (compare Figures 11.3 and 11.8). All of these are described below. Since these may be indistinguishable by imaging, the surgeon must always be prepared to divide both an atretic arch and a ligamentum arteriosum when these patterns are present.

**With dominant left arch**
Double aortic arch with dominant left arch (Figure 11.4) is relatively uncommon. However it is one of the rare, though important, exceptions to the general rule that vascular rings are always divided through a left thoracotomy. Because the smallest portion of the ring is usually the part posterior to the trachea and esophagus, it may be difficult to measure the caliber by echocardiography. This is further evidence for the importance of MRI in the diagnosis and assessment of vascular rings.

**Right aortic arch**
There are three types of vascular ring with a right aortic arch. These are typically not associated with intracardiac abnormalities but may be associated with the DiGeorge syndrome and chromosome 22q11.2 microdeletion.

**With diverticulum of Kommerell**
This is the second most common vascular ring after double aortic arch. While comprising only 5% of right arches, it accounts for nearly 20% of symptomatic vascular rings. The branching pattern has the left carotid artery as the first branch, followed by right carotid, right subclavian and then the left subclavian artery arising from the diverticulum of Kommerell. The ring is completed by a ligamentum arteriosum. Since the ligamentum has no lumen and therefore cannot be visualized with current imaging techniques, one might wonder how this can be distinguished from the non-ring right aortic arch with retroesophageal left subclavian artery – the sequence of branches is identical. The presence of a diverticulum, that is, a larger caliber vessel rapidly tapering to a smaller (normal) vessel, viz. subclavian artery, always signifies the presence of a ring (Figure 11.5). The simple aberrant subclavian artery has a uniform caliber throughout its thoracic course (Figure 11.6). During fetal life the ductus arteriosus “feeds” the descending aorta and therefore carries more than

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**Figure 11.2** Double aortic arch with dominant right arch. (a) Coronal image at level of the trachea (T) showing larger RAoA in cross-section. (Compare with Figure 11.1f.) RPA: right pulmonary artery. (b) More posterior coronal image showing larger RAoA joining LAoA to form DAo. DAo: descending aorta; LAoA: left aortic arch; LCA: left carotid artery; LPA: left pulmonary artery; LSCA: left subclavian artery; RAoA: right aortic arch; RPA: right pulmonary artery; T: trachea.
Figure 11.3 Double aortic arch with atretic left arch. (a) Left lateral view of 3-D volume rendering from static gadolinium acquisition shows posterior tethering of LSCA opposite dimple (open arrowhead) of distal left arch. (b) Posterior view of 3-D volume rendering shows right aortic arch (closed arrowhead) with left-facing dimple (open arrowhead) opposite the side of the arch indicating a vascular ring. This pattern is indicative of an atretic segment between LSCA and distal left arch. (Compare with Figure 11.8 below.) AAo: Ascending aorta; DAo: descending aorta; LCA: left carotid artery; LSCA: left subclavian artery; RCA: right carotid artery; RSCA: right subclavian artery.

Figure 11.4 Double aortic arch with dominant left arch. Coronal view showing larger posterior LAOA than RAoA. Compare with Figure 11.2b. DAo: descending aorta; LAOA: left aortic arch; LSCA: left subclavian artery; LVA: left vertebral artery; RVA: right vertebral artery.

50% of the combined output of the two ventricles; whereas the subclavian artery distal to the ductal insertion carries a small fraction of that, viz. flow to the left arm. After birth when the ductus closes, the aberrant subclavian artery has a large caliber proximal portion – the diverticulum of Kommerell – and a smaller distal portion. Although the flow in each portion is the same, the difference in caliber persists.

With left descending aorta (right circumflex aortic arch)
This is the third most common vascular ring. While it is true that nearly all cases of situs solitus (normal arrangement) of the viscera are associated with a left-sided descending aorta irrespective of arch sidedness, this refers to the descending aorta at and below the level of the diaphragm. Typically with right aortic arches, after crossing over the right mainstem bronchus the aorta gradually moves toward the left, reaching a leftward position near the diaphragm. However in the right arch-left descending cases, after crossing over the right mainstem bronchus, the aorta courses immediately to the left, passing behind the trachea and...
esophagus – so-called circumflex aortic arch – and then turns abruptly inferiorly so that the entire descending thoracic aorta is to the left of midline (Figure 11.7). Once again as in diverticulum of Kommerell the ring is completed by a ligamentum arteriosum, which is not visualized by current imaging techniques but is inferred from the presence of a descending aorta opposite the side of the arch. Two branching patterns are known: with left innominate artery and with aberrant left subclavian artery. Note that in this latter case the aberrant subclavian is not retroesophageal since it arises from the descending aorta after the arch itself has passed retroesophageally.
CHAPTER 11 Aortic arch anomalies

Left aortic arch
There are only two vascular rings associated with left aortic arch: rare and rarer – left arch with right descending aorta and left arch with right diverticulum of Kommerell. These two with the uncommon, but not as rare double aortic arch with dominant left arch, are the exceptions to the general rule that all vascular rings can be divided through a left thoracotomy.

With right descending (left circumflex aortic arch)
This is the mirror image of the more common right aortic arch with left descending aorta. This, like its counterpart, can also be called circumflex aortic arch. Because the orientation of the retroesophageal arch is upward to the left (or downward to the right), the impression on the barium-filled esophagus can mimic the more common right arch with retroesophageal diverticulum and lead to a mistaken diagnosis [10] when relying on that modality. We have only seen the aberrant subclavian variety of this anomaly, but a right innominate artery is theoretically possible.

Diverticulum of Kommerell
This too is the mirror image of the more common right aortic arch with left diverticulum giving rise to aberrant subclavian artery and ductus (ligamentum) (Figure 11.9). What is remarkable is that although this anomaly is quite rare compared with the right arch version, this is actually the anomaly first reported by Kommerell [11] – the first example of a vascular ring diagnosed (by barium esophagography) during life. However, the eponym has been applied to the right arch version as well.

Non-ring anomalies
While vascular rings are generally what come to mind when vascular causes of tracheo-bronchial compression are discussed, there are other causes that must be considered.

With tracheo-bronchial compression
Ascending and descending aorta in same sagittal plane
Normally the ascending aorta is right-sided and descending left-sided; however, in some cases with
Figure 11.7 Right aortic arch (RAoA) with left descending aorta (DAo). (a) Axial view showing RAoA with first vessel, left innominate artery (LInA) and retroesophageal circumflex arch (CAoA). (b) More inferior axial image showing right-sided AAo and distal CAoA with ductus dimple (open arrowheads). (c) More inferior axial image showing left-sided DAo. (d) Coronal view demonstrating normal rightward AAo. (e) More posterior coronal image showing RAoA. (f) More posterior coronal image showing retroesophageal CAoA and ductus dimple (open arrowheads). (g) More posterior coronal image showing left-sided DAo. (h) Posterior view of 3-D volume rendering of a static gadolinium acquisition demonstrating the abrupt angulation (open arrow) as the aorta goes from its retroesophageal circumflex portion to DAo. The ductus dimple (open arrowhead) is connected by the ligamentum arteriosum (not visualized) to the pulmonary artery seen in (f), completing the vascular ring. AAo: ascending aorta; LV: left ventricle; MPA: main pulmonary artery; RA: right artery.
Figure 11.7  Continued
Figure 11.8 Right aortic arch (RAoA) with retroesophageal left ductus/ligamentum. (a) Coronal image demonstrating typical mirror image RAoA branching pattern (see Figure 11.12 below): LinA, followed by right carotid artery (RCA), … (b) and right subclavian artery (RSCA). (c) More posterior coronal image shows right descending aorta (DAo). Note the subtle ductus dimple (closed arrowheads) on the left side of the DAo. The ligamentum arteriosum (not seen) completes a vascular ring between this dimple and the left pulmonary artery (LPA). There is an incidental left superior vena cava (LSVC) to coronary sinus (CoS). (d) Left lateral, (e) left posterior oblique, and (f) cranial views of 3-D volume rendering showing typical mirror image right aortic arch branching pattern except for the subtle dimple (open arrow) toward the left indicating a left ligamentum and therefore a vascular ring. (Compare with Figures 11.3a and 11.3b above with similar branching pattern but more superior dimple pointing toward LSCA.) LCA: left carotid artery; LinA: left innominate artery; LJv: left jugular vein; MPA: main pulmonary artery; RCA: right carotid artery; RPA: right pulmonary artery.
small antero-posterior chest diameter, the ascending aorta is displaced to the left so that the ascending and descending aorta are in the same sagittal plan. With the small antero-posterior chest diameter the left mainstem bronchus may be compressed between the ascending aorta and right pulmonary artery anteriorly and the descending aorta posteriorly (Figure 11.10).

Innominate artery compression of trachea
The innominate artery normally crosses the trachea. In cases where there is a narrow antero-posterior chest diameter or when there is tracheomalacia, the innominate artery may indent the trachea at the point where it crosses (Figure 11.11). Some investigators have postulated that this is a result of an abnormal origin of the innominate artery, but 3-dimensional reconstructions from MRI have shown no abnormality of the aorta and its arch vessels.

Ascending aorta compression of trachea
Normally the aorta passes from right to left anterior and to the left of the trachea. There is plenty of room in the mediastinum for both the aorta and trachea to exist without compression of either. In cases where the heart is displaced, especially with dextrocardia, the aorta may wrap around the trachea and compress it.

Without tracheo-bronchial compression
While all arch anomalies are interesting academically, and some have implications for various associations such as the DiGeorge syndrome, there are some clinically significant arch anomalies as well as clinical situations where arch sidedness or anatomy become relevant. The following are examples of non-compressive arch anomalies:

Mirror image right aortic arch
This is an arch anomaly with no physiological consequences except the small possibility of right mainstem bronchial compression as described in “Ascending and descending aorta in same sagittal plane” above. However about 25% of tetralogy of Fallot and up to 33% of truncus arteriosus cases have a right aortic arch, and most of those have mirror image branching pattern. In this anomaly the first brachiocephalic vessel is the left innomina-
Figure 11.9 Left aortic arch with retroesophageal diverticulum of Kommerell (right-sided). (a) Axial image showing roughly equal-sized carotid and subclavian arteries. (b) More inferior axial image showing larger diverticulum of Kommerell (Div). (c) More inferior image showing left aortic arch (LAoA) and juncture of Div. (d) Coronal view of LAoA and normal sized right subclavian artery (RSCA). (e) More posterior coronal image showing larger Div joining aorta. (f) Posterior and (g) cranial views of 3-D volume rendering showing abrupt tapering of Div to RSCA. (Compare with Figure 11.5d above – same anomaly in right aortic arch.) DAo: descending aorta; LCA: left carotid artery; LSCA: left subclavian artery; RCA: right carotid artery; T: trachea.
Figure 11.9  Continued
Cardiac MR of congenital and acquired pediatric heart disease

Figure 11.10 Left bronchial compression by ascending and descending aorta in same sagittal plane. (a) Steady-state free precession (SSFP) axial image demonstrating the AAo and DAo in the same sagittal plane, causing compression of RPA and left mainstem bronchus (LBr).

(b) Dark-blood off-axis axial image aligned with the left mainstem bronchus (open arrowheads), demonstrating narrowing as it passes between AAo and DAo. AAo: ascending aorta; DAo: descending aorta; RPA: right pulmonary artery; SVC: superior vena cava.

Figure 11.11 Innominate artery compression of the trachea. Sagittal view, turbo-spin echo (TSE) double inversion dark blood image demonstrating compression of the trachea (open arrowheads) in the anteroposterior dimension at the level that the right innominate artery (RInA) crosses it. Note complete loss of lumen (dotted arrowhead).

Retroesophageal subclavian

This and most of the following five anomalies are due to absence of the embryonic fourth aortic arch. The significance of this is that microdeletion of chromosome 22q11.2 has been associated with arch anomalies, particularly with absence of embryonic arch four [12]. In addition, nearly 40% of patients with trisomy 21 and intracardiac anomalies have retroesophageal right subclavian artery [13]. In this anomaly the branching pattern is the same as for diverticulum of Kommerell. However, the retroesophageal aberrant subclavian has the same caliber throughout its intrathoracic course, that is, there is no diverticulum, indicating that there is no ductus (embryonic sixth arch) attached to the subclavian artery (Figure 11.6; compare with Figure 11.5). Thus there is no vascular ring. Retroesophageal vessels are always opposite the
side of the aortic arch: left arches have retroesophageal right subclavian and right arches, retroesophageal left. In some cases (rarely in children) a condition known as dysphagia lusoria (swallowing difficulty from a trick [of nature]) is caused by compression of the esophagus by the adjacent subclavian artery in the absence of a diverticulum/ring.

**Retroesophageal innominate artery**

This rare anomaly is also derived from absence of the fourth embryonic arch plus absence of the proximal attachment of the third arch (common carotid artery) to the trunco-aortic sac. Therefore this may also be seen in 22q11.2 microdeletion. Though quite rare, this is an example of why the rule that “the first arch vessel usually includes a carotid artery opposite the side of the arch” is not absolute. The branching pattern shows that with a right aortic arch the first arch vessel is a right carotid artery (ipsilateral to the arch), then right subclavian, and finally the retroesophageal left innominate artery (Figure 11.13). While retroesophageal innominate is theoretically possible with either a right or left arch, we are only aware of the right arch variation.

**Isolated vessels**

Isolated subclavian or (very rarely) innominate artery means that the named vessel arises exclusively from the ductus arteriosus – always opposite the side of the arch. These anomalies occur from absence of the embryonic fourth arch and dissolution of the ipsilateral dorsal aorta. Since there is absence of a fourth embryonic arch this anomaly may be associated with 22q11.2 microdeletion. Isolated vessels may occur in isolation or in association with interrupted (contralateral) aortic arch. Clinically this anomaly is characterized by disappearance of the pulse in the affected distribution with closure of the ductus arteriosus. However, in the case of isolated subclavian artery, the arm is still supplied by way of the vertebral artery, sometimes resulting in so-called congenital subclavian steal when the arm is vigorously exercised. This is identifiable with phase-encoded velocity mapping by showing caudal flow in the vertebral artery connected to the isolated subclavian; cephalad flow in the other vertebral and carotid arteries is obvious on velocity mapping (opposite signal intensities i.e. black in one direction and white in another).

**Cervical origin of subclavian artery**

This is a subtle anomaly in which branching from the aorta is normal – innominate, carotid, subclavian from proximal to distal – but with an unusually high take-off of the subclavian artery from the innominate (Figure 11.14). The subclavian artery arises in the neck and descends back into the thorax before heading out to the arm. This anomaly, first described by Kutche and Van Mierop [14] in cases with interrupted aortic arch (see also Figure 11.20b) has been found exclusively in patients with 22q11.2 microdeletion [12], with or without interrupted arch. The connection is provided by the explanation of the presumed embryology by Kutche et al. [14] that this anomaly occurs with absence of the embryonic fourth arch on the opposite side from the definitive aortic arch with persistence of the third, more superior, arch.
Figure 11.13 Right aortic arch (RAoA) with retroesophageal innominate artery. (a) Axial image showing more anterior position of RCA and RSCA and more posterior position of LCA and LSCA. (b) More inferior axial view demonstrating conjunction of LCA and LSCA into LInA arising from a Div running posterior to trachea (T) and esophagus (not well shown). (c) More inferior axial view showing that RCA and RSCA have joined RAoA, and Div joins arch at this level. (d) Anterior and cranial view of 3-D volume rendering of a static gadolinium acquisition demonstrating the posterior origin of Div with ductal origin (open arrow) tethering LInA to MPA. LAA: left atrial appendage. (e) Posterior view of 3-D volume rendering showing LCA and LSCA arising as a LInA from a retroesophageal Div, that is, larger caliber vessel which also gives rise to ductus/ligamentum completing a vascular ring. AAO: ascending aorta; Az: azygos; DAo: descending aorta; Div: diverticulum; LCA: left carotid artery; LInA: left innominate artery; LInV: left innominate vein; LSCA: left subclavian artery; MPA: main pulmonary artery; RCA: right carotid artery; RSCA: right subclavian artery; SVC: superior vena cava.
Other non-ring, non-compressive, arch abnormalities

Persistent 5th aortic arch
Conventional embryology teaching has been that human beings only have embryonic arches one, two, three, four and six. However two anomalies are best understood if one invokes persistence of the embryonic fifth arch: double-barreled aortic arch and common origin of all four brachiocephalic vessels. The first has no physiological consequence but the latter is always associated with coarctation of the aorta [15].

Double-barreled aortic arch
This is an uncommon anomaly first described by Van Praagh et al. [15] in which there are two parallel aortic arches with a common origin at the level of the innominate artery and a common termination at the level of the left subclavian artery (Figure 11.15). Both are widely patent. The more superior of the two, derived from the embryonic fourth arch, gives origin to all three brachiocephalic vessels, and the inferior, from the embryonic fifth arch, has no branches. The only significance of this anomaly is that it presents an unusual appearance on imaging that should not be misinterpreted as a spontaneous dissection of the aorta.

Common origin of all brachiocephalic vessels
In this other variation of persistent fifth aortic arch, just as above, the superior (embryonic fourth) arch gives rise to the arch vessels, but its distal end is atretic. The more inferior (fifth) arch is patent throughout but has a discrete coarctation at its distal aspect. Clinically these patients present with coarctation of the aorta, but they have a distinctive appearance on imaging of all brachiocephalic vessels arising from a common stalk, since the atretic distal portion of the derivative of the embryonic fourth arch is invisible with current techniques (Figure 11.16) but is identifiable in the operating room or on postmortem examination.

Coarctation of the aorta Coarctation of the aorta is a discrete narrowing of the distal aortic arch typically at the level of the ligamentum arteriosum in

![Figure 11.14](image-url)

**Figure 11.14** Cervical origin of subclavian artery from innominate artery. (a) Right anterior oblique view of 3-D volume rendering of static gadolinium acquisition showing very long RInA before RSCA branches off (embryonic arch III) heading inferiorly from its origin in the lower neck. The normal origin of RSCA (embryonic arch IV) is depicted by dotted line. See text for complete description. (b) Posterior cranial view of 3-D volume rendering shows the unusual inferior orientation of the origin of RSCA from posterior aspect of RInA. AAo: ascending aorta; DAo: descending aorta; LCA: left carotid artery; LSCA: left subclavian artery; RCA: right carotid artery; RInA: right innominate artery; RSCA: right subclavian artery.
the native state but can occur at the superior aspect of the aortic arch after previous intervention. It is frequently associated with minor intracardiac abnormalities such as bicuspid aortic valve and/or mitral valve abnormalities including parachute mitral valve, double orifice mitral valve, or decreased interpapillary distance. Important features to be evaluated by MRI include the following:

1. Location relative to other arch vessels – left subclavian artery in unrepaired cases, left carotid artery in previously repaired cases where heterotopic “recurrence” may be present.
2. Extent of any pre-coarctation isthmic hypoplasia, which may necessitate a more extensive surgical or catheter intervention.
3. Presence and quantification of collateral flow around the coarctation.
4. Previously mentioned intracardiac abnormalities.
5. Presence of true aneurysms (with or without previous successful repair of coarctation) or pseudoaneurysms of the aorta (after previous surgical or catheter treatment).

Because the coarctation shelf itself may be very thin, it may not be visualized on dark-blood imaging because of partial volume effects but may be recognized by the turbulent jet seen on steady state free precession imaging (Figure 11.17). Collateral vessels are enhanced with gadolinium (Figure 11.18). Quantification of collateral flow as described by Higgins et al. [16] is accomplished by measuring flow in the descending aorta immediately beyond the coarctation and comparing that with flow just above the diaphragm (Figure 11.19). Normally there should be a small decrease owing to flow into the intercostals arteries, but in coarctation, the distal flow is higher than the proximal, owing to collateral flow usually originating from the left subclavian artery above the coarctation and flowing into the descending aorta by way of the intercostals.
Interrupted aortic arch type B This is an arch anomaly in which one or both embryonic fourth arches have involuted. As with other anomalies involving involution of the fourth embryonic aortic arch, a large number of these patients have DiGeorge syndrome [17]. Involvement of only one fourth arch is accompanied by the normal involu-
Figure 11.19 Quantifying collateral flow. (a) Orientation image showing location of coarctation. Short white line segments indicate locations for obtaining flow measurements. (b) Off-axis image plane just distal to coarctation indicating region of interest (open circle) for obtaining flow measurement. (c) Velocity-time curve permits quantification of flow through region of interest in (b): 1.0 L/min. (d) Axial image plane distal to likely entry of collateral vessels indicating region of interest (open circle) for obtaining flow measurement. (e) Velocity-time curve permits quantification of flow through region of interest in (d): 3.1 L/min. Therefore, collateral flow is 2.1 L/min.
Marfan syndrome is a disorder of fibrous connective due to mutation of the fibrillin-1 gene located on chromosome 15. Cardiovascular manifestations are typically dilation of aortic root and aortic arch; although aneurysms of the thoracic and abdominal descending aorta are known. In severe cases dissection of the aorta may occur. MRI has been used to follow aortic dimensions in a systematic manner. The relatively large field of view relative to that achievable with echocardiography facilitates measurement of a true cross-sectional diameter at

subclavians from the descending or one subclavian from the descending and the other from the ductus/ligamentum, that is, isolated subclavian artery. By definition there is no aortic arch, so the concept of arch-sidedness seems absurd. However the presence of an innominate artery, retroesophageal subclavian artery, or isolated subclavian artery all indicate contralateral arch-“sidedness.” The importance of this is that it has been shown that interrupted aortic arch type B with a “right arch” branching pattern is always associated with the DiGeorge syndrome.
consistent locations over time. Furthermore, one can image the entire aorta looking for localized dilatation. Dissections of the aorta can be recognized by slower flow in the false lumen segment of the aorta (Figure 11.21).

*Turner syndrome* Turner syndrome is a disorder caused by complete or partial loss of a second sex chromosome, affecting around 1 in 2000 girls. Cardiovascular manifestations are typically aortic valve abnormalities including bicuspid aortic valve and aortic coarctation. However, it has recently become increasingly recognized that these patients are at risk for aortic root and arch dilation. Furthermore, these patients are at significant risk for aortic dissection and a relatively young age, and the risk appears related to aortic diameter indexed to body surface area [18]. Therefore, routine screening and monitoring of the aorta in this patient population is warranted using CMR (Figure 11.22).

*Williams syndrome* Williams syndrome is a collection of abnormalities, usually associated with deletion of several contiguous genes on chromosome 7. The cardiovascular manifestations include sup-
ravalvar aortic stenosis and multiple stenoses or hypoplasia of the branch pulmonary arteries but can also include long segment hypoplasia of the ascending or descending aorta (Figure 11.23). This can be demonstrated with either black-blood or white-blood imaging sequences.

**Pulmonary artery anomalous origins**

One might wonder why a discussion of anomalous origin of a branch pulmonary artery is included in a chapter on aortic arch anomalies. Normally the branch pulmonary arteries arise from the embryonic trunco-aortic sac as do the embryonic aortic arches. The following three anomalies are thought to occur when a pulmonary artery branch has a different origin. All can be easily imaged with CMR dark and bright blood images as well as 3D gadolinium imaging.

**From ascending aorta (so-called “hemitruncus”)**

In this rare anomaly one pulmonary artery branch arises normally from the pulmonary artery portion of the trunco-aortic arch, while the other branch arises from the aortic portion of the trunco-aortic sac. Two variations are described [19]: anomalous right pulmonary artery, comprising 82%, most of which do not have intracardiac disease, though some have interrupted aortic arch, mainly in association with aortico-pulmonary window; and anomalous left pulmonary artery, nearly three-quarters of whom have tetralogy of Fallot.

**From ductus (so-called “absent” pulmonary artery)**

This anomaly usually occurs in association with tetralogy of Fallot but may occur in isolation or occasionally with other anomalies. In this anomaly the branch pulmonary artery and the ipsilateral embryonic sixth arch have no connection to the trunco-aortic sac but instead the branch pulmonary artery connects to the embryonic sixth arch, which only has a distal connection to the (dorsal) descending aorta.

**Origin of left pulmonary artery from right (so-called pulmonary artery “sling”)**

If the left pulmonary artery arises from the right pulmonary artery instead of the main pulmonary artery, it passes between the trachea and esophagus forming a vascular sling around the trachea (Figure 11.24). This is often associated with severe tracheal narrowing and even complete cartilaginous rings. In cases without intracardiac disease there is a ductus or ligamentum between the main pulmonary artery and descending aorta which means that the trachea is completely surrounded by vascular tissue (although the ligamentum is not patent): main pulmonary artery anteriorly, left pulmonary artery to the right and posteriorly, and the ligamentum to the left. This does not meet the definition of a vascular ring since the esophagus is outside the “ring” of vascular tissue.

**Summary**

Abnormalities of the aortic arches and related branching patterns are readily diagnosed with magnetic resonance imaging. Direct visualization of the aorta and arch vessels in relationship to the trachea is more reliable than ultrasound and
conventional angiography where arch sidedness is frequently determined indirectly by branching patterns. What is more, vascular rings, while suspected by barium esophagography are directly visualized in all their detail so as to direct optimal surgical division. This chapter has identified all of the major arch anomalies and many of the less common ones as well as methods for identifying vascular rings even when part of the ring is non-patent and therefore not visible with any imaging modality. Magnetic resonance imaging is the optimal imaging tool for demonstrating all arch anomalies without the need for ionizing radiation.

References


CHAPTER 12

MR assessment of pulmonary circulation

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The purpose of the pulmonary circulation is to carry blood to and away from the lungs for gas exchange. Clinicians and researchers are increasingly viewing the right ventricle, the pulmonary arteries and veins, and the capillary bed as one functional unit rather than independent components. With this in mind, there are five facets in the anatomy and physiology of pulmonary circulation:

1. Vascular luminal anatomy.
2. Anatomy and function of the vessel walls.
4. Driving force behind this flow, usually generated by the right ventricle.
5. Oxygen saturation.

Many imaging modalities are employed in the assessment of the pulmonary circulation, each of which with their own strengths and limitations. Echocardiography is an excellent tool for the assessment of vessels in the mediastinum but cannot be used for the pulmonary vessels within the lungs because of the lack of acoustic windows. Even central pulmonary arteries and pulmonary veins may be difficult to visualize because of poor acoustic windows. Contrast-enhanced computed tomography (CT) clearly defines not only the pulmonary vascular anatomy but also the lung parenchyma, central airways and mediastinum. In addition, multidetector CT technology allows imaging of the whole thorax within a few seconds, rarely requiring the need for sedation or general anesthesia. However, CT hardly provides any information about the hemodynamics of the pulmonary circulation or cardiac function. There is also the major disadvantage of the high radiation dose. Radioisotope perfusion scan is merely a crude tool for the assessment of the perfusion defects in suspected pulmonary thromboembolism. It can be used to examine the distribution of the pulmonary blood flow between the lungs and within a lung in those with pulmonary arterial or venous pathologies, but offers little anatomical detail. Similar to CT, there is also the major disadvantage of the high radiation dose. Catheterization with angiography has been the gold standard diagnostic tool for the assessment of the pulmonary circulation as it not only provides images of the highest temporal and spatial resolutions, but it is also the only tool that measures the pulmonary arterial pressure directly. However, catheterization is used as the last resource because of the invasive nature of the study, exposure to ionizing radiation as with the previous two modalities, and potential harmful effects of iodine containing X-ray contrast medium.

MR, in comparison, overcomes most of these disadvantages [1]. The comprehensive assessment of all these aspects of the pulmonary circulation is one of the greatest advantages of cardiovascular
magnetic resonance (MR) imaging as compared to other imaging modalities. First, it is able to assess all mediastinal and peripheral vessels in any orientation without any significant interference by the lungs, bones or scars. Second, it provides comprehensive hemodynamic details including direction, velocity, volume and pattern of blood flow, the distensibility and recoilability of the vessel wall, and ventricular function. Third, MR is non-invasive and does not use radiation. However, this modality faces challenges in the examination of the pulmonary circulation including artifacts from metallic implants, from the interface between the air and tissue, as well as those from respiratory and cardiovascular motion. The greatest limitation of MR in the assessment of the pulmonary circulation is, as in echocardiography, its inability to directly measure pressures.

This chapter outlines the techniques and applications of MR in the assessment of pulmonary circulation in selected clinical situations.

**MR techniques**

The following MR techniques are routinely used for the assessment of the pulmonary circulation: gradient-echo white-blood cine techniques, spin-echo black-blood techniques, contrast-enhanced angiography, phase-contrast velocity mapping.

**Gradient-echo white-blood cine sequences**

We rely on cine gradient-echo imaging using balanced steady-state free precession (SSFP) sequence for anatomical assessment of the central pulmonary vessels (Figure 12.1). In addition to providing anatomic information of the larger vessels, it also allows perception of the dynamic changes in vessel size during the cardiac cycle [2]. This technique, however, is vulnerable to artifacts from turbulent flow. The SSFP sequence requires a line TR (repetition time) of less than 4 milliseconds, and to comply with this requirement, it may compromise spatial resolution. Ultrafast gradient-echo sequence can be used when balanced SSFP is associated with major artifacts. With recent advances in hardware and software, these limitations are typically overcome and with further advances, this should not be an issue.

**Spin-echo black-blood techniques**

Fast spin-echo black blood sequence, often combined with double inversion recovery technique, can be used for the anatomical assessment of the central pulmonary vessels with a higher spatial resolution than that provided by gradient-echo techniques (Figure 12.2). However, it only provides static images, which limits its use for pulsatile vessels. Double inversion recovery sequence suffers less from susceptibility artifacts introduced by metallic foreign bodies. Therefore, it can be used for the assessment of stented vessels. However, it should be remembered that the absence of any
signal from inside the stent in spin-echo images does not prove the absence of obstruction from intimal proliferation or clot. Spin-echo sequences are also suitable for imaging of the airways and lung parenchyma.

**Contrast-enhanced angiography**

Three-dimensional contrast-enhanced angiography is best suited for the anatomic assessment of pulmonary arteries and veins that show tree-like branching (Figure 12.3). It is also very useful in the assessment of frequently tortuous systemic-to-pulmonary collateral arteries [3,4]. It is worthwhile to perform angiography early during the study because the imaging planes for further assessment of the pulmonary vessels can be tailored more easily and precisely. Time-resolved angiography can be used for the qualitative and quantitative evaluation of the regional distribution of pulmonary blood flow (Figure 12.4) [5–8].

**Phase-contrast velocity mapping**

Phase-contrast velocity mapping is advantageous over Doppler ultrasound in the hemodynamic
assessment of the pulmonary circulation because it is not affected by the presence of air and bones [9,10]. Consequently, the pulmonary vessels within the aerated lungs, as well as those in the mediastinum, can be assessed [11]. Phase-contrast imaging across the target vessel, referred to as through-plane imaging, allows accurate measurement of the average blood flow velocity and blood flow volume. We have recently shown that pulmonary venous blood flow volumes can be measured accurately [12]. A temporal resolution that accommodates at least 20 real data points within a cardiac cycle should be sought for accurate quantification of the blood flow velocity and volume. The temporal resolution of the phase-contrast imaging is calculated by the equation: 

\[ \text{Temporal resolution} = 2 \times TR \times \text{VPS} \]

where TR is the repetition time and VPS is the views or lines in k-space per segment. In order to accommodate >20 data points per cardiac cycle, the VPS should be adjusted according to the heart rate. Imaging during free breathing, with two or three excitations (or averages), is recommended as pulmonary blood flow changes significantly with prolonged breath-holds [11]. Figure 12.5 illustrates the normal patterns of the pulmonary arterial and venous blood flow curves acquired from phase-contrast imaging.

**MR oximetry**

In addition to these commonly employed sequences, the patient’s blood oxygen saturation (%HbO2) can be determined by MR. This technique relies on the mathematical relationship between the %HbO2 and the T2 relaxation time of blood: 

\[ \frac{1}{T_2} = \frac{1}{T_{2O}} + K \left( \frac{1 - \%O_2}{100} \right)^2 \]

where T2O is the T2 signal decay of fully oxygenated blood and K is a constant [13]. It has been found that both T2O and the constant K can be estimated if the patient’s hematocrit and fibrinogen concentration are known [14]. Therefore, the %HbO2 can be calculated using the T2 relaxation time of the blood measured by MR and the T2O and K values estimated from the patient’s blood sample.
**MR in specific conditions affecting pulmonary circulation**

**Pulmonary arterial anatomy in tetralogy of Fallot with pulmonary atresia**

When tetralogy of Fallot is associated with pulmonary atresia, the pulmonary arteries are supplied by either a patent ductus arteriosus or major aortopulmonary arterial collateral arteries (MAPCAs). If the source of pulmonary blood supply is a patent ductus arteriosus, the pulmonary arteries are usually confluent in the mediastinum and show a normal branching pattern. Rarely there can be non-confluent branch pulmonary arteries which are supplied by separate right and left arterial ducts. If the source of pulmonary blood flow is via MAPCAs, the pulmonary arterial anatomy is variable and often complex, with each lung supplied by multiple MAPCAs. The lung regions supplied by the MAPCAs may be connected to each other through communicating channels, or they can be compartmentalized or isolated from each other without communicating channels. The true branch pulmonary arteries and pulmonary trunk may or may not be present. Rarely, one lung is supplied by a patent ductus arteriosus, and the other lung by multiple MAPCAs.

MR is very useful in preoperative assessment of tetralogy of Fallot with pulmonary atresia and MAPCAs (Figure 12.6) [15]. Complete mapping of the anatomy of the collateral and pulmonary arteries is crucial as the surgical aim is to unifocalize the pulmonary arterial tree by uniting the MAPCAs to a common channel that can then be connected to the right ventricle via a conduit. Three-dimensional contrast-enhanced angiography can be used for noninvasive assessment of the origin, size and course of the MAPCAs [15,16]. Unfortunately, MR angiography is unable to clearly define whether the pulmonary segments supplied by more than one MAPCA are interconnected by communicating channels. Quantification of blood flow volume and assessment of the blood flow pattern of individual MAPCAs is possible using phase-contrast imaging. The total blood flow to each lung can be estimated by measuring the pulmonary venous return through the individual pulmonary veins. Although
Figure 12.6 Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries in a 7-month-old infant. The patient underwent unifocalization surgery for the left pulmonary artery by using a pericardial roll in the neonatal period. (a) Volume-rendered contrast-enhanced angiogram seen from front in a shows that the modified Blalock-Taussig (BT) shunt connects to the aneurysmally dilated pericardial roll, and the left pulmonary vein (LPV) is stenotic. A posterior view in (b) shows that the right lung is supplied by a large collateral artery (MAPCA #1) arising from the descending aorta (Ao), with the non-confluent right pulmonary artery (RPA) seen posterior to the ascending aorta (Asc Ao). Values for flow, including Qp and Qs, are in liters/minute/m² of body surface area. LA: left atrium; LIA: left innominate artery.

Abnormal origin, branching and course of the pulmonary arteries
Various forms of abnormal origin of a branch pulmonary artery have been described. These include left pulmonary artery slings, anomalous origin of a branch pulmonary artery from the ascending aorta, and unilateral absence of a pulmonary artery.

In patients with a left pulmonary artery sling, the left pulmonary artery arises distally from the right pulmonary artery on the right side of the distal trachea and courses to the left lung hilum through the space between the distal trachea and the esophagus (Figure 12.7) [17,18]. A left pulmonary artery sling is commonly associated with an abnormal branching pattern of the trachea and bronchi. Airway obstruction in left pulmonary artery sling is more commonly an intrinsic stenosis of the trachea due to complete cartilaginous rings and less commonly due to extrinsic compression from the aberrant left pulmonary artery with or without tracheomalacia.

A branch pulmonary artery, more commonly the right than the left, may arise from the proximal or distal part of the ascending aorta [19]. Anomalous origin of the right pulmonary artery from the ascending aorta is uncommonly associated with other structural defects such as aortopulmonary window and tetralogy of Fallot. In contrast, anomalous origin of the left pulmonary artery is almost always associated with tetralogy of Fallot, a right aortic arch, or both. When the anomalous pulmonary artery has an unobstructed origin from the aorta, both the affected and unaf-
the cases occur as an isolated anomaly. Absence of the left pulmonary artery is more frequently associated with another cardiovascular malformation, tetralogy of Fallot with or without pulmonary atresia being the most common. The absent pulmonary artery is almost always on the opposite side of the aortic arch and therefore connected to the ductus arteriosus arising from the innominate artery. The remnant of the ductus arteriosus is seen as a small diverticulum projecting from the base of the innominate artery. The blind ending proximal pulmonary artery is usually identifiable at the lung hilum.

These abnormalities can be best defined by contrast-enhanced angiography with three-dimensional reconstruction (Figures 12.7 and 12.8) [18]. Alternatively, a stack of static contiguous axial SSFP images can be used to assess this at a lower resolution than the contrast enhanced angiography. Associated airway malformation can be assessed by using a fast spin-echo sequence or a
one or both branch pulmonary arteries is not an uncommon complication of the arterial switch operation using a “LeCompte maneuver” in which the transected main pulmonary artery and its branches are brought forward to drape around the newly reconstructed ascending aorta (Figure 12.10) [22]. Isolated stenosis of a branch pulmonary artery unrelated to surgery is uncommon. Stenosis of the left pulmonary arterial origin is often seen in tetralogy of Fallot with pulmonary atresia and occasionally in severe forms of tetralogy of Fallot and pulmonary atresia with intact ventricular septum. This condition is known as “juxtaductal stenosis” because the stenosis is at the site of the ductal insertion and related to the closure of the ductus arteriosus [19]. Peripheral pulmonary artery stenosis is usually multifocal (Figure 12.11). It can occur as an isolated disease but it is more commonly a manifestation of a systemic condition, such as Williams–Beuren, Alagille, Ehlers–Danlos, Holt–Oram, Keutel, Noonan, or post–rubella syndrome, or Takayasu arteritis [23].

Contrast-enhanced three-dimensional MR angiography is ideal for the morphologic assessment of pulmonary arterial stenosis especially when multifocal peripheral stenoses are suspected. Breath-holding is crucial for this sequence when peripheral pulmonary arteries are involved. It should be noted, however, that the contrast-enhanced angiography typically underestimates the size of vascular structures as this non-gated static sequence averages the data from multiple cardiac cycles during which the size of the pulmonary arteries change. Some vendors offer ECG triggered sequences which increase fidelity but at the cost of speed. If the pulmonary artery is uniformly hypoplastic along its course, accurate measurement are best performed on ECG-gated cine images in a plane perpendicular to the vessel course. The velocity across the stenotic lesion can be assessed by using phase-contrast imaging and then translated into an approximate pressure gradient, using the modified Bernoulli equation. It must be remembered, however, that neither the pressure gradient across the stenosis, nor the pulmonary arterial size, necessarily reflect the severity of the stenosis because of redistribution of blood flow from the pathological area to other parts of the lung [24]. The reduced size of the pulmonary...
Figure 12.10 Pulmonary arterial stenosis in a 5-year-old child after the arterial switch operation using the LeCompte maneuver for complete transposition of the great arteries. Contrast-enhanced angiograms reformatted in an oblique coronal (a) and axial (b) planes show that the right and left pulmonary arteries (RPA and LPA, respectively) drape around the anterior aspect of the ascending aorta (Ao). The left pulmonary artery is diffusely small. The blood flow volume through the branch pulmonary arteries and individual pulmonary veins are shown in (b). Blood flow volumes are in liters/minute/m² of body surface area. Note that the blood flow volumes through the branch pulmonary arteries are close to the volumes drained through the pulmonary veins. LLPV: left lower pulmonary vein; LUPV: left upper pulmonary vein; RLPV: right lower pulmonary vein; RUPV: right upper pulmonary vein; MPA: main pulmonary artery; SVC: superior vena cava; LV: left ventricle.

Figure 12.11 (a & b) Peripheral pulmonary arterial stenosis in a 7-year-old child who previously underwent surgery for supravalvar aortic stenosis. The patient did not show other features of William-Beuren syndrome. Contrast-enhanced angiograms show hypoplastic right and left pulmonary arteries (RPA and LPA, respectively) and multifocal stenosis of the peripheral pulmonary arteries. The repaired ascending aorta (Ao) is still hypoplastic. LA: left atrium; LV: left ventricle; RV: right ventricle.

vein draining the pathological area reflects the hemodynamic consequence of the pulmonary arterial stenosis [25]. The blood distribution between the right and left lungs can be accurately assessed with phase-contrast imaging of the branch pulmonary arteries [24,26]. When the blood flow through a branch pulmonary artery cannot be
However, the maximum velocity and pattern of the blood flow through the affected pulmonary vein does not necessarily predict the severity of obstruction because pulmonary venous obstruction results in redistribution of blood flow to the unaffected area of the same lung and to the contralateral lung such that the affected lung segment has a reduced amount of blood flow [30,31]. Therefore, the size of the pulmonary artery as well as the blood flow through both the affected and unaffected lungs should be assessed in the presence of an obstructed pulmonary vein. The flow redistribution in the pulmonary arteries is characterized by reduced forward flow in systole and reversal of flow in diastole in the pulmonary artery to the affected lung area (Figure 12.12c) [31]. The pulmonary artery to the unaffected lung shows increased systolic blood flow and continuous forward flow in diastole. This can all be assessed using phase contrast velocity mapping. Severe pulmonary venous obstruction can lead to pulmonary hypertension, the MR signs of which are discussed later in this chapter.

Total anomalous pulmonary venous connections

Total anomalous pulmonary venous connection is relatively uncommon. In one third of the cases, it is associated with other major cardiac defects. It is a frequent feature of the heterotaxy syndromes. The sites of anomalous connections include the left innominate vein, the coronary sinus, the ductus venosus-portal venous system, the right superior vena cava, and the right atrium in order of decreasing frequency [32,33]. In 5–10% of cases, the anomalous connection is to multiple sites. In total anomalous pulmonary venous connection, systemic output can only be maintained by right-to-left shunting, which is almost always at atrial level. Obstruction in total anomalous pulmonary venous connections can occur at any anatomical level from the individual pulmonary veins, the site of the abnormal connection, or at the atrial level. Obstruction is an important risk factor for adverse outcomes, especially in right isomerism.

Echocardiographic diagnosis can be a challenge and may take a long time especially when the anomalous pulmonary venous drainage is to multiple sites or occurs in the setting of complex con-
MR in particular, is advantageous over ultrasound in defining the complex vascular anatomy in this malformation. Each individual pulmonary vein, the venous confluence and the drainage channel to the site of the anomalous connection must be assessed to exclude obstruction (Figure 12.13).

Precious time wasted in searching for anomalous pulmonary veins by echocardiography, especially in severely compromised infants such as these may be, can potentially mean the difference between a good and bad outcome. MR, contrast-enhanced
PART II Cardiac MR of congenital and acquired pediatric heart disease

Arterial pressure is significantly elevated and the right ventricle is typically pressure, rather than volume, overloaded.

Partial anomalous pulmonary venous connections

Partial anomalous pulmonary venous connection is less common than total anomalous connection [32]. Partial anomalous pulmonary venous connection is usually from one lung, more commonly the right. The anomalous pulmonary vein(s) from the right lung connect to the superior vena cava, and less commonly to the azygos vein, right atrium, coronary sinus, or inferior vena cava. The anomalous pulmonary vein(s) from the left lung connect to the innominate vein, coronary sinus, right atrium, inferior vena cava, or ductus venosus-portal venous system. The anomalous veins can also connect to multiple sites. Obstruction is rarely, if ever, encountered. Anomalous connection of the upper or all veins from the right lung to the superior vena cava is usually associated with a sinus venosus type of atrial septal defect (Figure 12.15). Other forms of partial anomalous connection are often associated with a secundum type of atrial septal defect or patent foramen ovale.
Figure 12.14 Total anomalous pulmonary venous connection in a newborn baby with right isomerism and dextrocardia on contrast-enhanced angiography. Maximum intensity projection images (upper panels) and volume rendered 3D images (lower panels) show complex pulmonary and systemic venous anatomy. The right upper, right lower and left lower pulmonary veins (RUPV, RLPV and LLPV, respectively) make a confluence (C) behind the atrium and drains to the upper part of the left superior vena cava (LSVC) through a vertical vein (VV). The upper half of the left lung is drained by two veins (LUPV[A] and LUPV[P]) that have separate connections to the left superior vena cava. The vertical vein and left upper pulmonary veins are narrowed as they connect to the left superior vena cava. The right superior and inferior vena cavae (RSVC and RIVC) connect to the right-sided atrium (R-A). The LSVC connects to the left-sided atrium (L-A). The hepatic veins draining the left lobe of the liver form a confluence to connect to the left-sided atrium. The patient previously underwent left Blalock-Taussig (BT) shunt. Ao: aorta; PA: pulmonary artery; RPA: right pulmonary artery; LPA: left pulmonary artery.
diagnosis, and can easily assess when the management is based on the Qp/Qs [34–39]. Many times, the only suspicion on echocardiography is a qualitatively dilated right atrium or ventricle. Cine gradient-echo imaging and contrast-enhanced angiography depict the vascular anatomy and the spatial relation of the anomalous pulmonary

The scimitar syndrome is a particular form of partial anomalous pulmonary venous connection that will be discussed in the following section.

MR is the diagnostic method of choice in patients with partial anomalous pulmonary venous connection, can confirm the echocardiographic diagnosis, and can easily assess when the management is based on the Qp/Qs [34–39]. Many times, the only suspicion on echocardiography is a qualitatively dilated right atrium or ventricle. Cine gradient-echo imaging and contrast-enhanced angiography depict the vascular anatomy and the spatial relation of the anomalous pulmonary

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**Figure 12.15** Partial anomalous pulmonary venous connection to right superior vena cava. (a) Volume-rendered contrast-enhanced angiogram seen from the front shows three pulmonary veins (asterisks) connecting to the superior vena cava (SVC). (b) Cine gradient image through the anomalous vein and atrial septum shows the anomalous right upper pulmonary vein and a sinus venosus defect (D). The flow volume data are summarized in (c). Ao: ascending aorta; ao: descending aorta; BSA: body surface area; LA: left atrium; LV: left ventricle; PA: main pulmonary artery; RA: right atrium; RV: right ventricle; LUPV, LLPV, RUPV and RLPV: left upper, left lower, right upper and right lower pulmonary veins respectively. LPA and RPA: left and right pulmonary arteries; SV: sinus venosus.
vein(s) to the left atrium and, if present, the atrial septal defect (Figure 12.15b). Contrast-enhanced angiography requires good simultaneous visualization of the systemic and pulmonary veins, which can be achieved either by prolonged injection of contrast so that both pulmonary and systemic veins are opacified or by time-resolved technique. The size and function of the volume loaded right ventricle can also be assessed. Phase-contrast imaging of the right and left pulmonary arteries and ascending aorta provides accurate data for calculation of Qp/Qs. The hemodynamic effects of the anomalous pulmonary venous connection and the atrial septal defect, when present, can be separated by obtaining the flow volumes of individual pulmonary veins or the flow volumes upstream and downstream to the anomalous connection as well as the total pulmonary blood flow volume.

**Scimitar syndrome**

Scimitar syndrome is a complex association of: (a) a varying degree of right lung hypoplasia; (b) anomalous and sometimes obstructed pulmonary venous drainage of a part or all of the right lung to the post-hepatic segment of the inferior vena cava; and (c) aberrant systemic arterial supply to the right lower lung [40]. It is often associated with abnormal bronchial branching and lung parenchymal pathologies, such as horseshoe lung and crossover lung segment. A rare variant of the scimitar syndrome involves a scimitar-shaped right pulmonary vein that connects correctly to the left atrium. This condition has been called “meandering pulmonary vein” and shares many of the other features of scimitar syndrome [41].

MR not only defines the vascular anatomy and associated pathology of the lungs and airway, but also provides the information regarding the hemodynamic effects of the constellation of abnormalities (Figure 12.16) [42,43]. The severity of right lung hypoplasia can be appreciated in the scout images in axial and coronal planes. The lung volumes can be measured from the stack of scout images in axial plane or by using a volumetric interpolated three-dimensional gradient sequence that can acquire a whole lung dataset during a single breath-hold. The airway can be further assessed by using double-inversion recovery black-blood technique. It is worthy assessing whether...
there is any right diaphragmatic abnormality or abnormal fusion of the right lower lung and the underlying liver. Contrast-enhanced angiography should be tailored to include the upper abdomen to the level of the origins of the celiac axis and superior mesenteric artery where the aberrant systemic artery usually arises. We prefer a static to a dynamic sequence for contrast-enhanced angiography in order to achieve a good spatial resolution as the abnormal vessels can be small. When a static sequence is used, a larger volume of contrast medium (0.2 mMol/kg of gadolinium compound) is injected for a prolonged period of time so that all the pulmonary arteries, veins and systemic arteries are visualized at the same time. The remainder of the assessment using phase-contrast and cine MR is similar to that for other forms of partial anomalous pulmonary venous connection.

**Pulmonary regurgitation**

Pulmonary regurgitation is an inevitable complication of surgical repairs such as those for tetralogy of Fallot and truncus arteriosus as the repair involves augmentation of the right ventricular outflow tract with or without pulmonary valvotomy or placement of a right ventricle-to-pulmonary artery conduit. Pulmonary regurgitation also complicates the Ross and Ross–Konno procedures in which the native pulmonary valve is used for aortic valve replacement and a valved-conduit is placed in the pulmonary outflow tract. As chronic pulmonary regurgitation is associated with right and left ventricular dysfunction, exercise intolerance, arrhythmia, and increased risk of sudden death, it is the major determinant of the long-term outcome after a surgical repair [44–46]. Therefore, regular follow-up of the severity of pulmonary regurgitation and right ventricular function is crucial. Both pulmonary regurgitation and right ventricular function can most accurately and quantitatively be assessed with MR (Figure 12.17) [45–48]. Phase-contrast imaging is performed across the main, right, and left pulmonary arteries to quantify the systolic forward, diastolic retrograde, and net forward flow volumes through each vessel. Precise targeting of the main pulmonary artery for through-plane phase-contrast imaging can be difficult and the flow data may have some inaccuracies because the short main pulmonary artery moves up and down and tumbles back and forth in the presence of important regurgitation. This mechanism can lead to an underestimation of the regurgitant fraction by as much as 10–15%, as

![Figure 12.17 Pulmonary regurgitation in a patient who previously underwent Ross–Konno operation. (a) Diastolic frame from cine gradient imaging shows a large degree of regurgitant turbulent flow in the right ventricular outflow tract. (b) Time–flow volume curve from phase-contrast imaging of the main pulmonary artery shows pulmonary regurgitation. In late diastole, there is antegrade flow, which represents restrictive physiology of the stiff right ventricle. In this period, the flow generated by atrial contraction is directly forwarded into the pulmonary artery. Ao: aorta; MPA: main pulmonary artery; RA: right atrium; RV: right ventricle.](image-url)
well as to an overestimation of net forward flow in the main pulmonary artery [10,11]. When the main pulmonary artery is difficult to target for accurate measurement, total pulmonary regurgitant volume and regurgitant fraction can be calculated by adding the volumes measured for the right and left pulmonary arteries. However, it should be remembered that not all of the retrograde flow through the pulmonary artery reaches the right ventricle.

Both regurgitant volume and regurgitant fraction are equally important parameters in the evaluation of pulmonary regurgitation [45]. The regurgitant volume is the exact amount of volume overload that pulmonary regurgitation causes to the right ventricle. The regurgitant fraction is the regurgitant volume divided by the systolic forward flow volume. The pulmonary regurgitant fraction varies according not only to the absolute pulmonary regurgitant volume but also to the right ventricular systolic and diastolic function. Therefore, the regurgitant fraction alone is meaningless unless the regurgitant volume and right ventricular function are known. For instance, a patient with a modest amount of absolute pulmonary regurgitant volume and a right ventricle with relatively normal volume and function may have a regurgitant fraction identical to that seen in a patient with a huge regurgitant volume but a proportionally larger right ventricular stroke volume [45]. Conversely, reduced systolic function with a decreased stroke volume results in a disproportionately high regurgitant fraction for the severity of disease. When the right ventricular systolic function is preserved, pulmonary regurgitation results in increased ejection fraction. The so-called corrected ejection fraction is calculated by dividing the net forward flow volume by the end-diastolic right ventricular volume, thus taking both systolic ventricular function and the degree of regurgitation into account [49]. Use of the corrected ejection fraction, as mentioned elsewhere in this textbook, is controversial regarding how physiologically meaningful it is.

Severity of pulmonary regurgitation is not only related to the anatomical integrity of the pulmonary valve but also affected by the diastolic function of the right ventricle. The pressure gradient for regurgitation across the pulmonary valve is reduced with increased pressure of the dysfunctional right ventricle in the later part of the diastole. With further impairment of the right ventricular diastolic function, the stiff right ventricle acts as a passive conduit during atrial contraction. This causes the blood flow generated by atrial contraction to be directly transmitted into the pulmonary artery. This forward flow in the pulmonary artery during the late diastolic phase characterizes the so-called restrictive physiology of the right ventricle (Figure 12.17b) [50,51]. As the stiff right ventricle limits pulmonary regurgitation and is resistant to dilatation, the restrictive physiology appears to carry a favorable prognosis in patients with chronic pulmonary regurgitation, albeit this observation is not without controversy [50,51].

Interestingly, pulmonary regurgitation derives more regurgitant blood flow from the left lung than from the right lung in the majority of cases with repaired tetralogy of Fallot (Figure 12.18) [52,53] and has been demonstrated in other lesions which also result in pulmonary regurgitation [54]. Infrequently the regurgitation is exclusively from the left pulmonary artery. Although its mechanism remains uncertain, differential regurgitation between the right and left pulmonary arteries contributes to differential lung perfusion. Therefore, the size of a branch pulmonary artery during systole does not mirror the net blood flow volume to the lung parenchyma when there is pulmonary regurgitation [52]. It has been suggested that in the absence of pulmonary artery or vein stenosis, that the branch pulmonary artery regurgitation fraction reflects the individual lung resistances [54]. The ratio of the branch pulmonary arterial size is poorly predictive of the ratio of lung perfusion [24,25,52,54]. It has recently been suggested that phase-contrast imaging may underestimate the net forward flow volume when there is disproportionately severe regurgitation from a branch pulmonary artery [53]. Likewise, when there is severe dilatation of the pulmonary arteries in association with absent pulmonary valve syndrome, swirling blood flow within the capacious pulmonary arteries may not allow accurate quantification (Figure 12.19). Quantification of pulmonary venous blood flow then can be used as more accurate tool for the assessment of the net blood flow to each lung [12].
Both surgical pulmonary valve replacement and percutaneous implantation of a valved stent requires preoperative assessment of the right ventricular outflow tract morphology and its relationship to the major coronary arteries. For this purpose, a three-dimensional volume rendered image or plastic model of the right ventricular outflow tract can be produced from three-dimensional SSFP or contrast-enhanced imaging [55,56].

Despite the ability to accurately assess the anatomy of the right ventricular outflow tract and quantify pulmonary regurgitation, it remains con-
MR assessment of pulmonary circulation

For surgical planning of a bidirectional cavopulmonary connection and that invasive pressure measurements are not necessary in these patients [60]. If needed, the pulmonary arterial pressure can be measured simply by advancing a central venous cannula into the pulmonary arteries without performing a full catheterization procedure.

When performed, the MR should determine anatomy and: (1) address ventricular function and the degree of atrioventricular valve regurgitation, particularly in the systemic ventricle; (2) rule out the presence of thromboembolic occlusion, intrinsic stenoses and extrinsic compression of the superior and inferior vena cavae, the Fontan tract, and the pulmonary arteries and veins; (3) determine the blood flow volumes to both lungs; (4) determine systemic arterial collateral blood flow to the lungs; and (5) identify systemic venous-to-systemic venous or systemic venous-to-pulmonary venous collateral blood flow.

The morphology of the Fontan pathway is best imaged using contrast-enhanced MR angiography (Figure 12.20). When undiluted contrast medium is injected through an arm vein in the presence of a bilateral cavopulmonary connection, the pulmonary arteries fail to be visualized during the first pass of contrast medium because the bolus of gadolinium compound causes a paramagnetic rather than T1-shortening effect [1]. Therefore, pulmonary arteries are imaged in the recirculation phase of the contrast medium. Goo et al. suggested the use of diluted contrast medium in patients with superior cavopulmonary connection or Fontan circulation, a method that is also applied to image deep and superficial venous systems in the upper and lower extremities [57]. Depending on the surgical technique and on pulmonary artery anatomy, blood from the superior vena cava flows preferentially into the left pulmonary artery while the inferior vena cava empties mostly into the right lung in many patients. Simultaneous injection of a reduced amount of diluted gadolinium into the upper and lower extremities is helpful in delineating the complete pulmonary arterial tree at the same time. Time-resolved angiography provides some hemodynamic information regarding flow dynamics through the Fontan circuit. On an investigative level, the flow dynamics in Fontan pathways have been evaluated using velocity vector...
gies, which is to volume-unload the single ventricle. In a study of 93 Fontan operations, two out of three fatal outcomes were associated with unrecognized or recognized excessive collaterals [60]. Although there is some controversy regarding the significance of the collaterals on the post-operative course and long-term outcome, the collaterals are frequently coil embolized before or after the Fontan operation [61,62]. However, coil embolization at cardiac catheterization is based on angiographic findings which vary significantly with the site of injection and the amount of contrast medium. Although contrast-enhanced MR angiography may show large collaterals, visualization is typically poor. We [63] and others [64] have recently showed that the blood flow volume through the systemic-to-pulmonary arterial collaterals can be measured by using phase-contrast imaging (Figure 12.21).

The collateral blood flow volume is simply the volume difference between the pulmonary arterial supply and pulmonary venous drainage, although this amount also includes normal bronchial arterial blood flow volume.

Systemic venous collaterals are common after bidirectional cavopulmonary anastomosis and they are likely to be progressive over time with increased systemic venous pressure [59]. As such, superior-to-inferior vena caval shunting leads to reduce the effective pulmonary blood flow and therefore systemic desaturation, large collateral channels may warrant coil embolization. Large venous collaterals are usually well visualized at contrast-enhanced MR angiography. The blood flow volume through the large collaterals can be directly measured using phase-contrast imaging. Theoretically, the difference between the volumes through the descending aorta at the diaphragm and the inferior vena cava above the diaphragm should be the superior-to-inferior vena caval runoff. However, the blood flow through the inferior vena cava is often difficult to assess because of its short length and turbulence from the hepatic venous confluence although some have done this successfully [64]. Occasionally collateral channels are seen between the systemic veins to the pulmonary veins or pulmonary atrium.

The one-and-a-half ventricular repair consists of a bidirectional cavopulmonary connection without isolation of the pulmonary artery from the pulmonary arterial trunk and intracardiac repair.
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(b)

Figure 12.21 Massive development of systemic collateral arteries secondary to pulmonary venous obstruction in a 5-year-old child with right isomerism, unbalanced atroventricular septal defect, double outlet right ventricle and pulmonary stenosis. The patient underwent bilateral bidirectional cavopulmonary anastomosis. (a) Frontal volume-rendered contrast-enhanced angiogram shows unobstructed connection of the right and left superior venae cavae (SVC) to the confluent pulmonary artery (PA). The pulmonary arterial branches in the right lung are well opacified out to the pleural surface. The left lung is obviously underperfused. Note the prominent collaterals in the left chest wall (arrows) which are the major source of blood flow to the left lung. (b) Angiogram reformatted in axial plane shows membranous obstruction of the left pulmonary vein (LPV). The right pulmonary vein (RPV) is unobstructed. Flow volume assessment using phase-contrast imaging is shown in (b). The left lung takes a large amount of systemic collateral arterial flow but is drained to the right lung through the left pulmonary artery. Ao: aorta. (This case has been reported in Cardiology Young 2007;17:548–550.)

After the operation, the systemic venous return through the superior vena cava flows passively into the pulmonary artery through the cavopulmonary connection and the systemic venous return from the inferior vena cava is pumped into the pulmonary artery by the ventricle. This procedure unloads the hypoplastic or dysplastic right ventricle, while a degree of pulsatile flow is maintained in the pulmonary circulation. In a limited number of cases, right ventricular growth after one-and-a-half ventricle repair allows biventricular repair later in life. Prior to a possible conversion to a biventricular circulation, it is essential to assess to which extent the right ventricle contributes to the pulmonary blood flow. The right ventricular cardiac output in this situation can be determined by: (a) measuring flow in the main pulmonary artery; (b) subtracting flow in the superior vena cava from the sum of the right and left pulmonary arterial flows distal to the cavopulmonary connection; or (c) by volumetric measurement of stroke volume. The last method is relatively inaccurate owing to the coarse trabeculations within the right ventricle.

Pulmonary arteriovenous malformations

Arteriovenous malformations within the lung can be congenital, as in Osler–Rendu–Weber syndrome, or acquired, especially in patients with cavopulmonary anastomoses. Large arteriovenous malformations can be identified by MR angiography, while visualization of small fistulous arteriovenous communications measuring less than 5 mm in size is challenging (Figure 12.22) [65,66]. Small arteriovenous malformations are often difficult to identify even on X-ray angiography with temporal resolution that is superior to that of MR. Time-resolved MR angiography may show prompt filling of the particular pulmonary vein draining the malformation prior to those veins draining the normal lung segments [57]. Phase-contrast imaging of the pulmonary arterial branches supplying the malformations may show continuous flow [1]. The use of MR oximetry, showing undersaturated blood in the affected vein, is potentially useful, but has not been reported.

Pulmonary hypertension

Pulmonary hypertension is a disease of the pulmonary vasculature resulting in a progressive increase
Nevertheless, MR shows a considerable number of abnormal findings in patients with pulmonary hypertension including: changes in pulmonary arterial blood flow pattern, dilatation and reduced distensibility of the central pulmonary arteries, right ventricular enlargement and hypertrophy with impaired systolic and diastolic function, and dilatation of the right atrium and inferior vena cava (Figure 12.23) [67–76].

Recognized hallmarks of the disease during flow assessment include an early systolic peak with decreased maximum velocity, a rapid deceleration with an additional peak(s) in later part of systole, a pronounced flow reversal in early diastole, and irregular oscillations of flow throughout diastole (Figure 12.23b) [68–70]. Sanz et al. showed that the average velocity of pulmonary arterial flow was the most useful single parameter in the evaluation of pulmonary hypertension [70]. Changes in flow pattern following administration of one or more pulmonary vasodilators such as oxygen, nitric oxide, prostacyclin or sildenafil can be monitored during an extended examination, but are usually subtle.

A simple measurement of the branch pulmonary artery diameter is also a good screening tool for pulmonary hypertension [71]. Murray et al. found a strong correlation between the mean pulmonary arterial pressure (PAP) and the ratio of the diameters of the main pulmonary artery (MPA) and descending aorta (AO): mean PAP = 24 × MPA/AO + 3.7 (r = 0.7, P < 0.01) [72]. Increased pulmonary artery pressure causes distension with possible wall remodeling, both resulting in stiffening of the proximal pulmonary arteries. Gan et al. showed that the relative area change ([maximum area – minimum area]/minimum area) is a good predictor of mortality [73]. Sanz et al. revealed a good correlation between the minimum pulmonary arterial cross-sectional area and the pulmonary arterial pressure [70].

The severity of right ventricular hypertrophy and the position of the interventricular septum are also predictors of pulmonary arterial pressure (Figure 12.23c). Frank et al. showed good linear correlations between the mean pulmonary arterial pressure and the end-diastolic thickness of the right ventricle (r = 0.83) as well as the diameter of the inferior vena cava (r = 0.73) [74]. Saba et al. demonstrated that mean pulmonary arterial pres-
closure and tricuspid valve opening, has been suggested as a marker for the severity of pulmonary hypertension [77]. An increased IVRT may be easily appreciated by simultaneous sampling of both tricuspid and mitral valve flows in which the tricuspid valve shows delayed onset of its E-wave, indicating an increased right ventricular isovolumetric relaxation time. LV: left ventricle; RV: right ventricle.

Contrast-enhanced MR angiography can be helpful in excluding pulmonary arterial and venous pathologies that may be responsible for pulmonary hypertension. When pulmonary veno-occlusive disease is suspected, contrast-enhanced CT is preferred to MR [78]. Diffusely increased interstitial markings in the peripheral lung field with ground-glass appearance of the lung parenchyma in the
presence of patent central pulmonary veins are diagnostic of this rare disease.

Finally, it is advised not to do breath-holding maneuvers during MR or CT as ensuing hypoxia and increase in intrathoracic pressure can trigger a pulmonary hypertensive crisis.

Summary and future perspective

MR is a potent tool for the evaluation of the pulmonary circulation. In addition to detailed anatomical images of every level of the pulmonary arteries, veins, and airways, a great variety of functional information can be obtained. This includes flow volumes, velocities, and patterns, and right ventricular size and function. The course of flowing blood can be traced with time-resolved MR angiography, and oxygen content can be measured.

Stress imaging, using either MR-compatible supine ergometers or drugs such as dobutamine or adenosine, is now available to assess ventricular function and flow volumes not only at rest, but also in an active state, mimicking more closely the demands of everyday life. With the advent of blood pool contrast agents, higher spatial resolution will be possible and help with the depiction of small peripheral vessels [79]. Small and medium size arteriovenous malformations, collaterals and abnormal arborization patterns seen in patients with pulmonary hypertension and pulmonary atresia with ventricular septal defect will be shown in greater detail. Using the same innovation, serial acquisitions will reveal the effect of breathing, exercise, or pharmacological stimuli [79]. Time-resolved 4-dimensional velocity mapping has started to give new insights into streaming in pulmonary regurgitation, bidirectional cavopulmonary shunt, and the Fontan circuit [80]. Ventilation scans, using hyperpolarized noble gases such as 3-Helium or 129-Xenon, reveal mismatches with perfusion which are common in stenosed pulmonary arteries or veins as well as in pulmonary hypertension and arteriovenous malformations [81].

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CHAPTER 13

Valvular heart disease

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Introduction

Quantification of valvular function is important for the clinical management of valvular heart disease. Although valvular stenosis may be evaluated by echocardiography, it does not provide consistently reliable and accurate quantification of valvular regurgitation. Thus, therapeutic decisions are mainly based on symptoms and indirect echocardiographically derived measurements of the valvular regurgitation. Cardiac magnetic resonance imaging (CMR) does have the capability of precisely quantifying valvular regurgitation. Moreover, it is highly accurate and reproducible in measurement of ventricular function and volumes. For pediatric patients, its capability for assessing pulmonary regurgitation as well as right ventricular volumes and function is especially attractive.

Clinical status is often misleading in valvular disease, as symptoms appear only in the advanced stages, when irreversible ventricular damage might have already occurred. Predicting when to operate on an incompetent valve is one of the most difficult decisions in cardiology. CMR quantification of valvular function together with ventricular burden are critical steps for guiding this decision.

CMR techniques

Valvular function can be measured using breath-hold and free breathing sequences. Breath-holding requires co-operation, which is not possible in infants and many children. Recently, a navigator-gated technique was introduced as an alternative, in which breathing motion can be monitored by exciting a small column of tissue perpendicular to the diaphragm. With use of an edge-detection algorithm, the position of diaphragm–lung interface is determined and data are included for reconstruction only from selected diaphragm positions during the respiration. These navigator echoes are usually interleaved with the cine sequence. Use of navigator technique effectively limits motion artifact, but the drawback is longer acquisition and the total examination time. Alternatively, multiple excitations can be used to “average” out respiratory motion, again at the expense of increased scan time.

Spin echo sequences (black-blood sequences)

Spin echo and its newer modification, double-inversion fast (or turbo) spin echo, utilize pulse sequences, in which flowing blood appears black and more stationary tissue produce MR signals displayed in shades of grey and white. Spin echo sequences produce a single image with high tissue contrast and spatial resolution. Therefore, this imaging is used only for assessment of valvular morphology and consequently has only a minor role in evaluating valvular heart disease. Gradient echo pulse sequences have mostly replaced spin echo imaging for the evaluation of valvular heart disease.

Gradient echo and balanced steady state precession cine sequences (white-blood sequences)

Gradient echo sequences utilize rapid repetition of radiofrequency pulses resulting in partial satura-
tion of stationary tissues of the slice, but blood that flows to the slice contains unsaturated spins. Therefore, stationary tissue generates weak signal (displayed grey) and flowing blood strong signal (displayed bright). Another important feature of gradient echo sequences is high imaging speed allowing cine display. Depending on the heart rate, 16–30 phases per cardiac cycle are acquired to generate cine loop. As a general rule, a temporal resolution of 50 ms is usually accurate enough for clinical purposes (lower limits of what is needed) in older patients but needs to be much lower in infants with high heart rates.

Signal loss can be seen in gradient echo images due to intravoxel dephasing in cases of high velocity jets due to valvular stenosis and regurgitation. The area of signal loss (void) is only a rough semi-quantitative measure of valvular regurgitation, which is dependent on a number of variables of which the most important is echo delay time (TE). The area of the signal loss cannot be used to estimate the degree of stenosis. It is necessary to know the details of the applied sequence when reading studies, because technical parameters of pulse sequence such as TE, TR, voxel size and flip angle, and plane selection influence the size of signal void. Moreover, it is affected also by hemodynamic parameters and the area or shape of jet origin.

Steady-state free precession (SSFP) pulse sequences have replaced gradient echo cine imaging in the past 10 years. It has better signal-to-noise ratio and contrast between blood and myocardium compared to conventional gradient echo sequences. Commercially different acronyms are used for them such as balanced FFE [fast field echo, Philips Medical Systems, Best, The Netherlands], FIESTA [Fast Imaging Employing Steady-state Acquisition, GE Medical Systems, Milwaukee, Wis] or true FISP [fast imaging with steady-state precession, Siemens Medical Systems, Iselin, NJ]. They are less sensitive to turbulence-related signal loss. However, flow artifacts may exist near regions of rapid blood flow acceleration and problems occur especially in infants and young children since they have significantly higher blood acceleration in great vessels compared to adults. Another consideration for SSFP sequences is that from some manufacturers, a reduction in field of view (FOV) or slice thickness can greatly prolong TE and TR leading to degradation of image quality. Larger FOV and matrix can be used to avoid these problems.

**Parallel imaging**

Parallel imaging is used mainly in conjunction with many sequences to reduce scanning and breath-hold time by a factor of 2 or more. Parallel imaging has been used also with velocity mapping, and excellent correlations and slopes even with accelerator factors of 2, 3 and 4 have been found: $r = 0.96$, $r = 0.94$ and $r = 0.90$ and all slopes $y = 0.99$, respectively [1]. Different acronyms for parallel imaging are used depending on manufacturer: SENSE in Philips, ASSET in General electric and IPAT in Siemens.

**Velocity mapping**

Velocity mapping (also known as velocity-encoded cine MRI [VEC-MRI] and phase contrast MRI [PC-MRI]) is based on the principle of phase shift between moving and stationary tissue. Hydrogen protons moving along a magnetic field gradient acquire a phase shift relative to stationary protons. The phase shift is directly proportional to the velocity of moving protons and can be transformed to voxel velocity at any point of cardiac cycle. The uncontrolled phase errors must be removed to detect only the velocity phase shift, so typically two scans, a reference and velocity-encoded (sensitized) are acquired together. Velocity map is generated by subtracting the reference phase image from the velocity-encoded phase image.

VEC-MRI provides accurate results in the presence of laminar flow but is detrimentally affected by turbulent flow patterns innate in valvular diseases. Flow-encoding directions are typically restricted. For volumetric flow, imaging plane and encoding direction should be perpendicular to target vessel. However, when the angle differs slightly from perpendicular to the vessel, through-plane velocity is underestimated, but is mainly compensated by the larger region of interest (ROI) area. Through-plane velocity method is used to determine velocity mapping flow data. Instantaneous flow in ml/s during heart cycle is calculated by drawing a ROI around the vessel and integrating the velocity over the vessel area for each
time frame. It has been recommended as small an ROI as possible be used to enclose the vessel [2]. Previous calculations have shown that at least 16 voxels must cover the cross-section of lumen of the vessel to keep the error within limits of 10% [3,4]. A flow curve is displayed as the flow data versus time. From a flow curve of the proximal ascending aorta stroke volume (SV) and cardiac output can be measured.

VEC method can be applied for estimation of severity of stenosis. In-plane VEC method using a thin slice, preferably narrower than the jet, is preferred to identify peak velocity. Typically, several in-plane and possibly through-plane VEC images are obtained to attempt peak velocity detection. Underestimation of peak velocity is to be expected because of limitation of temporal resolution and slice thickness. To quantify velocity through a stenotic valve, it is necessary to adjust imaging parameters so that signal is present in the jet core. It requires a very short TE to prevent signal loss. Velocity-sensitivity scale should be set above the expected peak velocity to avoid phase-wrap. Phase-wrap artifact is usually discernible due to its sharp border between wrapped and non-wrapped pixels in velocity mapping image. The modified Bernoulli formula is employed for calculation of a pressure gradient based on the peak velocity (peak pressure gradient = 4*(peak velocity)^2). Slight error in angulation of the imaging plane in relation to flow direction does not usually hamper the measurement of peak velocity, because the velocity remains usually constant over a distance of at least five times the jet diameter [5].

By adjusting the sequence settings, the error relating to variables such as temporal resolution, velocity encoded sensitivity, partial volume effect, signal loss and ghosting can be changed. The measurement may be affected also by concomitant gradients or by cardiac motion. Some errors are intrinsic to the method and anatomy and cannot be fixed by using currently available technology. However, repeatability of velocity mapping studies is superior to the most other clinical measurements of flow volume. Optimization of VEC-MRI sequence has recently discussed in detail by article of Kilner and colleagues [6]. This technique has been validated in vitro using flow phantoms, and correlation of 0.998 has been found between weighed flow volume and CMR derived flow volume [7]. It provides the most accurate information on cardiac output and shunts [8,9]. This method is considered as the gold standard for measurements of regurgitation of pulmonary, tricuspid and aortic valves, and the gradient of pulmonary stenosis [10–14]. This particular MR technique has been available at some level for over 20 years but is probably still under-utilized in clinical pediatric cardiology.

**Principles of quantification**

Regurgitant volume can be determined by comparing left and right ventricular stroke volumes when there is a single regurgitant valve and no intracardiac or extracardiac shunt co-exist. This measurement from CMR volumetric data is based on the physiological fact of almost equal outputs of left and right ventricle. The difference in left and right ventricular stroke volumes by volumetric technique. These calculations are demonstrated by several examples in Table 13.1. In patients with combined aortic and mitral regurgitation, the volumetric method assesses only total regurgitation. Assessment of left sided regurgitation is underestimated in the case of a concomitant regurgitant valve on the right side by utilizing the volumetric approach. Aortic and pulmonary valve regurgitation can also be measured directly using VEC MRI. Regurgitant fraction (%) is determined by dividing reverse flow, which represents regurgitant volume, with systolic forward flow. Regurgitant fraction is generally the primary parameter used in assessing severity of valvular regurgitation. Direct velocity mapping of atrioventricular valve regurgitation may be problematic. Therefore, the combination of ventricular volumetric stroke volume and great vessel velocity
Table 13.1 Examples of determination of regurgitant volume and fraction using two CMR techniques, volumetric (VOL) and velocity mapping (VEC).

<table>
<thead>
<tr>
<th>Single valve affected</th>
<th>Regurgitant volume (ml)</th>
<th>Regurgitant fraction (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe pulmonary regurgitation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alternative #1</td>
<td>VOL RV stroke volume 211ml – VOL LV stroke volume 108ml</td>
<td>103ml</td>
</tr>
<tr>
<td>Alternative #2</td>
<td>VEC PA forward flow 215ml – VEC PA backward flow 98ml</td>
<td>98ml</td>
</tr>
<tr>
<td>Alternative #3</td>
<td>VEC PA forward flow 215ml – VEC AO stroke volume 107ml</td>
<td>108ml</td>
</tr>
<tr>
<td>Mean regurgitation volume, mean regurgitation fraction for example, #1 and #2</td>
<td></td>
<td>101ml</td>
</tr>
<tr>
<td>Moderate-severe tricuspid regurgitation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alternative #1</td>
<td>VOL RV stroke volume 160ml – VOL LV stroke volume 104ml</td>
<td>56ml</td>
</tr>
<tr>
<td>Alternative #2</td>
<td>VOL RV stroke volume 160ml – VEC PA forward flow 96ml</td>
<td>64ml</td>
</tr>
<tr>
<td>Alternative #3</td>
<td>VEC TV forward flow 172ml – VEC TV backward flow 66ml</td>
<td>66ml</td>
</tr>
<tr>
<td>Alternative #4</td>
<td>VEC TV forward flow 172ml – VEC PA stroke volume 96ml</td>
<td>76ml</td>
</tr>
<tr>
<td>Mild-moderate aortic regurgitation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alternative #1</td>
<td>VOL LV stroke volume 96ml – VOL RV stroke volume 79ml</td>
<td>17ml</td>
</tr>
<tr>
<td>Alternative #2</td>
<td>VEC AO forward flow 98ml – VEC AO backward flow 16ml</td>
<td>16ml</td>
</tr>
<tr>
<td>Alternative #3</td>
<td>VEC AO forward flow 98ml – VEC PA stroke volume 80ml</td>
<td>18ml</td>
</tr>
<tr>
<td>Severe mitral regurgitation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alternative #1</td>
<td>VOL LV stroke volume 180ml – VOL RV stroke volume 89ml</td>
<td>91ml</td>
</tr>
<tr>
<td>Alternative #2</td>
<td>VOL LV stroke volume 180ml – VEC AO stroke volume 86ml</td>
<td>94ml</td>
</tr>
<tr>
<td>Alternative #3</td>
<td>VEC MV forward flow 200ml – VEC MV backward flow 94ml</td>
<td>106ml</td>
</tr>
<tr>
<td>Alternative #4</td>
<td>VEC MV forward flow 200ml – VEC AO stroke volume 86ml</td>
<td>114ml</td>
</tr>
<tr>
<td>Severe mitral regurgitation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alternative #1</td>
<td>VOL LV stroke volume 150ml – VEC PA stroke volume 90ml</td>
<td>60ml</td>
</tr>
<tr>
<td>Moderate-severe mitral regurgitation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alternative #1</td>
<td>VOL LV stroke volume 130ml – VEC AO forwarded volume 90ml</td>
<td>40ml</td>
</tr>
<tr>
<td>Severe mitral regurgitation</td>
<td></td>
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<tr>
<td>Alternative #1</td>
<td>VEC AO forward flow 160ml – VEC AO backward flow 40ml</td>
<td>40ml</td>
</tr>
<tr>
<td>Alternative #2</td>
<td>VEC MV forward flow 160ml – VOL RV stroke volume 110ml</td>
<td>50ml</td>
</tr>
<tr>
<td>Moderate aortic regurgitation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alternative #1</td>
<td>VOL LV stroke volume 200ml – VOL RV stroke volume 110ml</td>
<td>–</td>
</tr>
</tbody>
</table>

nary insufficiency, systolic forward flow in the pulmonary artery should equal right ventricular stroke volume. Of course, the sum of the flows in the branch pulmonary arteries should equal flow in the main pulmonary artery. In addition, in the absence of intracardiac shunts, net pulmonary blood flow should equal net aortic flow. Numbers will not match exactly, however, if they come within 10%, this is generally considered adequate.

Although no clear consensus exists on grading of pulmonary stenosis, some papers suggest that 20–50 mmHg peak gradient is considered mild, 50–75 mmHg moderate and >75 mmHg severe [16]. Our laboratory considers severity of valvular regurgitation as follows: mild – less than 20%; moderate – 20 to 40%; severe – >40%. The following grading has been proposed for aortic and mitral valve regurgitation severity: mild ≤15%, moderate >15–25%, severe to moderate 26–48% and severe >48% [17]. Corresponding values for regurgitant volumes are <30 ml/beat, 30–59 ml/beat and >60 ml/beat, when excluding the category of mild regurgitation. Recommendation of grading of aortic stenosis with classes of mild, moderate and severe stenosis by mean gradient <25 mmHg, 25–40 mmHg and >40 mmHg respectively, and by valvular opening area >1.5 cm², 1.0–1.5 cm² and <1.0 cm² respectively, have been proposed. Valve opening area index <0.6 cm²/m² is considered severe [18]. The mitral valve is considered stenotic when valve area is less than 2.0 cm² and graded severe if less than 1.0 cm².

**Pulmonary valve**

Pulmonary valve regurgitation or stenosis is observed by a diastolic or systolic signal void, respectively. Standard short axis cine CMR is used to determine RV volumetrics and ejection fraction. VEC-MRI is applied for measurement of pulmonary valve regurgitant fraction or valvular gradient. As a general rule, a plane 2 cm distal to pulmonary valve is recommended, when estimating pulmonary valve regurgitation by VEC-MRI in older children.

**Pulmonary valve stenosis**

Pulmonary stenosis is divided to three categories: (1) subvalvular; (2) valvular; and (3) supravalvular stenosis. The valvular type is the most common. Pulmonary stenosis may be part of several systemic syndromes such as Williams, Alagille, DiGeorge or Noonan’s.

**Pulmonary regurgitation**

Clinically significant pulmonary valve regurgitation is common in patients with a previous pulmonary valve operation, especially in those with tetralogy of Fallot (TOF). TOF consists of pulmonary valve stenosis (right ventricular outflow tract obstruction), right ventricular hypertrophy, ventricular septal defect and over-riding aorta. Obstruction can occur as one or more levels including supravalvular, valvular, annular and infundibular level. It is not uncommon for infundibular, annular, and valvular obstructions to coexist.

CMR is the method of choice for post-operative monitoring of patients after correction of TOF. Pulmonary regurgitation is the most common post-operative complication. Pulmonary regurgitation has become very frequent in survivors of repair of TOF (Figures 13.1 and 13.2) and other operative and transcatheter interventions on the pulmonary valve.

CMR has a leading role in the assessment, quantification and follow-up of patients with pulmonary regurgitation, as well as in determination of residual stenosis after interventional procedures for pulmonary valve stenosis. The CMR method has been validated for the assessment of severity of pulmonary regurgitation [19].

Some authors have recommended early pulmonary valve replacement to prevent irreversible right ventricular dysfunction, when right ventricular total ejection fraction decreases below 40%, right ventricular systolic pressure exceeds half of the left ventricular systolic pressure or right ventricular volume index increases >150 ml/m² [20,21]. The following volume limits have been proposed for pulmonary valve replacement: right ventricular end-diastolic volume index >160 ml/m² or end-systolic volume index >82 ml/m² [22]. Other authors have suggested that pulmonary valve replacement should be performed before right ventricular end-diastolic volume index reaches 170 ml/m² or end-systolic volume index reaches
Additional post-operative complications such as aneurysm of the right ventricular outflow tract, functionally abnormal tissue with fibrosis in the outflow tract or peripheral stenosis of the branch pulmonary arteries (which may affect the amount of PR) can be detected by CMR. The CMR method is recommended as the primary method for post-operative evaluation of TOF.

85 ml/m² [23]. A right ventricular end-systolic volume index greater than 95 ml/m² was always associated with a reduction of RV total EF to below 40% and effective EF to below 25% [24]. Effective EF is calculated as follows: $\text{EF}_{\text{eff}} = (\text{RVSV} - \text{pulmonary regurgitant volume})/\text{RV EDV}$. As mentioned elsewhere in this textbook, the use of $\text{EF}_{\text{eff}}$ is controversial.

Figure 13.1 Follow-up study after previous operation due to tetralogy of Fallot. SSFP sequence showed marked right ventricular dilation, hypertrophy and decreased ejection fraction. Four chamber (upper) and short axis (middle) images are presented in diastole (left) and systole (middle). Volumetric analysis showed severe right ventricular enlargement with end-diastolic volume of 213 ml and ejection fraction of 41%. Focal left pulmonary artery stenosis with a diameter of 1.1 cm is present in the axial SSFP image (right upper image). Dilation of main and right pulmonary artery was detected. Magnitude and phase images (bottom left and right respectively) and VEC-MRI derived flow curve (middle right) from same patient with pulmonary valve regurgitation. Selective VEC-MRI from pulmonary branches indicated that main pulmonary artery shared 66% of its flow to the right and 34% to the left.
Cardiac MR of congenital and acquired pediatric heart disease

Tricuspid valve

Cine CMR in the four chamber orientation is the most useful sequence in the evaluation of the tricuspid valve. Tricuspid regurgitation is quantified either by (a) the difference of right ventricular stroke volume calculated by the volumetric method and forwarded flow in pulmonary artery in VEC-MRI or (b) by using VEC-MRI alone in short axis orientation of tricuspid annulus. Tricuspid stenosis is easily identified in four chamber cine-CMR. Planimetric measurement of tricuspid valve area can, at times, be problematic. Short axis cine CMR is used to assess related right ventricular and right atrial involvement in tricuspid valve disease. Long axis cine (RV two chamber plane), can be used to demonstrate the length of atrialized portion of the RV in Ebstein anomaly (Figure 13.3).

The tricuspid valve consists of anterior, posterior and septal leaflets. The tricuspid annulus is weaker when compared to mitral annulus and therefore dilates more easily with pressure or volume overload. Congenital tricuspid valve regurgitation might be found whenever pulmonary artery pressure or right ventricular pressure is elevated. Congenital tricuspid valve defects include tricuspid stenosis and atresia, and Ebstein anomaly. Infective endocarditis can cause tricuspid pathology and is especially found in patients with history of intravenous drug abuse.

Perhaps the most frequent case of tricuspid regurgitation is secondary to pulmonary hypertension. It can be easily diagnosed from tricuspid regurgitation velocity achieved by VEC MRI. Right ventricular to atrial pressure gradient (RVAPG) is estimated by using modified Bernoulli equation. Pulmonary artery pressure can be estimated (when there is no pulmonary artery stenosis) by adding RVAPG to estimation of right atrial pressure.

Tricuspid atresia and stenosis

Tricuspid atresia is always associated with some level of right ventricular hypoplasia. Tricuspid valve stenosis or hypoplasia may exist with pulmonary valve atresia with intact septum. CMR may be critical in determining RV volume in order to decide upon the appropriate surgical approach.

Tricuspid regurgitation

Ebstein anomaly of the tricuspid valve is an uncommon congenital heart defect that occurs in approximately 0.5–1.0% of congenital heart disease. In Ebstein anomaly, septal and possibly the posterior leaflet of tricuspid valve is partly fixed on right
ventricular wall resulting in a more apical position of the functional valve plane. It leads to enlargement of the right atrium and corresponding decrease in right ventricular volume. Hemodynamic consequences are related to grade of tricuspid regurgitation, to the small size of remaining functional right ventricle and its outflow, and to the subsequent degree of shunting through the atrial septum.

A number of possible methods exist for surgical correction of Ebstein anomaly. The type of repair depends on the extent of disease. Therefore, CMR is useful in determining leaflet size and location, annulus and atrial size, and right ventricular function. CMR currently provides details about anatomy that echocardiography cannot and is therefore recommended in evaluation of Ebstein anomaly [25,26].

**Aortic valve**

Short-axis cine CMR is used to visualize aortic valve morphology and to assess ventricular burden in aortic valve disease. Recently, it has been shown that CMR could detect all 14 bicuspid valves identified with transesophageal echocardiography, of which 56% were missed with transthoracic echocardiography [27]. Cine CMR in three-to-five chamber orientations offers the easy way for visual observation of aortic regurgitation and stenosis. Aortic regurgitation can be measured directly using VEC-MRI, which should be performed in a plane as close as possible to the aortic valve below the coronary ostia [28]. Others have advocated a plane at the level of the sino-tubular junction. Regurgitation can be measured alternatively by differences in left and right ventricular stroke volumes using the volumetric method when no atrioventricular valve insufficiency or intracardiac shunts are present. Regarding the plane selection in an aortic valve study, coronary flow in systole is missed when a plane distal to coronary orifice is used, and coronary flow in diastole is missed by proximal plane. Aortic stenosis can be graded using valve area planimetry or using measures based on stenosis related acceleration in blood flow velocity. Short-axis cine CMR is optimal for direct aortic valve planimetry, which is more reproducible with CMR than transesophageal echocardiography [27]. Peak velocity could be transformed to transvalvular gradient (pressure drop) using modified Bernoulli equation ($P_{\text{gradient}} = 4v^2$), where $P$ is pressure in millimeters of mercury (mmHg) and $v$ is velocity in meters per second. Aortic valve area can be measured also indirectly using continuity equation and is recommended when left
ventricular systolic function is diminished. It is derived from the following formula: \( \text{Ao}_{\text{area}} = (\text{LVOT}_{\text{area}})^2 / (\text{LVOT}_{\text{flow}}) / (\text{Ao}_{\text{flow}}) \), in which \( \text{Ao}_{\text{area}} \) and \( \text{Ao}_{\text{flow}} \) are aortic valve area and flow, and \( \text{LVOT}_{\text{area}} \) and \( \text{LVOT}_{\text{flow}} \) are left ventricular outflow tract area and flow, respectively. It requires use of two additional velocity mapping sequences, one from the left ventricular outflow tract and another from just beyond the aortic valve plane. Although aortic valve area measured by planimetry and the continuity equation correlates well together, continuity equation results in about 15% overestimation of the stenosis severity [29], which is related to continuity equation, but not to the imaging method itself. Previous studies have shown a close agreement (\( y = 0.97x + 0.5; r = 0.97 \)) between velocity mapping assessment of aortic pressure gradient by comparing them to invasive technique [30]. In the pediatric population, the valvular opening area should be indexed to body surface area (BSA).

The normal aortic valve has three cusps – left, right and non-coronary cusps – according to their relation to normal orifices of coronary arteries. Bicuspid aortic valve (Figures 13.4 and 13.5) can be functional, consisting of some degree of fusion in two of the cusps, or a morphological two-leaflet valve. Bicuspid aortic valve can result in either stenosis, regurgitation or both and can be associated with ascending aortic dilation. Treatment for moderate to severe congenital aortic stenosis is val-

![Figure 13.4](image1.png)

**Figure 13.4** 39-year-old male with bicuspid aortic valve and annuloaortic ectasia. Bicuspid aortic valve in short axis image using an SSFP sequence, (left); signal void due aortic regurgitation using same sequence in axial orientation (middle) and maximal diameter of 58 mm in ascending aorta (right) indicating severe dilation using spin echo sequence in sagittal orientation. Furthermore, cine images showed fusion of the left and right coronary cusps and a regurgitant fraction was 33% indicating moderate-to-severe regurgitation.

![Figure 13.5](image2.png)

**Figure 13.5** 14-year-old female was studied in follow-up for a bicuspid aortic valve with mild aortic regurgitation, aortic stenosis, and dilated ascending aorta. Aortic diameter of 38 mm in coronal spin-echo image (left), bicuspid aortic valve with a fusion between the right and non-coronary cusps (middle) and axial VEC-MRI image (right) at the level of right pulmonary artery showed a moderate aortic stenosis with a peak pressure gradient of 49 mmHg and mild aortic regurgitation with a regurgitant fraction of 10% (8 ml). Normal biventricular volume and function was observed by volumetric analysis.
votomy or valvuloplasty, which results usually in increased valvular regurgitation. Aortic regurgitation, annuloaortic ectasia, and or aneurysm of sinus valsalva are associated with Marfan syndrome.

**Aortic stenosis**

Generally, aortic valve stenosis develops slowly due to the calcification process. However, marked calcification can be rarely seen in the early course of bicuspid aortic valve. It takes usually several years before it becomes symptomatic. Congenital aortic stenosis is the 9th commonest CHD representing approximately 4% of CHD. In contrast to valvular aortic stenosis, the other two forms of aortic stenosis are subvalvular (Figure 13.6) and supravalvular stenosis. Hypertrophic obstructive cardiomyopathy can be considered a form of subvalvular aortic stenosis, in which systolic anterior movement (SAM) of the anterior mitral leaflet results in mitral regurgitation. However, SAM is not present in membrane-like subvalvular aortic stenosis, which is usually characterized by left atrial enlargement and severe left ventricular hypertrophy. Supravalvular aortic stenosis is the least common form of aortic stenosis and can be found as an isolated defect or as a part of Williams syndrome.

CMR has been useful in the evaluation of pulmonary homograft stenosis after Ross procedure, in which a patient’s diseased aortic valve is replaced with his or her own pulmonary valve [31] (autograft). CMR is recommended, because late complications such as aortic insufficiency, right ventricular outlet obstruction, aortic autograft dilatation, supravalvular aortic stenosis and pulmonary allograft stenosis are difficult to evaluate comprehensively using echocardiography. Ross procedure related aortic root dysfunction and its effect on left ventricular function have also been recently evaluated by CMR [32].

**Aortic regurgitation**

Etiology of aortic regurgitation includes those of aortic stenosis and the diseases leading to dilation of the ascending aorta including Marfan syndrome, Ehlers-Danlos and Loey-Dietz syndromes, relapsing polychondritis, ankylosing spondylitis and various forms of annuloaortic ectasia. Severe aortic regurgitation is present in about 10% of cases with aortic dissection. Chronic aortic regurgitation leads to increased left ventricular volume. Left ventricular mass is increased, although left ventricular wall thickness may appear normal. Ejection fraction remains normal for many years, but when decreased, prognosis is poor. CMR has been found useful to measure aortic regurgitation by comparison with both invasive and non-invasive techniques \( r = 0.97 \) [33]. Aortic regurgitant volume and regurgitant fraction assessment by VEC-MRI

![Figure 13.6](image_url) Follow-up CMR was performed after surgical treatment for coarctation of the aorta and aortic valve stenosis in a morphologically bicuspid aortic valve. The left ventricular outflow tract seems normal in axial SSFP image in diastole (left), but the systolic image (middle) shows a signal void from the outflow tract and valve area. There was a subaortic membrane producing a 36 mmHg gradient and the anterior mitral leaflet was normal. VEC-MRI (right) shows turbulence in outflow tract. Furthermore, pre- and post-coarctation flow measurements using VEC-MRI indicated 13% collateral flow in the thorax.
has high interstudy reproducibility \( (r = 0.98 \text{ and } r = 0.99) \) and close correlation \( (r = 0.98 \text{ and } r = 0.99) \) to the corresponding assessment by volumetric CMR \([14]\). There are significant variations in absolute values of regurgitation fraction between VEC MRI and echocardiography. Therefore, reference values for all parameters are not interchangeable between different methods \([34]\).

**Mitral valve**

Cine CMR in the four chamber orientation is used for evaluation of the mitral valve, but short axis images may be useful, too. Mitral regurgitation is quantified either by the difference of left ventricular volumetric stroke volume and forwarded flow in aorta in VEC-MRI (in the absence of intracardiac shunts) or by using VEC-MRI alone in short axis orientation of mitral annulus. The accuracy rate of CMR derived mitral regurgitation assessment was reported to be 91% and 94%, respectively. Other studies have shown a close correlation \( (r = 0.96) \) between VEC-MRI measurement of mitral regurgitation fraction and that by using ultimate invasive measures incorporating Fick, thermodilution and ventriculography \([35]\). Suspicion of mitral stenosis is easily identified in four chamber cine-CMR and can be quantified correspondingly to direct measurement of mitral regurgitation by VEC-MRI. CMR with short axis orientation can be applied to quantify a valve area for the mitral valve, a morphological measure of the severity of mitral stenosis. Short axis cine CMR is used to assess related left ventricular and atrial involvement in mitral valve disease. Accuracy rate of peak velocity across a stenotic mitral valve was found to be over 87% by CMR. Another study indicated close agreement between transmitral inflow velocities \( (r = 0.90) \) and stenotic mitral valve area \( (r = 0.94) \) measurements between CMR and echocardiography \([36]\).

The mitral valve consists of two leaflets with the shorter but broader posterior leaflet and the longer anterior leaflet. The posterior leaflet can be divided to segments according to ridges P1-P3. The anterior leaflet is smooth and an A1-A3 nomenclature is used according to its coaptation surface relation to posterior leaflet’s segments. Normal maximal opening area of mitral valve is 2.0–5.3 cm²/m² in mixed and populations using different modalities. Reference values have been published for children \([37]\). Up to 20–50% of patients with marked mitral valve disease develop atrial fibrillation, which usually hampers conventional CMR imaging, since it relies on electrocardiographic gating. However, recent upgrades in MR technology have made real time CMR a practical and reliable tool to overcome this problem.

**Mitral stenosis**

Congenital mitral stenosis is rare, and is observed usually in infants and children as a part of complex congenital heart disease, for example left ventricular hypoplasia or univentricular heart. In general, mitral valve stenosis leads to increased left atrial pressure, which is reflected in increased pulmonary arteriolar resistance resulting in precapillary pulmonary hypertension. In advanced stages of the disease, right ventricular hypertrophy might be present together with increased pulmonary artery pressure. However, changes in the right heart may differ depending on the presence of atrial shunt.

**Mitral regurgitation**

In general, acute mitral regurgitation can be differentiated from chronic mitral regurgitation by small left atrial and ventricular volumes, low cardiac output and the presence of pulmonary congestion. In these cases, infective endocarditis, papillary muscle or chordal rupture or ischemic posterolateral wall motion abnormality together with restriction in the posterior leaflet of the mitral valve should be suspected. Mitral valve prolapse and myxomatous degeneration of mitral valve leaflets are the most common causes of primary chronic mitral regurgitation. Mitral regurgitation is also present in congenital atroventricular septal defect with a cleft in the anterior leaflet. Secondary forms of chronic mitral regurgitation are more common than the primary ones. Dilated cardiomyopathy, ischemic heart disease, hypertension and atrial fibrillation are the most common reasons for mitral annular dilatation leading to secondary mitral regurgitation. Left atrial dilatation prevents an increase of left atrial pressure and is considered an adaptative change in chronic mitral regurgitation.
Polyvalvular disease

Determination of the volume of valvular regurgitation in polyvalvular disease can be performed by using a combination of difference in stroke volume of the two ventricles and measurements of inflow and outflow of the ventricles, and flows in the aorta and in the pulmonary artery. Difference in stroke volume of the two ventricles gives a total volume of regurgitation on one side of the heart. The stroke volume is calculated from cine CMR volumetrics. Retrograde flow in either the aorta or the pulmonary artery can be used to determine the volume of semilunar valve regurgitation to the volume of atrioventricular valve regurgitation. The effect of each valve lesion on cardiac function can be measured by direct VEC measurement or by using the calculations described in the section on Principles of quantification or in Table 13.1. Intracardiac shunting complicates matters in apportioning the contribution of each valve on the burden of disease.

Prosthetic cardiac valves

All prosthetic valves can be studied safely using CMR at 1.5 T, because no substantial heating or magnetic interaction is present [38]. Using higher strengths of magnetic fields at least one artificial valve was considered unsafe. Therefore, it is recommended that MRI compatibility be checked from the website dealing with MRI safety issues (MRIsafety.com). Signal loss and artifacts are produced by metallic valve components. Regurgitation volume and regurgitant fraction can be measured from planes proximal to prosthetic valve by using VEC MRI with valve tracking techniques. CMR research has produced important physiologic data of mechanical valve function. It has been found that the blood volume required for leaflet closure is significantly higher in bileaflet mechanical valve compared to native valves [39]. CMR has been proved to offer additional value beyond the standard measurements in a pediatric patient with prosthetic valve dysfunction [40].

Endocarditis

Endocarditis is unusual in pediatric compared to adult patients and is usually related to a congenital valve abnormality. Endocarditis with medium to large vegetations can be demonstrated by CMR (Figure 13.7), but transesophageal echocardiography is considered to be the premier method. CMR might be useful when evaluating perivalvular changes associated with infective endocarditis such as perivalvular abscess.

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CHAPTER 14

Imaging coronary arteries in children

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Introduction

Clinical coronary artery imaging using magnetic resonance (MR) started in the early 1990s [1–3]. The focus has been to develop magnetic resonance coronary artery imaging (MRCA) as a non-invasive imaging tool as part of the diagnostic armamentarium for adult ischemic heart disease. This has proven to be technically challenging in the adult population. Nonetheless, many centers have reported non-invasive MRCA commanding a diagnostic role in anomalous origin of the coronary arteries [4,5], native coronary artery disease [6], coronary bypass graft patency [7], evaluation of the coronaries after other surgical procedures such as the Ross operation and the arterial switch operation for transposition of the great arteries and Kawasaki disease [8,9]. Surgical training and pre-operative planning tools could also benefit from good delineation of the coronary artery tree for precise incision planning [10,11]. The recent rapid development of cardiac computed tomography (CT) with multi-detector row CT (MDCT) scanners has increased the use of this modality despite concerns for radiation exposure [12]. Although cardiac catheterization still remains the gold standard, it is invasive requiring ionizing radiation, iodinated hyperosmolar intravascular contrast agents and is quite often performed under general anesthesia [16]. It offers only imaging of the lumen and lacks 3D information of the coronary arteries and the surrounding vessels and heart chambers. Echocardiography is not ideal given the limitations of dependence of available acoustic window and the limited visualization of only the most proximal extent of the coronary arteries [17]. In addition, false negatives have been noted, for example, with echocardiographic determination of anomalous coronary arteries. Cardiac CT with ECG gating also requires intravenous administration of iodinated contrast agent and ionizing radiation. Many
patients with congenital heart disease may well require repeated non-invasive imaging throughout their childhood if not into adulthood. The cumulative radiation dose from multiple cardiac MDCT examinations can be hazardous unless there is continued improvement in the techniques for dose reduction [18–20]. Moreover, the high heart rates of an infant or a young child can be a major challenge to the current MDCT scanners [21,22]. Therefore, MR can be the optimal, reproducible [23], non-invasive imaging method in the pediatric population for visualization of the blood pool and surrounding tissue of the heart, great vessels and the coronary arteries. Additionally to imaging the lumen, the possibility of visualization of the vessel wall may offer future options regarding risk stratification and monitoring of different treatment methods [24,25]. This chapter will first discuss the MR techniques involved in coronary artery imaging followed by clinical examples.

Techniques for magnetic resonance coronary angiography (MRCA)

There are certain boundary conditions that have hampered the clinical success of MRCA and coronary vessel wall imaging. These include the small diameter [26] and wall thickness [24,27] of the coronary artery vessels, their highly tortuous pathway, cardiac and respiratory motion, and the proximity of the coronary arteries to epicardial fat and myocardium. Recent advances in both hardware and software, which will be discussed in this section, allow for consistent visualization of the proximal and mid portions of the native coronary arteries even in children.

Advanced motion compensation strategies to meet the challenges of coronary vessel wall imaging have been developed, allowing more consistent visualization of the proximal and mid portions of the native coronary vessel walls.

Cardiac motion

Due to the intrinsic motion of the heart, synchronization of data acquisition to the ECG is an absolute requirement for coronary artery imaging. Coronary artery motion is minimal during end-systole and mid-diastole [28]. Even during these quiescent periods the rest period can be less than 50 ms depending on the patient’s heart rate [28,29]. In infants and young children with very high heart rates, there is no sufficiently long rest period during diastole to permit artifact-free imaging. Mid-diastole is a period of rapid coronary flow thus enhancing the signal from coronary blood between successive RF pulses. Thus the trigger delay is set to end-systole for these patients with high heart rates. The trigger delay should be optimized specific for each patient by acquiring a high temporal resolution cine scan in the 4-chamber orientation [28,30].

Respiratory motion

As the diaphragmatic excursion (up to 3 cm) may exceed a multiple of the coronary vessel diameter (1–4 mm depending on patient age) [26], respiratory motion compensation is also critical for high-resolution coronary artery imaging [31]. To compensate for respiratory motion, the simplest technique is to have the patients hold their breaths. However, in the pediatric population, this translates to having the infant and young children who need sedation for the MR examination to be placed under general anesthesia with paralysis. This not only limits the availability of the coronary MRA examinations to facilities where pediatric anesthesia can be supported but also increases the complexity and risk of the examination. In addition, the achievable volume of coverage of anatomy, the spatial resolution and the signal-to-noise ratio (SNR) will be dictated by the breath-hold duration. The use of MR respiratory navigators during free-breathing, first proposed by Ehman and co-workers [32], removes the time constraints of a breath-hold. There are two options: The MR navigator, ideally a pencil like beam [33], can be either placed on the dome of the right hemidiaphragm [34–38] or directly on the heart [39,40]. Data are accepted if the diaphragm–lung or myocardium–lung interface is within a user defined window (usually 3 to 5 mm), that is preferably positioned around the end-expiratory interface position. Particularly in younger children during sedation or general anesthesia the user defined window should be as small as possible (2 to 3 mm) to get optimal imaging results for the coronary arteries. Temporal position of the navigator immediately prior to
the image acquisition portion of the sequence is beneficial.

Compared to breath-hold approaches, free-breathing navigator gating eliminates registration errors due to inconsistent breath-hold levels or artifacts due to diaphragmatic drift, which are common during sustained breath-holds [31,41]. Free-breathing navigator methods are particularly well suited for more prolonged 3D approaches, combining the post-processing benefits of thin adjacent slices with improved SNR and allowing for high-resolution MRCA. Our clinical experience in the pediatric population indicates relatively high respiratory navigator efficiency of greater than 60% is common in patients who are sedated and freely breathing. General anesthesia with paralysis and mechanical ventilation is usually not necessary.

**Suppression of signal from epicardial fat**

As the coronary arteries are surrounded by epicardial fat with short T1 and resultant MR signal intensity similar to that of flowing blood, fat suppression is crucial for accurate definition of the coronary lumen. This is usually accomplished with a frequency selective small-banded pre-pulse that saturates fat signal and immediately precedes the imaging sequence [2,42].

**Suppression of signal from myocardium**

The underlying myocardium and coronary blood have similar T1 relaxation values (850 ms and 1200 ms, respectively), making delineation of the coronary arteries difficult for 3D techniques, where blood exchange between successive RF excitations is reduced. There are different methods that can be used to enhance the contrast between the coronary arteries and myocardium. Most promising are pre-pulses such as a T2 preparation [43,44], spin-locking [45] or magnetization-transfer-contrast (MTC) [46].

**Coronary MRA acquisition sequences**

Coronary MRA sequences can be conceptualized as a building block of components that include: (1) cardiac (ECG) gating for cardiac motion suppression; (2) respiratory motion suppression (breath-holding, respiratory bellows, navigators); (3) pre-pulses to enhance contrast-to-noise ratio (CNR) of the coronary arterial blood from surrounding tissue (fat saturation, T2 preparation, MTC, selective "tagging" of blood in the aortic root, exogenous contrast agents); and (4) image acquisition that optimizes coronary arterial SNR. The image acquisition scheme includes bright-blood gradient echo sequences, black-blood fast spin-echo techniques. These can be implemented as 2D (typically breath-hold) and 3D (typically non-breath-hold) acquisitions. While these more conventional acquisitions have received the most attention during the last decade, novel techniques such as steady-state-free precession (SSFP) in combination with parallel imaging have become the method of choice since the beginning of 2000. Continued exploration of novel imaging techniques such as spiral, radial, Simultaneous Acquisition of Spatial Harmonics (SMASH) [47], SENSitivity Encoding (SENSE) [48], generalized autocalibrating partially parallel acquisitions (GRAPPA) [49], k-t space Broad-Use Linear Acquisition Speed-Up Techniques (kt-BLAST) [50] have and will continue to receive increasing attention, but their clinical role remains to be defined and will therefore only briefly be mentioned.

**2D and 3D segmented k-space gradient echo CMRA**

Advances in hardware (gradient strength/slew rate, receiver coils) and software (motion suppression, pre-pulses, acquisition schemes) over the past decade have allowed for successful MRCA of the proximal and mid regions of the major epicardial coronary arteries in all subjects in sinus rhythm. Currently, the vast majority of reported MRCA methodologies have utilized bright-blood cardiac MR approaches using 3D segmented k-space gradient-echo acquisitions. Though exogenous contrast agents have been studied (and have become the standard for many non-coronary MRA applications), their role in MRCA is presently under investigation in adult patients [51].

The first robust approach to MRCA was the 2D segmented k-space gradient echo acquisition scheme first described in an isolated heart and in-vivo animal model by Burstein [52] and subsequently in humans by Edelman [1] and Manning...
has been introduced by various MR scanner manufacturers as “TrueFISP”, “FIESTA” or “Balanced FFE”. Fast imaging SSFP seems to be a highly effective and reproducible technique in imaging the coronary vessel lumen and has been successfully applied in children (Figure 14.1) [23,25,54].

Black-blood magnetic resonance coronary angiography (MRCA)
“Black-blood” coronary MRA [55] takes advantage of the negative contrast between flowing coronary blood and epicardial fat/myocardium. Promising preliminary results have been reported [56,57]. “Black-blood” methods may be particularly suited for imaging of patients with metal implants such as bypass graft clips, intracoronary or intravascular stents as these methods are less sensitive to the associated susceptibility artifacts compared to “bright-blood” gradient echo methods. As intracoronary artery stents are rare in children with acquired and congenital heart disease this technique might be useful when other metal implants are problematic for delineation of the origin, size and course of the coronary arteries.

Contrast enhanced magnetic resonance coronary angiography (MRCA)
The application of Gadolinium-DTPA leads to a considerable reduction of the T1 relaxation time of
blood (T1 = 1200 ms without contrast; T1 = 50–100 ms with contrast), thereby resulting in an improved contrast between coronary blood and myocardium. Imaging can either be done using saturation [58] or inversion recovery (IR) pre-pulses [59–61]. After contrast injection, the magnetization of coronary blood returns more rapidly to the equilibrium than for the myocardium. The result is a positive contrast between coronary blood and myocardium. Extracellular agents appear adequate for first pass breath-hold approaches, while equilibrium free-breathing MRA methods benefit from intravascular agents [60–62]. Recent developments in contrast agents with prolonged vascular retention (such as Gadofosveset) offer possibilities in combining this contrast agent with IR pre-pulse sequences for high spatial resolution imaging of the coronary arteries [51,63]. Even though these contrast agents have not yet been approved in children their future use may be very promising for imaging the heart, great vessels and the coronary arteries.

Vessel targeting
Due to the highly tortuous paths of the left and right coronary artery system, multiple double oblique projections are acquired along the natural axis of the coronary artery system during an X-ray angiographic examination. To acquire coronary MRA along these axes, a vessel targeting approach was proposed by Bornert [64], Wielopolski [65] and Stuber [66]. In all protocols, a low-resolution coronary MRA is first acquired to define the anatomic position of the coronary arteries for subsequent planning of the higher-resolution coronary MRA. This is especially valuable for the right coronary artery. Stuber and co-workers utilize an image plane semi-automatically defined by three points along the major axis of the coronary artery [66]. This plane is then used for the subsequent double oblique high resolution coronary MRA of the right coronary artery with a second imaging plane for the left coronary artery.

Weber et al. [67] introduced the whole-heart approach that provides a more user-independent method to image the heart and great vessels including the coronary arteries. This technique was successfully applied in children of different age groups (Figures 14.1–14.3). The isotropic image resolution is especially suited for imaging coronary arteries in infants and children. It is easy to implement and requires minimum user input. In addition, the acquisition of isotropic data facilitates image reformation in any arbitrary imaging plane and makes it particularly useful for follow-up examinations [67,68]. Usually longer segments of the coronary arteries compared with the 3-point plan scan method can be visualized.

Imaging speed
For routine clinical applications, imaging speed is an important issue as the pressure for cost-effective examinations has grown in the last decade. Faster acquisitions also facilitate scanning in less cooperative patients. There are several methods currently under investigation to reduce the imaging time for MRCA. These approaches can be subcategorized in ultra-fast imaging sequences, for example EPI [69,70] or SSFP [46,71] imaging and parallel imaging techniques such as SMASH [47], SENSE [48] or GRAPPA [49].

Parallel imaging techniques offer the potential for markedly enhancing the speed of image acquisition. Both approaches require minimal hardware modifications and can be applied to any MR sequence. While traditionally, hardware/gradient performance was translated into imaging speed, SMASH [47] or SENSE [48] take advantage of the
sensitivity maps of multi coil arrays, thus allowing for image reconstruction with acquisition of a fraction of k-space. The remaining k-lines are then reconstructed by using the sensitivity information of the individual coil elements. The “cost” is the expected loss of signal-to-noise (SNR), with a large benefit of imaging speed. These two novel parallel imaging techniques have demonstrated promising results that have already been reported with 3D coronary MRA particularly in combination with

Figure 14.3 Good agreement was found between invasive selective right coronary cine angiography (a, lateral view) and the corresponding MRCA (b) for the detection of right coronary artery aneurysms in a 7.8-year-old boy (see also Figure 14.2) with Kawasaki disease. MR images were acquired with a whole-heart 3D steady-state free precession technique without the use of contrast agents. Sequential cross-sectional views of the aneurysm demonstrate a thickened vessel wall (arrow) at three different locations (b1, b2 and b3). Images were acquired with a two dimensional (2D) double inversion recovery (DIR) black-blood segmented turbo spin echo technique. Please note the appearance of the normal vessel wall of the descending thoracic aorta (DAo) for comparison. Reproduced with permission from [25].
coronary MRA [67]. K-t BLAST has been introduced recently for time-resolved 3D imaging of the heart and great vessels [72,73]. Ultra-fast imaging techniques for the coronary arteries need to be investigated in the future.

High field MRI
Although 3T MR scanners are now clinically available and successfully utilized in the routine imaging of the nervous system and the musculoskeletal system, wide spread adaptation of cardiac imaging and coronary artery imaging has not yet occurred. Although high field strength, in general, provide increase signal that can be traded off for higher spatial resolution and/or higher temporal resolution, it is more sensitive to magnetic field homogeneity and in particular SSFP sequence implementation is more challenging. Nonetheless, early successes in adult coronary artery imaging has been reported [74,112]. To date there has not yet been reported extensive experience in the pediatric population for the evaluation of congenital heart disease.

Coronary plaque imaging
Early in-vivo coronary vessel wall imaging studies used a 2D fat suppressed fast spin echo (FSE) technique [27,75]. Coronary blood signal was suppressed using a double inversion pre-pulse leading to optimal contrast between lumen and vessel wall. This “black-blood” approach has been implemented both in a 2D breath-hold [75] and a free-breathing mode [27], and allows for visualization of cross-sectional images of the left anterior descending (LAD) and right coronary artery (RCA) vessel wall both in healthy subjects and patients with CAD. In-plane spatial resolution of these first implementations varied from 0.46 × 0.46 mm to 0.5 × 1 mm with a typical slice thickness of 3–5 mm. Coronary wall thickness was found to be higher in subjects with CAD when compared to normal healthy subjects [27,75]. Significant vessel wall thickening was also found in children with severe coronary artery aneurysms after Kawasaki disease (Figure 14.3) [25].

Because of the highly tortuous paths of the coronary artery system, cross-sectional vessel wall imaging of the coronary arteries is time inefficient, making a vessel targeted 3D approach more desirable. Such an approach was implemented using a 3-point plan scan method [66] and combined with a modified black-blood pre-pulse (local inversion) [24], which allows acquisition of 3D stacks along the major axis of the coronary artery system. This approach allowed for visualization of the proximal and mid portions of the RCA and LAD coronary artery walls with good contrast between coronary blood and the vessel walls [24]. Free-breathing 3D black-blood coronary CMR identified an increased coronary vessel wall thickness with preservation of lumen size in patients with non-significant CAD [76–78].

Recently, advances have been made with in vivo imaging of arterial thrombus by fibrin-binding molecular magnetic resonance contrast agents [79,80]. Other contrast agents are developed for detection of early endothelial activation have been developed and are currently tested in rats with brain inflammation [81], and in focal ischemia in mice brains [82]. This Gd-labeled contrast agent, Gd-DTPA-B(sLe\(^x\))A [83], consists of the Sialyl Lewis\(^x\) (sLe\(^x\)) carbohydrate, which interacts with both E- and P-selectin. These promising results encourage further studies including assessment of E- and P-selectin up-regulation in early atherosclerotic lesions in the coronary arteries which would be of great benefit in children with vasculitis such as patients with Takayasu or Kawasaki disease.

Delayed enhancement imaging
Myocardial late enhancement in contrast enhanced cardiac MRI using Gd-DTPA has the ability to precisely delineate myocardial scar. Areas of late enhancement correlate well with areas of fibrosis and thereby enable differentiation between transmural and subendocardial scarring [84]. It is based on the fact that infarcted areas enhance vividly 10–15 minutes after the administration of intravenous paramagnetic contrast agents. This enhancement represents the accumulation of gadolinium contrast in the extracellular space, most likely due to the loss of membrane integrity in scarring tissue. For imaging the contrast-enhanced areas, a gradient echo sequence with an inversion recovery gradient echo pre-pulse is currently used. The inversion times will be adjusted to null the signal of normal myocardium with a nonselective 180° pre-pulse. For 3D localization of the infarcted area
4-chamber and short-axis planes of the myocardium are currently performed. See Chapter 5 on contrast enhancement cardiac MRI for further details.

**Clinical applications**

**Kawasaki disease**

Kawasaki disease is an acute vasculitis of unknown etiology that predominantly occurs in young children and produces coronary artery aneurysms (CAA) in 15 to 25% of untreated cases [85]. CAA may rupture, thrombose, or develop stenotic lesions that cause myocardial ischemia. Serial evaluation of the distribution and size of CAA is necessary for risk stratification and therapeutic management [85,86]. Although transthoracic echocardiography is often utilized for this purpose initially, visualization and characterization of the coronary arteries becomes progressively more difficult as children grow. Serial evaluation with X-ray angiography is costly, invasive, and radiation intensive [16]. The clinical utility of coronary MRA in Kawasaki disease has been shown in several feasibility studies [8,9,25]. Additionally to lumen imaging, vessel wall thickness can be assessed with MR which allows further assessment of the coronary artery system for risk stratification and monitoring of treatment (Figure 14.3) [25]. In a recent study CAAs were successfully imaged and significantly increased vessel wall thickness compared to normal volunteers was shown [25]. Abnormally thickened vessel walls were shown weeks, months and years (Figure 14.3) after onset of the disease [25,87]. Coronary artery stenoses have been shown to develop in the area of CAA as a long-term consequence of Kawasaki disease [85,87,88]. This clinical observation has been confirmed by pathology studies [88]. Imaging the vessel wall may help to quantify future risk for coronary artery stenosis not only based on CAA size but also on diseased vessel wall thickness. Recently, imaging of acute inflammation in Kawasaki disease has been shown in a case report [89]. Combined with the established ability of MRCA to visualize aneurysms in the coronary artery system, and also delineate thrombus within the aneurysms with black-blood technique, MRI has the potential to play an increasing role in initial assessment and follow-up of patients with Kawasaki disease [90]. Currently, there is no gold standard available to confirm vessel wall thickness in children. Intravascular ultrasound is not an alternative in infants and children due to its invasive nature and limited access to peripheral coronary arteries [91]. Transthoracic echocardiography is very limited to reliably depict vessel wall abnormalities in CAA [87]. Multidetector computed tomography (MDCT) is an alternative imaging method of coronary arteries in adults [92]. High heart rates in the pediatric population is a major challenge to MDCT [21,22,92,93,94]. Additionally, general anesthesia would be needed for suspension of respiration and risks for radiation-induced fatal cancer from cumulative computed tomography are not negligible [11].

**Anomalous origin and course of coronary arteries**

Several studies in adults and adolescents have shown the high accuracy of coronary MRA for determining the origin and delineating the proximal course of the anomalous coronary arteries even in cases that X-ray angiography fails to achieve definitive results [95]. Anomalous origin of the coronary arteries has great clinical significance in routine clinical work. In our practice, exclusion of anomalous coronary arteries in the work-up of adolescent patients with unexplained syncope is a main indication for cardiac MR examination. Anomalous course of coronary arteries between the aorta and the pulmonary arteries are correlated to sudden death and surgical correction is recommended in these patients (Figure 14.4) [96]. Excellent results can be achieved in early diagnosis of an anomalous origin of the coronary artery from the pulmonary artery as in patients with Bland–White–Garland syndrome [97]. Delayed diagnosis may lead to repeated coronary infarction with significant morbidity and mortality [97]. Also, for pre-operative planning, imaging of the coronary arteries is of clinical importance. Patients with tetralogy of Fallot, the most common cyanotic congenital heart defect, have abnormal coronary arteries in 4 to 5% [98]. This influences the operative strategy in these patients as a Rastelli operation might be needed in the case of an infundibular course of a coronary artery branch. In addition, there are numerous variations of
coronary artery anatomy in patients with transposition of the great arteries [99,100]. Intramural courses of coronary arteries is considered as an additional risk factor for the arterial switch operation [100]. Increasingly, complex cardiac surgery needs to be performed in many of the patients with congenital heart disease. Some surgical procedures, such as arterial switch operation for repair of transposition of the great arteries, involve excision and re-implantation of the coronary arteries. This may cause coronary artery stenosis in the long-term follow-up of these patients (Figure 14.5) [101]. As a non-invasive screening tool, MRCA would be of great benefit in these patients.

**Coronary artery fistulas**

Coronary artery fistulas are common findings in a variety of congenital heart defects (Figure 14.6) [102–105]. Several modalities are described for imaging these coronary abnormalities including echocardiography [106], computed tomography [107,108] and MRI [103]. Current MRI techniques are limited to good sized coronary artery fistulas as motion artifacts and spatial resolution are limiting factors (Figure 14.6). X-ray angiography remains currently the gold standard in imaging particularly

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**Figure 14.4** The abnormal course of the left main coronary artery (LM) between the Aorta (Ao) and the pulmonary artery (PA) in a 13-year-old patient is clearly demonstrated on the reformatted three-dimensional Steady-State-Free Precession (3D SSFP) magnetic resonance coronary artery angiography (MRCA) image (a) in a patient with repeated syncope after physical stress. After reimplantation of the LM the patient was free of symptoms (b).

**Figure 14.5** A 7-year-old patient after an arterial switch operation for complete transposition of the great arteries is shown. Echocardiography could not adequately visualize the coronary and pulmonary arterial anatomy. A navigator gated and ECG triggered whole heart 3D SSFP MRCA was performed. The anatomy of the proximal coronary arteries with the circumflex coronary artery (Circ) arising from the right coronary artery (RCA) and courses posterior to the neo-aortic root (Ao). The Circ has an acute proximal angulation with a mild narrowing. The left coronary artery (LCA) arises from the left anterior facing sinus. Cube shows spatial orientation: A: anterior; F: foot; L: left; MPA: main pulmonary artery.
small coronary artery fistulas particularly in newborns and children.

**Delayed enhancement myocardial imaging**

Additional information of coronary artery imaging can be provided by delayed enhancement myocardial imaging. This technique provides information regarding the viability of myocardial tissue. In pediatric cardiac MRI, there are many applications for this technique. Most frequently encountered clinical situations in a busy pediatric cardiac MR program where delayed enhancement technique is utilized include: myocardial infarction after surgical intervention (Figure 14.7), coronary artery thrombosis in patients with Kawasaki disease, follow-up examinations in patients after arterial switch operation [101] or after correction of tetralogy of Fallot [109], and diagnosis of cases with arrhythmogenic right ventricular cardiomyopathy (ARVC) [110,111].

**Summary**

MRCA is particularly suited in pediatric patients given its non-invasive nature, absence of radiation, and its independence of acoustic windows. As a 3D imaging technique and the ability to differentiate between various soft tissues, MR is the ideal imaging technique...
modality to visualize the coronary artery system in relation to the surrounding tissue and vasculature. Major challenges in imaging the coronary artery system of pediatric patients are small vessel size, high heart rates, and respiratory motion artifacts. However, several new technical developments in the field of MRCA allow successful application of this technique even in young pediatric patients.

In the future new MR contrast agents [57,62], high field MRI [74,112], faster image acquisition techniques [50], new coil developments [113] and new motion correction techniques [114] may help to increase image contrast, reduce motion artifacts and decrease image acquisition time. MRCA may then be a faster, more robust, non-invasive, and more reproducible imaging tool for the diagnosis, treatment planning and monitoring of the coronary artery system in all age groups of patients with acquired and congenital heart disease than it is, even today.

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PART II

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CHAPTER 14 Imaging coronary arteries in children

CHAPTER 15

Other complex congenital heart disease – heterotaxy, complex spatial relationships, conjoined twins and ectopia cordis

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Introduction

Heterotaxy, L-looped ventricles, ectopia cordis and conjoined twins represent some of the most complex alterations in cardiovascular morphology in humans. Not infrequently, echocardiography (echo) cannot provide a comprehensive picture of relevant cardiovascular anatomy in these situations due to the narrow field of view, lack of optimal acoustic windows, difficulty in delineating complex spatial relationships, and incomplete visualization of the extracardiac vasculature. Magnetic resonance imaging (MRI) performs an important complementary role to echocardiography in these situations. MRI is also increasingly being used as a sole imaging modality for comprehensive pre-surgical evaluation. This chapter will focus on the imaging of heterotaxy by MR, followed by a brief discussion of the imaging of twisted atrioventricular connections, conjoined twins, and ectopia cordis.

Heterotaxy

Definition

Situs refers to the position of the atria and viscera relative to the midline. There are three types of situs: solitus, inversus, and ambiguous. Atrial and visceral situs are almost always the same, i.e. both solitus, or both inversus, or both ambiguous. Heterotaxy is synonymous with situs ambiguous, and is simply defined as “situs other than solitus or inversus”. In the Greek language, “heteros” means “other” and “taxis” means “arrangement”. Hence, heterotaxy literally means “other than the usual order or arrangement”. It is present in 3% of neonates with congenital heart disease.

Situs solitus and situs inversus

In order to diagnose heterotaxy, one must exclude the possibility of situs solitus and inversus. Visceral situs solitus is characterized by the presence of a right-sided liver, a single or dominant left-sided spleen, 3-lobed right lung with an eparterial bronchus, and a 2-lobed left lung with a hyparterial bronchus. Atrial situs solitus is characterized by the presence of the systemic venous atrium (morphologic right atrium) on the right, and the pulmonary venous atrium (morphologic left atrium) on the left. Situs inversus is defined as the “mirror-image” of situs solitus. Although the location of the cardiac apex and the stomach is usually on the left in situs solitus, and on the right in situs inversus, they are generally not considered reliable markers of visceral situs, because their position is frequently altered by diseases involving the thorax and abdomen.
A third syndrome of heterotaxy manifested by levocardia with a single right-sided spleen has also been described [4]. Features of this syndrome are similar to that of right isomerism. In an autopsy series of 109 cases of heterotaxy syndrome by Van Praagh, 53% were asplenia, 42% were polysplenia, and 5% were single right-sided spleen with levocardia [5].

A diagnosis of heterotaxy is clinically significant due to the following associations: poor splenic function [6], poor pancreatic function [7], midgut malrotation, extra-hepatic biliary atresia [8], and abnormal visceral and vascular anatomy encountered during routine surgery [9] (Figure 15.1). However, the major prognostic determinant in heterotaxy is the type of congenital heart disease, which is reportedly present in 50–100% of cases [10], with the major causes of mortality being heart block, complex extracardiac vascular pathology, and complications resulting from a single right ventricle.

**Diagnostic approach to heterotaxy**

Although the classification of heterotaxy into left and right isomerism is helpful for teaching purposes, the high frequency of exceptions to this syndromic approach to diagnosis results in poor clinical utility [11]. Therefore, most centers have discarded this syndromic approach in favor of a segmental approach to delineating the visceral and cardiovascular anatomy in heterotaxy, in which there is an independent assessment of every involved organ system without any preconceived notions.

Pre-operative imaging of heterotaxy typically occurs in the newborn period, and consists of chest radiography, echocardiography, and an abdominal ultrasound to determine abdominal visceral situs, splenic situs and abdominal vascular anatomy.
rial collaterals. MRI is complementary in this setting, providing a high resolution 3-dimensional dataset, with excellent delineation of complex spatial relationships and the extracardiac vascular anatomy with a high degree of diagnostic confidence. Because of radiation considerations including the fact that these patients, who generally have complex heart defects, will get exposed to multiple imaging studies involving radiation such as cardiac catheterization, CT scanning is avoided unless absolutely necessary. In a paper comparing MRI to echocardiography and cardiac catheterization in pre-surgical planning of heterotaxy, accurate delineation of pulmonary venous connections was not achieved by echocardiography in 42% of patients, and by catheterization in 25% of patients. The interobserver agreement was also better on MRI relative to echocardiography or catheterization [14].

Conventional radiographs of the chest and abdomen are not accurate for diagnosis of heterotaxy [12]. The only reliable clue to the presence of heterotaxy on conventional radiography is recognition of an isomeric bronchial branching pattern. But accurate determination of bronchial situs by chest radiography is achieved in only a minority of cases [13]. A discordant position of the cardiac apex and the gastric air shadow is suggestive of the diagnosis, but inconclusive because isolated dextrocardia or isolated levocardia would also have similar manifestations. Additionally, the presence of a normal position of the cardiac apex and gastric bubble does not exclude heterotaxy.

Echocardiography is limited due to a narrow field of view, lack of acoustic windows, inability to delineate complex spatial relationships, as well as the extracardiac vascular anatomy, especially pulmonary veins, systemic veins, aortic arch, and arterial collaterals. MRI is complementary in this setting, providing a high resolution 3-dimensional dataset, with excellent delineation of complex spatial relationships and the extracardiac vascular anatomy with a high degree of diagnostic confidence. Because of radiation considerations including the fact that these patients, who generally have complex heart defects, will get exposed to multiple imaging studies involving radiation such as cardiac catheterization, CT scanning is avoided unless absolutely necessary. In a paper comparing MRI to echocardiography and cardiac catheterization in pre-surgical planning of heterotaxy, accurate delineation of pulmonary venous connections was not achieved by echocardiography in 42% of patients, and by catheterization in 25% of patients. The interobserver agreement was also better on MRI relative to echocardiography or catheterization [14].

![Figure 15.1 Visceral manifestations of heterotaxy.](image)

(a), (b) Transverse liver with a midline gallbladder. (b) Occult polysplenia (two tiny splenic fragments in the left upper quadrant), which was wrongly diagnosed as asplenia on a prior abdominal ultrasound (blue arrow). (c) CT of the abdomen demonstrating right-sided polysplenia, transverse liver, and a blunted appearance (black arrow) of the pancreas (lack of development of the distal body and tail). (d) Transverse ultrasound image of the upper abdomen demonstrating a horseshoe adrenal gland (white arrow). (e) MR angiogram showing common origin of the celiac and superior mesenteric arteries from the abdominal aorta. (f) Bilateral hyparterial bronchi in a patient with polysplenia, with the right upper lobe bronchi arising inferior to the branch pulmonary arteries (white arrows) on either side. (g) Bilateral eparterial bronchi in a patient with asplenia, with the right upper lobe bronchi arising superior to the branch pulmonary arteries (white arrows) on either side.
Goals of imaging
Since any imaginable combination of visceral, cardiac, and vascular morphology can occur, the study of heterotaxy is an excellent exercise in the segmental approach to heart disease. Table 15.2 describes a modified segmental approach to evaluation of visceral and cardiovascular morphology in heterotaxy by MRI, along with common findings in heterotaxy involving each segment [10] as well as the best MR sequence(s) for their evaluation. The ability of MRI to provide a comprehensive evaluation of cardiac and vascular anatomy, and thereby aid therapeutic decision making, is well demonstrated in Figures 15.2 and 15.3.

Table 15.2 Segmental approach to diagnosis in heterotaxy, with typical findings at each segmental level.

<table>
<thead>
<tr>
<th>Visceral or cardiac segment</th>
<th>Asplenia</th>
<th>Polysplenia</th>
<th>Comment</th>
<th>Best MRI sequence(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bronchial anatomy</td>
<td>Bilateral eparterial</td>
<td>Bilateral hyparterial</td>
<td>Up to 15% of cases of heterotaxy may have solitus, inversus, or bronchus suis bronchial branching pattern</td>
<td>Thin slice coronal black-blood</td>
</tr>
<tr>
<td>Lung lobation</td>
<td>Bilateral 3-lobed lungs</td>
<td>Bilateral 2-lobed lungs</td>
<td>Accessory fissures and lobes may be present</td>
<td>None</td>
</tr>
<tr>
<td>Spleen</td>
<td>Absent</td>
<td>Right-sided or left-sided polysplenia, polylobated spleen, rudimentary splenic fragments, anisosplenia</td>
<td>Single right-sided spleen present in 5% of heterotaxy. Splenules may be present in normal individuals</td>
<td>Axial single shot fast spin echo T2 through abdomen.</td>
</tr>
<tr>
<td>Liver</td>
<td>Transverse liver, solitus, inversus</td>
<td></td>
<td></td>
<td>Axial single shot fast spin echo T2 through abdomen.</td>
</tr>
<tr>
<td>Pancreas</td>
<td>Blunted pancreas, Normal</td>
<td></td>
<td></td>
<td>Axial single shot fast spin echo T2 through abdomen.</td>
</tr>
<tr>
<td>Bowel</td>
<td>Midgut malrotation</td>
<td></td>
<td>None (upper GI study)</td>
<td></td>
</tr>
<tr>
<td>Abdominal vasculature</td>
<td>Preduodenal portal vein, common origin of the celiac artery and SMA, replaced hepatic artery, absent splenic vein</td>
<td></td>
<td>3D MRA</td>
<td></td>
</tr>
<tr>
<td>IVC</td>
<td>IVC on same side as aorta, tortuous intrahepatic course, duplicated IVC, left IVC to unroofed coronary sinus</td>
<td>Interrupted IVC (80%), intact (20%)</td>
<td></td>
<td>Axial static or cine FGRE stack through abdomen, MRA, 3D SSFP</td>
</tr>
</tbody>
</table>
### Table 15.2 Continued

<table>
<thead>
<tr>
<th>Visceral or cardiac segment</th>
<th>Asplenia</th>
<th>Polysplenia</th>
<th>Comment</th>
<th>Best MRI sequence(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hepatic veins</td>
<td>Drainage to: 1. Unilateral IVC 2. Ipsilateral IVC 3. IVC and atrium 4. Unilateral atrium 5. Ipsilateral atria likely through unroofed coronary sinus 6. Separate insertion into floor of common atrium</td>
<td>Axial static or cine FGRE stack though abdomen, MRA</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SVC</td>
<td>Bilateral SVC with or without connecting vein, Left SVC to coronary sinus which may be unroofed, Left SVC to superior aspect of common atrium</td>
<td>Axial cine fast FGRE stack though chest, MRA, 3D SSFP</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Coronary sinus</td>
<td>Absent (unroofed) in 95%</td>
<td>Absent coronary sinus septum in 28%</td>
<td>LSVC or hepatic veins may drain into unroofed coronary sinus, must be addressed during 2 ventricle repair</td>
<td>Axial cine FGRE stack though chest, MRA, 3D SSFP</td>
</tr>
<tr>
<td>Pulmonary veins</td>
<td>TAPVC to systemic vein, TAPVC to RA, TAPVC to common atrium</td>
<td>Malposition of septum primum causing PAPVC or TAPVC to RA, TAPVC to RA, Normal.</td>
<td>Axial cine FGRE stack though chest, MRA, 3D SSFP</td>
<td></td>
</tr>
<tr>
<td>Atria</td>
<td>2 separate atria with large ASD, common atrium with rudimentary atrial septum, symmetric appendage morphology, malposition of atrial septum in polysplenia</td>
<td>4 ch cine SSFP, cine FGRE, axial black-blood</td>
<td></td>
<td></td>
</tr>
<tr>
<td>AV canal</td>
<td>Partial or complete CAVC 90%, usually with right dominance</td>
<td>2 AV valves with intact AV septum 35%, CAVC 60%</td>
<td>4 ch cine SSFP, cine FGRE</td>
<td></td>
</tr>
<tr>
<td>Ventricles</td>
<td>Ventricular hypoplasia 55%, L looped ventricles 38%</td>
<td>Ventricular hypoplasia 35%, L looped ventricles 30%</td>
<td>4 ch and SA Cine SSFP, cine FGRE, axial black-blood</td>
<td></td>
</tr>
<tr>
<td>Outflow tract</td>
<td>DORV/TGA, subaortic conus, pulmonary stenosis or atresia</td>
<td>Normally related great arteries, DORV, subpulmonary conus, subaortic obstruction</td>
<td>cine SSFP, cine FGRE, coronal black-blood</td>
<td></td>
</tr>
<tr>
<td>Aorta</td>
<td>Coarctation in the setting of subaortic stenosis, double aortic arch, right aortic arch, aortopulmonary collaterals</td>
<td>cine FGRE, thin slice black-blood, 3D SSFP</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary arteries</td>
<td>Ductal dependent pulmonary blood flow in pulmonary stenosis or atresia, branch pulmonary stenosis</td>
<td>cine FGRE, thin slice black-blood, 3D SSFP</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Figure 15.2 Illustrative case: newborn male with heterotaxy, asplenia, unbalanced right dominant common atrioventricular canal, double outlet right ventricle, severe pulmonary stenosis, obstructed total anomalous pulmonary venous return to the left sided atrial appendage, status post Blalock–Taussig shunt on day 1 of life. He was referred for MRI on day 2 of life to delineate systemic and pulmonary venous return. (a) A cardiac coil was used. Coronal EPI T1 black-blood sequence with 2 mm thick slices shows bilateral right-sided bronchial morphology with short mainstem bronchi, and eparterial take-off of the upper lobe bronchi (arrows). (b) Axial cine FGRE sequence demonstrates an anomalous pulmonary vein (curved arrow) carrying the entire pulmonary venous return, inserting into the base of the left-sided atrial appendage (not shown). The vein was partially obstructed at the site of insertion. (c) Posterior view of a 3-D MRA sequence demonstrates the anomalous pulmonary vein (curved arrow) traveling ventrally, a right-sided IVC which drains the right hepatic vein and inserts into the right sided atrium (long white arrow), three remaining hepatic veins, which come to a common confluence inserting into the left-side atrial appendage (not shown). The Blalock–Taussig shunt is also demonstrated (arrowhead). Repair of the total anomalous pulmonary venous return was performed, with anastomosis to the posterior wall of the common atrium. An atrial septectomy was also performed. At 6 months of life, the patient underwent a bidirectional superior cavopulmonary anastomosis, with takedown of the Blalock–Taussig shunt, and main pulmonary artery isolation. At 3 years of life, he was referred for a MRI to plan a completion Fontan procedure. (d) Posterior view of a 3-D MRA sequence demonstrates a patent superior cavopulmonary shunt (short arrow). The anomalous pulmonary vein (curved arrow) is also shown. Its previous insertion into the left atrial appendage was now completely occluded (not shown), with the pulmonary veins draining via the surgical anastomosis to the posterior wall of the left atrium. (e) A free breathing navigator 3-D steady state free precession sequence was performed. A vertical long axis plane demonstrates moderately severe obstruction of the common pulmonary vein anastomosis to the posterior wall of the atrium, with proximal pulmonary vein dilatation (arrow). (f) The SSFP sequence shows the right hepatic vein inserting separately into the atrium (arrow), but the IVC is not visualized. (g) A thick slab MIP image of the 3-D SSFP sequence demonstrates thromboses of the entire length of the IVC, with numerous venous collaterals draining the renal veins (thin arrows) through the paravertebral veins and the azygos vein (thick arrow). A number of abdominal wall venous collaterals are also noted, draining the lower extremities (curved arrow). (h) and (i) The patient also had moderately severe common atrioventricular valve regurgitation, demonstrated on the magnitude (h) and phase (i) images. The regurgitant jet is marked by the arrow on both images. Based on the above findings, the patient was considered a poor candidate for a completion Fontan procedure. Due to the poor prognosis associated with a systemic right ventricle with atrioventricular valve regurgitation, the patient was counseled for a possible cardiac transplant in the future.
CHAPTER 15  Other complex congenital heart disease

In the immediate post-natal period, IV access in the arm or leg is preferred to an umbilical venous catheter which could potentially terminate in an occluded ductus venosus. Heterotaxy patients have a high incidence of duplicated superior vena cavae without a connecting vein, duplication of the inferior vena cavae, or isolation of the hepatic veins, and are prone to developing acquired thrombosis of the major systemic veins. Since the first pass magnetic resonance angiography (MRA) is ideal for evaluation of the extra cardiac vasculature, selecting an appropriate route of contrast administration is critical to the success of the study.

MRI technique
Cardiac MRI, in the setting of heterotaxy, is a dynamic user-dependent exam, and changes to the protocol will be needed based on real-time evaluation of the initial sequences.

Patient preparation
Prior to start of a MR examination in a patient with heterotaxy, review of chest radiographs helps to screen for devices such as endovascular coils and stents which can create artifacts on the MR images, and to confirm position of the heart in the thorax, which will determine positioning of the EKG electrodes. As in any cardiac MR examination, robust synchronization with EKG signal utilizing vectorcardiogram triggering [15] is required. Respiratory bellows can be optimally placed on the patient for pulse sequences needing respiratory triggering; for those manufacturers who offer it, the respiratory navigator technique can also be used.

Intravenous access
In the immediate post-natal period, IV access in the arm or leg is preferred to an umbilical venous catheter which could potentially terminate in an occluded ductus venosus. Heterotaxy patients have a high incidence of duplicated superior vena cavae without a connecting vein, duplication of the inferior vena cavae, or isolation of the hepatic veins, and are prone to developing acquired thrombosis of the major systemic veins. Since the first pass magnetic resonance angiography (MRA) is ideal for evaluation of the extra cardiac vasculature, selecting an appropriate route of contrast administration is critical to the success of the study.

Patients who have been repaired for complex heart...
part ii  cardiac mr of congenital and acquired pediatric heart disease

disease with a Fontan procedure will require separate injection of intravenous contrast into both upper and lower extremities to optimally evaluate the upper and lower venous pathways of the Fontan circuit [16].

Sedation
In general, sedation is necessary in patients under 8 years of age. If there is a need for assessment of very small vascular structures such as aortopulmonary collaterals, breath-holding will reduce motion blurring, and yield sharper, and potentially more diagnostic images. But, breath-holding requires general anesthesia with controlled ventilation. In our experience, most pulse sequences can be performed in sedated and freely breathing patients yielding diagnostic images.

Coil selection
A phased array receive coil that provides adequate coverage should be used. The coverage should extend from the middle of the neck to the level of the renal vessels in all patients with heterotaxy so that important findings outside the thorax such as infra-diaphragmatic total anomalous pulmonary venous connection or collaterals from systemic venous obstruction are identified. Ideal coverage is provided by the head or shoulder coil in neonates and small infants, the cardiac coil in the young pediatric age group, and the torso coil in larger patients. Phased array coils are preferred since they allow for the use of parallel imaging techniques, which significantly reduce scan time while maintaining spatial resolution.

Pulse sequences
The following is a summary of pulse sequences used. All sequences have benefited from the advent of parallel imaging, which uses information from multiple coil elements to speed up acquisition or increase temporal resolution. Parallel imaging techniques such as SENSE [17,18] should be applied when possible to all these pulse sequences.

Black-blood sequences: The most commonly used sequences are fast spin echo double-inversion recovery sequence, performed with EKG gating and breath-holding, or a spin echo echoplanar imaging (EPI) sequence, performed with EKG gating and free-breathing. With free-breathing sequences, respiratory motion is compensated by multiple signal averages with/without respiratory triggering. Black-blood sequences provide multi-slice static images, with excellent spatial resolution even in neonates. They provide an overview of vascular anatomy, spatial chamber relationships, airway morphology, and abdominal visceral anatomy, and form an important part of the preoperative evaluation of heterotaxy. Post-processing with minimum-intensity-projection is helpful to demonstrate complex anatomical relationships.

Bright-blood sequences: As an alternative to the static black-blood sequences above, a set of static steady state free precession (SSFP) images, usually a contiguous axial set, can be performed; this can then be reformatted into any plane to delineate the salient points of the anatomy or as “localizers” for subsequent dark-blood or cine imaging.

The most commonly used cine sequence is segmented k-space SSFP. It has excellent temporal resolution, allowing multiphase evaluation across the cardiac cycle, and optimal myocardial and blood pool contrast. However, cine SSFP can be limited by out-of-plane flow-related phase incoherence artifact [19] especially in small patients with rapid flow [20]. In addition, the spatial resolution achievable in cine SSFP can be limited as well. Therefore, the old “workhorse” sequence, cine fast gradient echo (FGRE) with segmented k-space sequence, is preferred to the SSFP sequence when thin slices with high resolution are required in small patients. It can be acquired with free breathing and multiple signal averaging which all comes at the expense of imaging time. It is more sensitive to in-plane intravoxel dephasing signal loss from flow turbulence, which can be used as an important diagnostic clue to the presence of shunts, stenosis and regurgitation. In heterotaxy, the cine FGRE sequence is frequently performed in the axial plane with overlapping thin slices to track the course of the extracardiac vasculature, and to determine veno-atrial connections.

Alternatively, isotropic whole heart coverage utilizing a 3D SSFP sequence with respiratory navigator gating has been used in patients with congenital heart disease [21] with good results (Figure 15.4). It has the potential to provide comprehensive static, high-resolution morphologic bright-blood evaluation of intra-cardiac morphology and
coronary flow, and pressure gradients across stenoses using the modified Bernoulli equation.

MR angiography: The most common sequence used is the contrast-enhanced MRA (CEMRA) performed in dynamic fashion following bolus injection of gadolinium using a 3-D T1-weighted fast gradient echo sequence. CEMRA provides a high-resolution 3D dataset, with its main application being evaluation of the extracardiac thoracic vasculature, including the pulmonary arteries, pulmonary veins, aorta, systemic veins, and collateral blood supply to the lungs (Figure 15.4). Techniques of post-processing include multiplanar reformatting, maximum-intensity-projection, volume rendering, and virtual endoscopy. While the post-processed images are helpful to convey information to the referring physicians in an extracardiac vascular anatomy in one sequence. Its performance has not been as robust in the setting of small patients with high heart rates, which constitutes the bulk of patients with heterotaxy requiring pre-operative evaluation; a low number of segments/view are used to increase the chances of optimal imaging. This of course adds time to the scan.

Flow velocity mapping is accomplished by a gradient echo based pulse sequence known as phase contrast (PC). PC imaging is based on the principle that moving protons such as flowing blood, when experiencing a specific bi-polar gradient, will accumulates a predictable phase shift that is proportional to its velocity. PC imaging can be used in the post-operative setting to quantify stroke volume, valvular regurgitation, Qp/Qs, differential pulmonary artery flow, venous return, pulmonary artery (e), which was essentially a Kawashima procedure, and aortic arch augmentation (b). The repaired aortic arch was tortuous and dilated, causing mass effect on the pulmonary artery (images b, c, f and g). The 3-D SSFP sequence compares quite favorably to the time resolved MRA in delineation of the pulmonary arteries, aortic arch, the left cavopulmonary anastomosis and the azygos arch. (Az) in (i) A: aorta; P: pulmonary artery; S: superior vena cava.

Figure 15.4 The use of navigator three dimensional steady state sequence (top row of images) as an alternative to gadolinium enhanced time-resolved 3-dimensional magnetic resonance angiography (bottom row). 7-year-old male with polysplenia, bilateral superior vena cavae (SVC) (e and h), a left-sided interrupted IVC with azygos continuation (d), and a variant of hypoplastic left heart syndrome with aortic arch hypoplasia. His initial palliation was connecting the left SVC to the left
easily understandable format, they should be used to supplement, and not supplant a thorough evaluation of the raw data to extract diagnostic information from the CEMRA. This is due to the significant risk of artifacts being created during the post-processing, causing pseudo-stenoses, and false communications between vessels. On the 3D renderings, it is also important to remember that the MRA is a luminogram, and does not provide any information about the vessel wall. Gadolinium contrast agent has been recently discovered to be associated with development of nephrogenic systemic fibrosis predominantly in adult patients with acute or chronic renal failure [22]. Therefore, thoughtful determination of the benefits of using gadolinium is needed prior to administration, especially in a pediatric patient [23], although very few pediatric patients have been reported to have developed this disease. Prudence dictates that if there is any question regarding renal function, either a renal consultation should be obtained or gadolinium should be avoided.

**Imaging planes**

**Pre-operative evaluation:** Black-blood imaging is performed in the axial and coronal planes, with the latter being aligned along the trachea and proximal bronchi. Bright-blood imaging is performed in the axial plane. MRA is performed in the sagittal or coronal planes.

**Post-operative evaluation:** Vertical long axis, four chamber, and short axis planes are routinely used, with right ventricular and left ventricular outflow tract views, aortic root short axis views, and customized planes to image the atrial septum, atrial baffles, aortic arch, and external conduits being used whenever necessary. 3D MR angiograms may be performed in the sagittal or coronal planes.

**Indications and protocols for MRI in the setting of heterotaxy**

The role of MRI in heterotaxy may be considered at four different time points.

**In the immediate post-natal period prior to initial palliation**

Initial palliation stands for first stage surgical procedures that are typically done immediately after birth to stabilize the patient including relieving pulmonary vein, pulmonary artery or aortic outflow obstruction, augmentation of pulmonary blood flow with a modified Blalock–Taussig shunt, or reducing pulmonary blood flow with a pulmonary artery band. MRI is always indicated especially when a comprehensive look at the organs is required. In addition, because of the complex nature of the heart disease in heterotaxy, MRI is used to confirm anatomy, view spatial relationships such as the ventricular septal defect relative to the semilunar valves, assess ventricular function and Qp/Qs, etc. Hence, the imaging question for MRI is typically quite discrete, pertaining to the status of the pulmonary and systemic vasculature (Figures 15.2, 15.3, 15.5–15.7) although general questions are also answered well with MRI. It is important to remember that the patient is potentially unstable, thereby requiring a short and efficient MRI study. The MRI protocol includes axial cine FGRE performed with 3 to 4 mm slice thickness with overlap to diminish errors related to volume averaging, thin section black-blood images in the axial and coronal planes through the chest, and a high-resolution gadolinium enhanced 3-D MR angiogram.

Indications for MRI evaluation of the pulmonary artery arise in the setting of pulmonary stenosis or atresia. MRI is indicated to screen for the source of pulmonary blood flow, confluence of the branch pulmonary arteries, and branch pulmonary artery stenosis prior to BT shunt placement. An axial cine gradient echo sequence with overlapping thin slices, and a contrast enhanced MRA are adequate for morphologic evaluation of the pulmonary arteries.

Indications for MRI of the aorta are usually in the setting of aortic stenosis or atresia, a characteristic of polysplenia, and to rule out coarctation. A cine gradient echo sequence is performed in the oblique sagittal plane, aligned along the aortic arch. MR angiography is the best sequence for the aortic arch, providing information on the location, extent and severity of stenosis.

In cases where there is a need to evaluate the abdominal manifestations of heterotaxy, a rapid single shot fast spin echo T2 weighted axial sequence through the abdomen can be added to the protocol (Figure 15.1b).
Prior to permanent palliation
After initial palliation, the patient proceeds to permanent palliation in the form of either a single ventricle repair or a two-ventricle repair [24]. Since a number of patients with heterotaxy have a functional single right ventricle, with procedures including bidirectional Glenn, Kawashima, or modified Fontan. The critical questions then deal with obstruction of the pulmonary arteries or pulmonary veins [25], and the feasibility of incorporating the systemic veins into the Glenn and Fontan circuit [26,27].
If a 2-ventricle repair is performed in the setting of heterotaxy, modifications are usually necessary to achieve separation of systemic and pulmonary venous return in the form of venous baffles, atrial baffles, or ventricular to pulmonary artery conduits. MRI helps in decision-making between two-ventricle and single ventricle repair in complex cases by providing an excellent overview of ventricular morphology (Figure 15.9), ventricular volume [28] as well as vascular mor-

**Figure 15.6** Pulmonary venous anomalies in heterotaxy.
(a) and (b) Coronal black-blood images demonstrating total anomalous pulmonary venous return (v) to the SVC in a patient with asplenia, who also has a functional common atrium with a rudimentary atrial septum (white arrow in (b), and separate insertion of the hepatic veins into the floor of the common atrium. (c) Malposition of the septum primum (black arrow) causing partial anomalous pulmonary venous return of the right-sided pulmonary veins to the right atrium in a patient with polysplenia. The patient also has two intact atrioventricular valves, with two well-developed ventricles. (d) Infradiaphragmatic total anomalous pulmonary venous return to the portal vein (black arrow) in a patient with asplenia.
CHAPTER 15 Other complex congenital heart disease

Figure 15.7 Status of the branch pulmonary arteries in heterotaxy. (a) Neonate with polysplenia, severe hypoplasia of the proximal right pulmonary artery (black arrow), and a ductus arteriosus (white arrow) arising from the innominate artery supplying the distal right pulmonary artery (RPA). (b) Newborn patient with pulmonary atresia. A large patent ductus arteriosus (PDA) (white arrow) is the source of pulmonary blood flow. The pulmonary arteries are confluent. Note the tight stenosis at the distal end of the PDA. Ao: aorta; LPA: left pulmonary artery.

Figure 15.8 Post-operative imaging of the systemic veins in heterotaxy. (a) and (b): Figure a is a posterior view of a volume rendered 3-D MRA, and figure b is a left lateral view. Patient with polysplenia, interrupted IVC with azygos (Az) continuation, and bilateral SVC (S), who underwent a single ventricle repair with a left-sided Kawashima procedure (left SVC to left pulmonary artery anastomosis), a right-sided bidirectional Glenn procedure (right SVC to right pulmonary artery anastomosis), and an extracardiac Fontan conduit (F) to return hepatic venous blood to the lungs. (c) Stenosis of the left SVC (arrow) in a different patient following a left-sided bidirectional Glenn procedure.
Valvular function – degree of regurgitation after common atrioventricular valve repair.

Flow – Qp/Qs, differential pulmonary flow, direction of flow.

Aorta and branches – re-coarctation, aorto-pulmonary collaterals.

Specific questions for MRI can focus on every aspect of cardiac morphology and function:

- **Ventricular function and mass** – following 2-ventricle repair with systemic RV, after single ventricle repair.
CHAPTER 15 Other complex congenital heart disease

- **Pulmonary arteries** — branch pulmonary artery stenosis after Blalock-Taussig and central aortopulmonary shunts, unifocalization.
- **Pulmonary veins** — recurrent stenosis after total anomalous pulmonary venous connection repair or transplantation.
- **Systemic veins** — IVC and hepatic venous anatomy to plan Fontan procedure, veno-venous collaterals after cavopulmonary connection.

There is increasing clinical evidence that MRI screening of cardiovascular morphology and function prior to cavopulmonary connection can serve as an alternative to catheterization in low risk patients [31,32]. Although the clinical question may be discrete, it is wise to perform a complete anatomical survey and functional evaluation to determine operability. Thin slice axial cine FGRE with overlap is an excellent means of tracking the course of the extracardiac vasculature, and to screen for stenosis. Thin slice coronal black-blood imaging provides an overview of morphology. Cine SSFP is used for functional evaluation. A whole heart 3-D SSFP with respiratory navigator is performed as a backup angiographic data set with the potential to better delineate aortopulmonary collaterals and systemic venous collaterals (Figure 15.11). Qp/Qs is routinely obtained prior to a Fontan procedure to screen for potential problems like aortopulmonary or venous collaterals, and to determine differential pulmonary flow. Finally a contrast enhanced MRA provides comprehensive evaluation of the extracardiac vasculature (Figure 15.4). Please see Chapter 16 on cardiac MRI in the single ventricles.

**In the late monitoring phase after permanent palliation**

Following permanent palliation, usually after a modified Fontan procedure, the MRI protocol is no different from any other patient with a single ventricle. The imaging question may occasionally be discrete, like cause of cyanosis after Fontan (Figure 15.11), but more often, it is to perform a comprehensive evaluation of morphology, function and flow in a stable patient. A significant number of these patients may not be able to obtain an MR due to the presence of coils placed previously for aortopulmonary and venous collaterals. The MRI protocol is similar to a pre-Fontan evaluation, with the exception being that phase contrast imaging of the SVC, IVC, ascending and descending aorta, and the branch pulmonary arteries are added to determine Qp/Qs ratio as well as the fraction of collateral arterial or venous flow. A delayed enhancement sequence is also added for myocardial fibrosis and can tell if intra-cardiac thrombi are present [33]. As noted above, please see Chapter 16.

**Pitfalls in MRI evaluation of the extracardiac vasculature in heterotaxy**

Questions regarding systemic and pulmonary venous return, the status of the aortic arch and branch pulmonary arteries constitute the most common indications for MRI in heterotaxy. Familiarity with the anatomy prior to planning the study, achieving adequate sedation to reduce motion artifact, choosing the right coil to provide the required coverage, and choosing the right sequences and planes are important pre-requisites for a successful study. The eye sees only what the mind knows. Hence, familiarity with anatomical variations at each segmental level in the setting of heterotaxy is also important.

Venous information is best obtained on a high resolution 3-D MR angiographic dataset acquired during the first pass of contrast agent through the vessel of interest. In heterotaxy, first-pass imaging of all the involved vasculature may be impossible to achieve even with simultaneous contrast injection into the upper and lower extremities, due to the aberrant vascular anatomy. The following situations may be encountered that could potentially compromise the quality of the MRA:

1. There is a high incidence of bilateral SVC, without a communicating vein, frequently draining into ipsilateral atria or pulmonary arteries. Simultaneous injection of contrast into both upper extremities would be required if first pass imaging is desired for optimal visualization.
2. The IVC is frequently interrupted in the setting of polysplenia, or may be occluded.
3. There is a high incidence of independent drainage of hepatic veins into the atria, the filling of which is dependent on systemic venous pressure, and the integrity of the portal circulation.
4. In the setting of Fontan physiology, there is significant stasis in the venous circulation, resulting
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available on all magnet platforms, and requires special expertise to perform successfully on small children, or on patients with high heart rates. An axial cine FGRE bright-blood sequence with overlapping thin slices is usually adequate for providing diagnostic information regarding systemic and pulmonary venous return. Thin slice black-blood images in the coronal plane oriented along the long axis of the SVC, IVC, and hepatic veins, are also helpful. Redundant information from different sequences is essential to achieve a comprehensive

5. The presence of hepatic or pulmonary parenchymal enhancement on the delayed dynamics further limits evaluation of vessels with diluted intravascular contrast.

A respiratory navigator triggered whole heart isotropic 3-D SSFP sequence has been used as an alternative (or backup) angiographic dataset, and is not subject to the vagaries of distribution of intravenous contrast. But this technique is not

Figure 15.11  (a) and (b): 7-year-old male with history of asplenia, status-post repair of infra-diaphragmatic total anomalous pulmonary venous connection, status-post single ventricle repair with a left-sided extracardiac Fontan conduit. Coronal 3-D SSFP image (a) shows an unobstructed inferior vena cava (IVC) draining via the Fontan conduit (F) into the pulmonary artery; (b) is a MIP image from a 3-D MRA showing patency of the anomalous pulmonary vein (white arrow) which communicated with the right portal vein in the abdomen. (c) and (d) are reformatted oblique coronal, and axial images respectively from a navigator respiratory triggered 3-D SSFP whole heart acquisition in a patient with asplenia, status-post completion Fontan surgery. There are numerous systemic to pulmonary venous collaterals (white arrow) lying ventral to the spine, arising from the abdomen and draining into the left lower pulmonary vein.
alignment and atrioventricular connections. The ventricular mass is rotated such that the axes of the atrioventricular inlets cross each other in the transverse plane. The majority of them have atrioventricular concordance; atrioventricular discordance may also be present. For instance, Figure 15.12 is a MRI of a patient with situs solitus of the atria and viscera, d-looping of the ventricles, and d-malposition of the great arteries. In this patient, one would expect atrioventricular concordance and ventriculoarterial discordance. But, because of the crossed atrioventricular connections, this patient has discordant atrioventricular connection, with the right atrium connecting to the left sided morphologic left ventricle, and the left atrium connecting to the right-sided morphologic right ventricle. The base to apex axis of the heart spirals almost 180 degrees, with the two atrioventricular blood streams crossing each other. In a less severe rotational anomaly of the ventricular axis, the ventricles may assume a superior–inferior orientation, with a horizontally oriented interventricular septum (Figure 15.13). This is referred to as the upstairs–downstairs or superior–inferior ventricles. Associated atrial and ventricular septal defects, and outflow tract anomalies are common, with the patients demonstrating either a concordant or discordant ventriculoarterial connection or double outlet right ventricle.

Conclusion
MRI has an increasingly important role in heterotaxy syndrome, in the pre-operative and post-operative period. It is currently used along with echocardiography in heterotaxy in the comprehensive management of these patients. The most common indications for MRI in heterotaxy are suspected anomalous systemic and pulmonary venous return, aortic arch morphology, source of pulmonary blood flow in pulmonary atresia, branch pulmonary artery stenosis, and decision making regarding single or 2-ventricle repair. The setting of heterotaxy is a virtual Pandora’s box, and knowledge of all the possibilities, and a dogged pursuit of the anatomic minutiae are essential for successful management.

Other complex congenital heart disease
Twisted atrioventricular connections
Twisted atrioventricular connections, otherwise known as criss-cross, or topsy-turvy hearts, refer to hearts with discordance between atrioventricular alignment and atrioventricular connections. The ventricular mass is rotated such that the axes of the atrioventricular inlets cross each other in the transverse plane. The majority of them have atrioventricular concordance; atrioventricular discordance may also be present. For instance, Figure 15.12 is a MRI of a patient with situs solitus of the atria and viscera, d-looping of the ventricles, and d-malposition of the great arteries. In this patient, one would expect atrioventricular concordance and ventriculoarterial discordance. But, because of the crossed atrioventricular connections, this patient has discordant atrioventricular connection, with the right atrium connecting to the left sided morphologic left ventricle, and the left atrium connecting to the right-sided morphologic right ventricle. The base to apex axis of the heart spirals almost 180 degrees, with the two atrioventricular blood streams crossing each other. In a less severe rotational anomaly of the ventricular axis, the ventricles may assume a superior–inferior orientation, with a horizontally oriented interventricular septum (Figure 15.13). This is referred to as the upstairs–downstairs or superior–inferior ventricles. Associated atrial and ventricular septal defects, and outflow tract anomalies are common, with the patients demonstrating either a concordant or discordant ventriculoarterial connection or double outlet right ventricle.

MRI is ideally suited for noninvasive evaluation of complex spatial relationships of the ventricular chambers and great vessels because it is not limited by acoustic windows, offers unlimited planes of evaluation, and a high-resolution 3-D dataset which can be processed and analyzed in multiple oblique planes. The protocol for evaluating complex spatial relationships of the chambers includes thin slice black-blood images in the axial and coronal planes, dynamic SSFP imaging in the ventricular and atrial short axis and four chamber planes, and gadolinium enhanced 3-D MR angiography. A navigator whole heart 3-D SSFP sequence is also useful in this setting, providing simultaneous multi-planar evaluation of intracardiac and extracardiac anatomy.

Conjoined twins
Conjoined twins are classified according to the location of the tissue that links the twins. Twenty-eight percent of conjoined twins are classified as
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rated thoracopagus twins, Chiu et al. reported an overall survival rate of 48% [44].

The current approach to the pre-operative evaluation of complex visceral, vascular, and bony anatomy in conjoining includes the use of non-invasive multi-planar techniques like ultrasound and MRI as first-line tools [45,46]. The cardiovascular anatomy frequently determines feasibility of successful surgical separation, and is well evaluated by gadolinium enhanced time resolved 3-D MR angiography [47]. The most important information provided by MRI is related to the spatial relationships of the shared anatomical structures in the heart, and the detailed anatomical structure of the biliary tracts. This is easily achieved by thin slice black-blood imaging in multiple planes (Figure 15.14), cine gradient echo imaging of the heart and extracardiac vasculature, single shot fast spin echo T2 of the shared liver for biliary information, and a time resolved contrast enhanced MRA. The information derived from MRI is indispensable for surgical planning, in which the major

thoraco-omphalopagus [40]. Congenital heart disease (most commonly, a ventricular septal defect or tetralogy of Fallot) is present in 75% of thoracopagus twins, with some degree of fusion of the pericardial sac in 90% [41,42].

Twins may be classified according to the degree of cardiac fusion: separate hearts and pericardium, separate hearts and common pericardium, fused atria and separate ventricles, and lastly, fused atria and ventricles, with the feasibility of successful surgical separation diminishing in the latter two groups [43].

Surgical separation of conjoined twins must be planned on a case-by-case basis with survival depending on the type and extent of conjoining and on the presence or absence of associated anomalies. The liver and bowel are shared in 81% of omphalopagus twins, and have a major bearing on outcome after surgical separation. Surgical separation is usually deferred for weeks or months, since survival improves with increasing age at surgery. In a review of 47 pairs of surgically separated thoracopagus twins, Chiu et al. reported an overall survival rate of 48% [44].

The current approach to the pre-operative evaluation of complex visceral, vascular, and bony anatomy in conjoining includes the use of non-invasive multi-planar techniques like ultrasound and MRI as first-line tools [45,46]. The cardiovascular anatomy frequently determines feasibility of successful surgical separation, and is well evaluated by gadolinium enhanced time resolved 3-D MR angiography [47]. The most important information provided by MRI is related to the spatial relationships of the shared anatomical structures in the heart, and the detailed anatomical structure of the biliary tracts. This is easily achieved by thin slice black-blood imaging in multiple planes (Figure 15.14), cine gradient echo imaging of the heart and extracardiac vasculature, single shot fast spin echo T2 of the shared liver for biliary information, and a time resolved contrast enhanced MRA. The information derived from MRI is indispensable for surgical planning, in which the major

Figure 15.12  Criss-cross ventricles. Three-month-old male with criss-cross atrioventricular valve relationship, with the right atrium draining via the mitral valve into the left ventricle, and the left atrium draining via the tricuspid valve into the hypoplastic right ventricle. (a) is a volume rendered 3-D MRA; (b) and (c) are axial cine bright-blood sequences; while (d), (e), and (f) are reconstructed images from a navigator three dimensional steady state free

precession (3-D SSFP) whole heart examination. (a) and (d) Demonstrate the d-malposed aorta arising from the hypoplastic right ventricle. The arrows in (b), (c), (e) and (f) denote the crossing A-V pathways. The 3-D SSFP sequence is comparable to conventional bright-blood sequences in demonstrating complex intracardiac anatomy as well as the status of the extracardiac vasculature.
**Figure 15.13** Superior inferior ventricles. 1-month-old female with dextrocardia, (L,L,I) segmental cardiac anatomy, with superior-inferior orientation of the ventricles. (a) Is a coronal black-blood image, and (b) is a cine steady state free precession short axis image showing the right ventricle, which is heavily trabeculated, lying superior to the left ventricle, which has a smooth septal surface.

**Figure 15.14** Imaging of conjoined twins. (a) “Double” sagittal black-blood image demonstrating extensive fusion of the atrial and ventricular chambers of the heart, as well as the liver in thoracopagus twins, decreasing chances of successful separation. (Performed at Children’s Hospital, Boston, MA, USA). (b) Volume rendered 3-D image of the heart of the surviving twin following separation of thoracopagus with a fused heart. A conduit (white arrow) was placed from the RV outflow tract (RVOT) of the surviving twin to the remnant aorta of the non-surviving twin to preserve intact coronary perfusion of the heart. (Courtesy of Dr. Mark Fogel, Children’s Hospital of Philadelphia, PA, USA.) Ao: aorta; LV: left ventricle.
challenges are separation of important shared organs such as the liver and the heart, and closure of defects in soft tissue and bone [48,49].

**Ectopia cordis**

Ectopia cordis is defined as complete or partial displacement of the heart outside the thoracic cavity. Four types of ectopia cordis [50] have been described according to the location of the heart: thoracic (65%), thoraco-abdominal (20%), abdominal (10%) and cervical (5%). Thoracic ectopia cordis constitutes the classic naked heart with no overlying somatic structures. Ventricular septal defects and tetralogy of Fallot are the most common cardiac defects, while omphalocele, cleft lip and cleft palate are other common associations [50,51].

In 1958, Cantrell *et al.* described a syndrome [52] called the pentalogy of Cantrell, consisting of the following: deficiency of the anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, a defect of the lower sternum, and congenital intracardiac abnormalities (Figure 15.15).

MRI can provide all the information relevant for surgical planning (fig. 15.15), including the size of the skin, sternal and pericardial defects, the presence of associated intracardiac anomalies, and the status of the liver and thoraco-abdominal vasculature. The protocol comprises thin slice axial and sagittal black-blood sequences, axial thin slice cine GRE, cine SSFP in the 4-chamber and short axis planes, and a contrast enhanced 3D MRA.

A two-stage correction is usually performed, first with a covering skin flap in the neonatal period, followed by a rib graft and pectoral muscle graft over the sternal defect [53]. Synthetic or prosthetic material have also been used, but with

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**Figure 15.15** Pentalogy of Cantrell with associated tetralogy of Fallot. MRI was performed to delineate intracardiac anatomy in a 9-day-old female with pentalogy of Cantrell. (a) Oblique sagittal black-blood image demonstrating a large omphalocele with ectopia cordis, sternal defect and pericardial defect. (b) Aorta (A) overriding the conoventricular VSD. (c) Infundibular RVOT obstruction. (d) Apical muscular VSD (arrow). (e–g) Hypoplastic branch pulmonary arteries, with stenosis of the origin of the left pulmonary artery (arrow in g). P: pulmonary artery.
an increased risk of infection. Survival is strongly correlated with the absence of intrinsic cardiac defects [54].

References

Cardiac MR of congenital and acquired pediatric heart disease

PART III

Special topics in cardiac MR of pediatric and congenital heart disease
Cardiac magnetic resonance of single ventricles

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Chapter 16

Introduction
The patient born with a single ventricle remains one of the last frontiers and the most complex aspects of all congenital heart disease. Nearly all patients require reconstructive surgery or heart transplantation. In addition, the stages of reconstructive surgery which eventually lead to the Fontan procedure [1] place varying loads on the ventricle (from a volume loaded ventricle to a volume unloaded one) as well as varying physiologies (Figure 16.1) (for example from the run-off lesion into the pulmonary circulation in the Norwood Stage I reconstruction [2] to one without one at Fontan completion). To further complicate matters, single ventricles are not one lesion but rather a collection of many types which fall under the same diagnostic category. It’s no wonder that the care and management of these patients, including their assessment using imaging, remains one of the greatest challenges in pediatric cardiology.

Cardiac magnetic resonance (CMR) has come to play a major role in the management of single ventricle patients as a group is the variable anatomy; for example: (A) D-loop vs. L-loop; (B) right (RV) vs. left ventricle (LV); or (C) anatomic true single ventricle (i.e. a common atrioventricular valve or both atrioventricular valves enter one ventricle in the presence of only one sinus portion of the heart) versus a “functional” single ventricle (any type of ventricular arrangement where from a “functional” standpoint, the ventricle acts as single pumping chamber).

For example, a true single LV would be a heart having a common atrioventricular valve connect-

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ing to a single, morphologic LV chamber that gives rise to at least one great vessel; a “functional” single LV would be a patient with D-looped tricuspid atresia. As can be seen, there can be a seemingly hopeless number of complex combinations; however, the one underlying theme is that only one usable ventricle is present or both ventricles are connected in such a way that separating them into two pumping chambers is impossible.

During the various stages of surgical reconstruction, as noted above, the physiology of the cardiovascular system changes dramatically (Figure 16.1). In the patient’s native state, if there is outflow obstruction or hypoplasia of one of the great vessels, blood flow to the obstructed circulation can be maintained by either flow through a ductus arteriosus (Figure 16.2), a stenotic pulmonary valve (allowing just enough blood to enter the pulmonary circulation) or a ventricular septal defect (for example bulboventricular foramen) if one or both great vessels arises from the hypoplastic ventricle. The ultimate goal of surgery is to completely separate the systemic and pulmonary circulations and place them in a “series circuit.” This is done by allowing passive blood flow into the pulmonary circulation via a conduit while the functional single ventricle pumps to the systemic circulation; this is performed in stages.

In the native state, surgery may or may not be needed if adequate but restricted pulmonary blood flow is present (for example double outlet RV with pulmonic stenosis and a subaortic ventricular

![Figure 16.1](image.png)

*Figure 16.1* Diagram of the different physiology at each stage of surgical reconstruction for single ventricles: At Stage I, the single ventricle pumps to both systemic and pulmonary circulations in parallel with a systemic to pulmonary artery shunt (top). After bidirectional superior cavopulmonary connection (BSCC), the single ventricle pumps only to the systemic circulation with blood from the brain and upper body supplying flow to the lungs as well (middle). After Fontan reconstruction, the systemic and pulmonary circulations are in series (bottom).
Figure 16.2 Patient with hypoplastic left heart syndrome prior to Stage I. (a) Double inversion dark-blood images demonstrate the hypoplastic aorta (Ao) in long axis (upper and lower left) and the large patent ductus arteriosus (PDA) in long axis (upper left and lower middle). These structures are demonstrated in cross-section in the upper middle image. The upper and lower right images are again double inversion dark-blood images which demonstrate the hypoplastic left ventricle (LV) in the 4-chamber and short axis views respectively. DAo: descending aorta. (b) FLASH images demonstrating the large PDA, flow in the hypoplastic Ao and the ventricles in short axis (from left to right). RV = right ventricle.
septal defect). Patients with hypoplastic left heart syndrome (Figure 16.2), however, will always require surgical intervention—the Norwood Stage I procedure [2], which includes a systemic to pulmonary artery shunt, an atrial septectomy (Figures 16.1 and 16.3) and an aortic to pulmonary anastomosis (Figure 16.3). An RV to pulmonary artery conduit (Sano procedure) has been substituted for the systemic to pulmonary artery shunt in some institutions [5]. At this stage, the single ventricle pumps to both the systemic and pulmonary circulation in parallel, causing a volume overload. Results of the Sano procedure have been mixed regarding its superiority over the systemic to pulmonary artery shunt with early hospital mortality [6–9] and no difference in midterm analysis after the next stage (see below) [10].

Once pulmonary vascular resistance has dropped adequately (∼4–6 months of age), the bidirectional superior cavopulmonary connection (BSCC) is
performed such as the hemi-Fontan procedure or the bidirectional Glenn (Figure 16.4). As the name suggests, this is a connection of the superior vena cava(e) to the pulmonary artery with exclusion of superior vena caval blood flow to the atrium and ligation of the systemic to pulmonary artery shunt. Since blood needs to go to the head/arms first before entering the pulmonary circulation, the ventricle does not pump directly to the pulmonary circulation and is therefore not technically volume loaded. It is not clear, however, that it remains volume unloaded throughout the time the patient is in this physiology [11]; indeed, it has been demonstrated that systemic to pulmonary collaterals are present [12]. This recently has been quantified by CMR (see below) [13]. Cardiac output is maintained at the expense of cyanosis as only a portion of the systemic venous return enters the lungs (i.e. flow from the head and arms and from aorto-pulmonary collaterals if present). This second stage was instituted when it was noted that when forming a Fontan directly after a stage I reconstruction, the ventricular wall thickness-to-chamber dimension ratio acutely increased [11] with low output, tachycardia, and hemodynamic deterioration; diastolic ventricular compliance issues were thought to play a major role. In some instances, an additional source of pulmonary blood flow is left in place, such as a systemic to pulmonary artery shunt or forward flow through a stenotic pulmonary valve; this physiology definitely places a volume load on the ventricle.

At approximately 2 years of age, directing inferior vena cava blood into the lungs is performed to complete the Fontan operation (Figure 16.5); this can be done in a number of ways such as by placement of a patch along the lateral wall of the atria (“lateral wall tunnel”), an extracardiac conduit, or an “atrio-pulmonary connection” (no longer used). The circulations are now separated and all blood must traverse the lungs by passive flow to maintain cardiac output. A connection in
between the systemic and pulmonary venous pathways, a “fenestration,” is purposely created to allow for shunting between the circulations during times of increased pulmonary vascular resistance. Cardiac output can be maintained at the expense of cyanosis (as in the BSCC). Fenestrations, in general, close on their own. Since the circulations are now separated, the ventricle is volume unloaded (after the fenestration closes), except for possible aorto-pulmonary collaterals.

**CMR protocol for imaging the single ventricle patient**

In general, the goal of CMR imaging in single ventricle patients is to delineate the anatomy and assess the physiology and function as with most other patient populations. At different stages of surgical reconstruction, certain structures and key points may be different, but the overall goal remains the same. At all surgical stages (including pre-surgical), the following is the minimum assessment:

- Pulmonary artery imaging, to assess for pulmonary artery stenosis, hypoplasia, discontinuity (Figures 16.3–16.5). CMR has been shown to be superior to echocardiography in delineating the pulmonary arteries in single ventricle patients and correlates better with images from angiography [3].
- Aortic arch imaging, aimed mostly at patients with an aortic to pulmonary anastomosis, to assess for aortic arch obstruction (Figure 16.3). This includes aortic to pulmonary collaterals.
- Ventricular outflow tract obstruction (especially in patients with a bulboventricular foramen).
- Pulmonary or systemic venous obstruction including the status of the atrial septal defect.
- Anomalous venous structures.
- Ventricular function.
- Velocity mapping to assess for cardiac index, Qp/Qs, relative flows to both lungs and regurgitant fraction of the semilunar (and indirectly) atrioventricular valve.

**Native state:** At this stage, it is important to do a detailed assessment of the anatomy since much less is known about the patient’s anatomy than at other stages. The presence of a left superior vena cava or other anomalous venous structures, delineation of visceral situs for heterotaxy, presence or absence of an inferior vena cava, etc. are all important issues. In addition, because some patients may have been resuscitated in the newborn period (which is sometimes the history), assessment of ventricular function and valve insufficiency is also extremely important to evaluate.

**After the Stage I procedure:** Aortic imaging is important at this point to evaluate the initial repair as arch obstruction may be present. In addition, the to pulmonary anastomosis as well as visualization of the aortic to pulmonary shunt needs to be assessed (Figure 16.3). The aortic to pulmonary shunt is generally performed with dark-blood imaging (or gadolinium) as the turbulence of the shunt by cine generally causes signal loss. An RV to pulmonary artery shunt should be evaluated in the same fashion. Qp/Qs is obtained by velocity mapping the ventricular outflow and by either velocity mapping across the shunt or in each pulmonary artery (and summing the flows) using a high VENC (for example 400 cm/sec), with as low an echo time (TE) as possible. The status of the atrial septal defect should be assessed and since this is a volume loaded stage, ventricular function is also a key imaging goal (Figure 16.4).

**After the BSCC:** (Figure 16.4) Since the superior vena cava to pulmonary artery anastomosis is the major reconstruction, this is the one of the major focuses of the exam. This can be done with dark-blood, cine or gadolinium sequences as the flows are generally low velocity. Qp/Qs can be calculated by flow in the superior vena cava or in both branch pulmonary arteries (note that this generally is very different from catheterization derived data). This, however, may be fraught with error since aortic to pulmonary collaterals which are not obvious on gadolinium enhanced imaging may be present and not taken into account by this method. Recently it has been shown that pulmonary venous flow is a more reliable means of measuring Qp in this population because of this [13]. To further bolster the evidence that collaterals are present, this study demonstrated that the addition of pulmonary venous flow and inferior vena cava flow equaled
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Figure 16.4 Patient with single ventricle after superior cavopulmonary connection. The left upper panel and lower middle panel are steady state free precession images of two patients after a bidirectional Glenn where the superior vena cava (SVC) is divided from the right atrium (RA) and connected in an end-to-side fashion to the right pulmonary artery (RPA). The upper middle image is an off-axis view of a patient who underwent a hemi-Fontan, where the SVC is not divided surgically from the RA but instead, is connected side-to-side with the RPA and a “dam” is created surgically between the SVC and the RA, preventing blood from entering the RA from the SVC. The right upper image is an anterior view of a volume rendered 3-dimensional gadolinium dataset which also demonstrates the SVC-RPA connection and its relationship to the rest of the heart and great vessels (for example the aorta [Ao]). The right lower image is a shaded surface display isolating the SVC to RPA connection. The left lower image is delayed enhancement imaging demonstrating myocardial scarring and the atrioventricular valves showing increased signal intensity (arrows). LPA: left pulmonary artery.

aortic flow. Contrary to data from cardiac catheterization which uses the superior vena cava saturation as mixed systemic venous saturation (which is extremely questionable), Qp/Qs was near 1. If a hemi-Fontan is performed, leaks into the atrium from the superior vena cava-pulmonary artery anastomosis should be assessed.

After the Fontan procedure: The most key structure to image is the entire systemic venous pathway for obstruction, thrombus and fenestration flow, since this is the major reconstruction (Figure 16.5). Since patients who have undergone the Fontan procedure have ventricular dysfunction [6], evaluating ventricular performance is an essential part of the exam. In addition, gadolinium enhanced imaging can help determine the presence of collaterals and to assess the aortic arch.

In general, it should be noted that two key retrospective studies, one in patients undergoing hemi-Fontan/bidirectional Glenn procedure and one in patients undergoing Fontan completion both demonstrated that cardiac catheterization should not be the “routine” in all single ventricle patients [14,15]. A combination of non-invasive imaging modalities such as CMR and echocardiog...
Conduct of the CMR exam for single ventricles (after localizers) which is typically performed in under an hour (in the older Fontan patient who can breath-hold, in even less time):

1. Steady state free precession contiguous axial images – this surveys the cardiovascular anatomy and is used as localizers for other imaging in the study. One of the important points is that if the therapy can clearly delineate all that is needed prior to surgery and can categorize patients into those that need a cardiac catheterization prior to their operation (such as an intervention in the lab) and those that do not. A prospective study by Brown et al. [16] further demonstrated this fact using CMR and echocardiography in patients prior to hemi-Fontan/bidirectional Glenn; this study also showed that it is less costly with the non-invasive approach. Patients who underwent catheterization had longer hospital stays and more adverse events; however, the surgical success rate for the catheterization and non-invasive imaging groups were similar.

Conduct of the CMR exam for single ventricles (after localizers) which is typically performed in under an hour (in the older Fontan patient who can breath-hold, in even less time):

1. Steady state free precession contiguous axial images – this surveys the cardiovascular anatomy and is used as localizers for other imaging in the study. One of the important points is that if the
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scan is terminated early due to technical problems or patient instability, at least a full anatomic volume data set has been obtained and can be reformatted to determine the important parts of the anatomy. Obvious anatomic issues can of course be evaluated in this manner at the outset. If there is turbulence in structures during diastole (for example systemic to pulmonary artery shunt), signal loss will occur which is one of the major drawbacks of this type of imaging. Systemic and pulmonary venous anatomy, the aorta and pulmonary arteries along with the size of the various cardiac chambers in diastole can be assessed at this point.

2. HASTE contiguous axial images – This is another type of axial imaging and is acquired while the imager loads the set of static steady state free precession contiguous axial images into multiplanar reconstruction for subsequent imaging. It is also used as axial images to view structures that may have diastolic turbulence in them.

3. Reformating of axial images – Our lab utilizes multiplanar reconstruction of the axial images to localize subsequent imaging. This includes the “2-chamber” long axis and short axis views of the heart for ventricular function (can be used to standardize this measurement), two orthogonal views of the ventricular outflow tract, the candy cane view of the aortic arch, long axis views of the pulmonary arteries, the superior vena cava to pulmonary artery anastomosis, etc. In addition, this is used to ensure image planes are as perpendicular as possible to flow for through plane phase encoded velocity mapping.

4. Double inversion dark-blood imaging – Used sparingly since this type of imaging takes a long time. It is used to image the systemic to pulmonary artery shunt, evaluate for clot or masses in the systemic venous pathway, or as an alternative to bright-blood/cine CMR imaging of various important structures such as the pulmonary arteries and the aortic arch.

5. Cine CMR – Generally, assessment of ventricular performance (cines of the “2-chamber” long axis and a set of contiguous short axis images from base to apex) to quantitate end-diastolic volume, mass, ejection fraction, stroke volume and cardiac index are obtained first. If the patient is sedated or under anesthesia and averaging is used, reformating of the axial images for other structures can be performed while this imaging is being obtained since it doesn’t require operator intervention. Afterwards, other anatomic questions are evaluated so that cines can be obtained of the pulmonary arteries (long axis, cavopulmonary anastomosis), the systemic and pulmonary venous pathways, the candy cane view of the aorta, the aortic to pulmonary anastomosis and ventricular outflow tracts as examples.

6. 3-dimensional gadolinium imaging – Used for 3-dimensional viewing of the pulmonary arteries, aorta and systemic venous structures and lays the foundation for subsequent viability imaging 5–10 minutes later. This may be performed as a static procedure (i.e. obtain 3–5 single 3D slabs) or as a time-resolved one (i.e. inject gadolinium and visualize its passage through the cardiovascular system). With the advent of new technology, even these time-resolved procedures can create high resolution 3D images with each time point (see below).

7. Velocity mapping – This can be done after gadolinium to add increased signal to the data. Flow is typically obtained: (a) across the branch pulmonary arteries for relative flows to both lungs; (b) across the semilunar valves (if two are present) for cardiac output; (c) across the superior and inferior vena cavae and/or across the atrioventricular valve can be obtained; and (d) across the pulmonary veins. After Stage I, flow in the systemic to pulmonary artery or RV to pulmonary artery shunt can be obtained using a high VENC and a low TE.

8. Viability (Figure 16.4) – Usually performed in the ventricular short axis view and the “2-chamber” long axis views to evaluate for myocardial scarring; the single ventricle undergoes cardiopulmonary bypass and deep hypothermic circulatory arrest in at least two if not more operations – this includes extensive intracardiac and extracardiac reconstruction. Myocardial scarring, therefore, is always a possibility. Viability sequences have also been used to identify patch material used for surgical reconstruction as well [17].

9. Special imaging – These are performed specific to the patient and may be fitted in at some
point along the scanning pathway. For example, selected coronary imaging utilizing navigator sequences can be performed if there is a question regarding the coronary arteries. Regional wall motion abnormalities can be assessed using myocardial tagging. "Real-time" interactive steady state free precession cine CMR can be used to "sweep" through the heart and great vessels similar to echocardiography to quickly identify areas of concern, detect turbulence, etc. (subsequent higher resolution scans can then target regions for further assessment). The previous examples should be performed before gadolinium injection to obtain better image quality. Time-resolved gadolinium imaging can be used to assess anatomy quickly or if, for example, there is question of discontinuity between the branch pulmonary arteries. Adenosine stress perfusion can be used if myocardial perfusion is an issue.

**The many faces of the Fontan operation and the single ventricle**

Figures 16.2–16.5 demonstrate the routine structures needed to be imaged throughout the course of staged Fontan reconstruction. Figure 16.2 shows images prior to any surgery, in this particular instance, a patient with hypoplastic left heart syndrome. Figure 16.3 demonstrates the systemic to pulmonary artery shunt and the aortic reconstruction of a patient who has undergone Stage I Norwood reconstruction.

Figure 16.4 shows the two most common forms of superior cavopulmonary connections – the hemi-Fontan and the bidirectional Glenn procedures. These two procedures, which lead to the same physiology, are different: (a) in the bidirectional Glenn, the superior vena cava is divided from the right atrium and connected in an end-to-side fashion to the right pulmonary artery while in (b) the hemi-Fontan procedure, the superior vena cava is not divided surgically from the right atrium but instead, is connected side-to-side with the right pulmonary artery and a "dam" is created surgically between the superior vena cava and the right atrium, preventing blood from entering the right atrium from the superior vena cava. The hemi-Fontan operation usually carries the patch across the pulmonary artery to augment its size and has the theoretical advantage of setting up the patient to complete the Fontan in the cardiac catheterization laboratory with a covered stent.

Figure 16.5 shows examples of a standard Fontan completion (upper images and lower left), where the inferior vena cava is baffled to the pulmonary arteries. The routine repair generally has an extracardiac conduit or lateral wall tunnel placed in the right hemithorax or right atrium connecting the two structures. Axial, off-axis coronal and off-axis sagittal views using steady state free precession imaging generally can yield all the anatomic information needed, including visualizing the fenestration.

There are, however, a wide variety of anatomic variations of single ventricle lesions, and it is important that the CMR physician understands the anatomic, functional and physiologic nature of the creative repairs surgeons undertake so that they can adequately be assessed with CMR. The lower middle and lower right images of Figure 16.5 show an example of one such anatomic variation; a single ventricle patient with a right inferior vena cava and bilateral superior vena cavae. The patient underwent anastomosis of the superior vena cavae to the ipsilateral pulmonary artery as well as placement of a rightward inferior vena cava to right pulmonary artery anastomosis. This results in the "missing leg H" coronal view in the figure.

Another approach when a left superior vena cava to coronary sinus is present is to connect the coronary sinus directly to the systemic venous pathway Fontan baffle instead of ligating and dividing the left superior vena cava and hooking it to the left pulmonary artery. Figure 16.6 demonstrates two functional results of this approach. The images on the left are two frames from a time-resolved 3-dimensional gadolinium injection showing this type of connection with a mildly dilated coronary sinus. The steady state free precession images in the middle and right are of a patient who was not so lucky, and underwent massive dilation of the coronary sinus because of this physiology. The high right-sided systemic venous pressures resulted in this coronary sinus expansion and the patient had decreased ventricular performance, possibly due to coronary sinus hypertension with decreased myocardial perfusion and/or mechani-
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The atrioventricular Fontan, in its variety of forms, connects the atria directly to the pulmonary artery (usually using the atrial appendage). One of the major problems with this surgery was the marked dilation the atria undergoes afterwards, as demonstrated in the figure, with the subsequent risk of arrhythmia, thrombus formation and decreased cardiovascular energetics and performance. The figure also shows the Bjork modification to this procedure, where in addition to the atrioventricular connection, there is a superior cavopulmonary connection as well.

As medicine becomes more successful in the treatment of patients with congenital heart disease, an ever increasing number of children are living to adulthood. The physician imaging these patients not only has to be knowledgeable about the latest medical and surgical management, but also needs to be aware of surgical treatments which have preceded present day practice. One such surgery is the atrioventricular Fontan, in its variety of forms, connects the atria directly to the pulmonary artery (usually using the atrial appendage). One of the major problems with this surgery was the marked dilation the atria undergoes afterwards, as is demonstrated in the figure, with the subsequent risk of arrhythmia, thrombus formation and decreased cardiovascular energetics and performance. The figure also shows the Bjork modification to this procedure, where in addition to the atrioventricular connection, there is a superior cavopulmonary connection as well.

**Figure 16.6** The left superior vena cava to coronary sinus (CS) to right-sided baffle (B) connection. The left two panels are time-resolved, 3-dimensional gadolinium injections demonstrating the anatomy. In the left upper panel, arrows point to the CS, the right-sided Fontan B and the branch pulmonary arteries while on the lower left panels, arrows point to the left and right superior vena cavae (on “levophase”) and the right pulmonary artery. The middle and right panels are steady state free precession images of a different patient with the same anatomy but with a severely dilated CS. The middle upper and middle lower panels are axial and ventricular (V) short axis views whereas the right most panels are two coronal views demonstrating this phenomenon. RA: right atrium; RV: right ventricle.
Another anatomic variation is shown in Figure 16.8. In this, a right inferior and left superior vena cavae are present, with absence of the right superior vena cava. The repair is performed by connecting the left superior vena cava to the left pulmonary artery and baffling right inferior vena caval blood to the right pulmonary artery. This gives rise to the “zig-zag” coronal appearance to the systemic venous pathway seen in the figure.

As a final example of the wide variety of anatomic connections an imager may encounter, Figure 16.9 shows images of a patient with dextrocardia, a single left ventricle with a right ventricular outflow chamber and left-sided juxtaposition of the atrial appendages with restrictive ventricular septal defects. The course of the systemic venous pathway is fairly complex: the conduit courses leftward and remaining posterior as it originates from the inferior vena cava and once in the left hemithorax, courses anterior and superior along the left border of the heart. The conduit’s anterior course takes it anterior to the pulmonary artery, where it finally takes a posterior course to insert anterior on this vessel. Figure 16.9 demonstrates the old saying that a picture is worth a thousand words (or in this case, at least 66 words).

Not only is there a wide variety of anatomic surgical connections, there is also a wide variety of morphologic single ventricles a CMR imager will likely come across. Figure 16.10 is just a
number of different ways; it has become less complex and temporal information is added. For example, multiple static 3D images can be obtained in 6–15 second intervals (too slow for time resolution) called static 3D or can be time-resolved reaching subsecond temporal resolution with high spatial resolution. This can be reconstructed in three orthogonal views (for example coronal, sagittal or axial). Figure 16.11 demonstrates a time-resolved gadolinium injection with 1.5 second temporal resolution and 1.3 cm isotropic spatial resolution in a patient with a single LV after a hemi-Fontan procedure. Each time point can be made into a high resolution 3D volume rendered image; each 3D data set can be added or subtracted from each other to optimize the 3D volume rendered image or the raw data of the structures of small sample of the myriad of morphologic variations. Most fall into categories as mentioned in the beginning of this chapter: (a) RV vs. LV; (b) D-loop vs. L-loop; or (c) true single ventricle vs. a “functional” single ventricle. However, there are other morphologic variations which even complicate this, such as dextrocardia vs. levocardia, supero-inferior ventricles, criss-cross atrioventricular relations, atrioventricular canal vs. two separate atrioventricular valves, etc. The figure shows examples from some of these categories.

With the wide variety of surgical corrections and morphologic single ventricles, there is also a wide variety of CMR procedures to visualize these. With the advances in hardware and software, 3-dimensional imaging can be performed in a number of different ways; it has become less complex and temporal information is added. For example, multiple static 3D images can be obtained in 6–15 second intervals (too slow for time resolution) called static 3D or can be time-resolved reaching subsecond temporal resolution with high spatial resolution. This can be reconstructed in three orthogonal views (for example coronal, sagittal or axial). Figure 16.11 demonstrates a time-resolved gadolinium injection with 1.5 second temporal resolution and 1.3 cm isotropic spatial resolution in a patient with a single LV after a hemi-Fontan procedure. Each time point can be made into a high resolution 3D volume rendered image; each 3D data set can be added or subtracted from each other to optimize the 3D volume rendered image or the raw data of the structures of
Figure 16.9
Figure 16.9 The curved Fontan connection. All panels are steady state free precession images (in “a”) of a patient with dextrocardiac, single left ventricle (V) with a right ventricular outflow chamber (RVOC) and left-sided juxtaposition of the atrial appendages. The ventricular septal defects are restrictive. (a) Selected axial images which progress from inferior to superior as the panels go from left to right (top more inferior than bottom images). The Fontan conduit (C) distally connects to the right-sided inferior vena cava (IVC) and as it progresses proximally, courses to the left hemithorax posteriorly and then courses superiorly and anteriorly on the left side to insert anteriorly into the left pulmonary artery (LPA). (b) The upper left panel is a coronal view of the entire extent of the baffle in one image (arrows) connecting to the pulmonary artery (PA). The upper middle panel is an off-axis 2-chamber view of the main pumping V. The upper right image is a coronal view demonstrating the anatomy with one single left V pumping chamber and the RVOC with restrictive ventricular septal defects (arrows). The only outlet from the single left V is through these ventricular septal defects. The lower left and middle panels are two views through the restrictive ventricular septal defects; note the abnormal jet visualized on the lower left image (arrowhead). The lower right panel is an in-plane phase encoded velocity map through this region at a velocity encoding of 400 cm/s with increased signal intensity indicating the blood flow exceeded this value. Ao: aorta; DAo: descending aorta; LAA: left atrial appendage; SVC: superior vena cava; RPA: right pulmonary artery.

Figure 16.10 (a) Various forms of single ventricles. (a) Two common forms of single ventricle are hypoplastic left heart syndrome (upper left image) and pulmonary atresia with tricuspid stenosis and hypoplastic right ventricle (RV). The former represents a “single RV” and the latter represents a “single left ventricle (LV)” form of single ventricle. The right image is a short axis view of a patient with a single LV. B: baffle.
Continued (b) Examples of more complex single ventricle lesions include patients with supero-inferior ventricles (upper left panel). The systemic venous return enters the atria which connects to the inferior morphologic LV (upper middle and upper right images) while the pulmonary venous return enters the atria which connect to the superior morphologic RV (lower right panel). The patient has obvious dextrocardia. Time-resolved 3-dimensional gadolinium injection demonstrates this as well along with pulmonary lung perfusion (lower middle panel). The lower left panel is an example of a patient with a malaligned atrioventricular canal resulting in a single RV which underwent Fontan reconstruction. Flow across the fenestration can be seen (black arrow). B: baffle.

Figure 16.11
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Figure 16.11 High resolution time-resolved gadolinium injection in a single left ventricle patient after hemi-Fontan. (a) Maximum intensity projections from multiple time points during the gadolinium injection viewed with a temporal resolution of 1.5 seconds and an isotropic spatial resolution of 1.3 cm. The upper images, viewed from a coronal aspect, are of the pulmonary phase (left), the systemic arterial phase (middle) and the recirculation phase (right). Note how clearly the right and left jugular (RJ and LJ respectively) as well as the innominate veins (In) can be seen. The pulmonary arteries (PAs) and aorta (Ao) can be visualized easily. Two lower images are reconstructed from the sagittal aspect demonstrating the PAs (left) and the aorta (middle). The lower right image demonstrates the PAs from the transverse aspect.

(b) Volume rendered images from the maximum intensity projections shown in (a). The pulmonary arteries in anterior, posterior and sagittal views are shown in the upper left, 2nd from left and 2nd from right respectively. The hemi-Fontan pouch (H-P), created by the right superior vena cava-right pulmonary artery anastomosis, is seen prominently. The upper right image is an image of the PAs tipped up towards the transverse aspect. The lower right panel is an image from the posterior aspect of the aorta without the PAs. The lower middle and right panels are images of the PAs and aorta combined in the anterior and posterior views respectively.

Examples of knowledge about single ventricles gained from CMR studies

As with the previous section, it would be impossible to outline all that CMR has contributed to our interest. A recirculation phase can also be seen. Note how both the pulmonary and venous structures can be separated from each other.

The examples in this section are just a sample of the wide variety of surgical reconstructions and morphologic single ventricle the CMR imager may come across. It is impossible to “memorize” all of them; however, if the CMR imager is schooled in basic principles of congenital heart disease as well CMR principles, the anatomic, physiologic, functional and surgical assessment of the single ventricle patient utilizing CMR can be performed successfully.
understanding of congenital heart disease. The examples which follow are just a taste of what has been learned and shed some light on the potential of CMR in the future to elucidate anatomy, physiology and function in congenital heart disease to improve patient care.

Flow
A perennial question always concerns the driving force behind “passive” pulmonary flow in single ventricle patients. It appears to be a combination of negative intrathoracic pressure during inspiration and either directly or indirectly, of the systemic ventricle’s contraction and motion [18–22]. The amount to which all these factors contribute to this passive flow remains debatable with reports of either atrial contraction, ventricular systole, ventricular diastole or the respiratory component determining this flow.

It is clear that dependence of pulmonary blood flow in Fontan physiology cannot be wholly based on respiration because a Fontan patient on a respirator receiving positive pressure ventilation would not survive. It is also clearly not wholly cardiac dependent. In a cardiac MRI study using bolus tagging [18], flow in the Fontan baffle was imaged both gated to the respiratory cycle using bellows at end-inspiration, and end-expiration and triggered to the cardiac cycle via ECG. If flow was solely respiratory controlled, then an ECG triggered study would demonstrate similar images at all cardiac phases. If flow was solely cardiac controlled, then a respiratory gated study would demonstrate similar images at end-inspiration and end-expiration. Both types of scans demonstrated different images during the cardiac cycle and the respiratory cycle. Because cardiac and respiratory cycles occur simultaneously, flow dependency in this study was defined as the percentage change in flow during imaging triggered to the cardiac cycle (or gated to the respiratory cycle) as a fraction of the sum of flow changes noted during both cardiac triggering and respiratory gating. Using this definition, nearly 70% of flow was cardiac dependent, with the rest of the flow being respiratory dependent. Maximum flow occurred during late systole-early diastole (2nd quarter of the cardiac cycle) with the slowest flow occurred during diastasis in the 3rd quarter of the cardiac cycle.

This flow appears to be fairly complex. In a recent article published by Zelicourt et al. [23] CMR anatomic data was used to create a model using transparent stereolithography. Power loss, digital particle velocimetry and flow visualization revealed complex, unsteady and a highly 3-dimensional flow structure with high pressure drops and power losses. Most of the dissipation of the energy occurred in the pulmonary arteries.

Many of the studies utilizing mathematical models and flow optimization in Fontan patients rely on the knowledge of the caval contribution of flow to each lung, to systemic venous return and the relative flow to each lung. Using presaturation tagging to “label” blood from each cava, a CMR study was performed on 10 single ventricle patients with lateral wall tunnel Fontans to determine just those parameters, published in 1999 [24]. Sixty percent of superior vena caval blood was found to flow into the right pulmonary artery and 67% of inferior vena caval blood flowed towards left pulmonary artery (i.e. superior vena caval blood was directed to the right pulmonary artery and inferior vena cava blood was directed towards left pulmonary). The inferior vena cava contributed 40% to total systemic venous return in these approximately 2-year-olds and the distribution of blood to each lung was nearly equal. Three-plane velocity mapping is also an extremely valuable tool in visualizing and analyzing these flows (Figure 16.12), although it can’t “label” blood from each cava.

Ventricular function
The total heart volume concept (i.e. that the combined volume of both atria and ventricles does not change during the cardiac cycle) is a measure of the integrated function of the heart: the change is as little as 5% [25]. This occurs by reciprocating volume changes in the atria and ventricles during the cardiac cycle and results in minimizing the energy needed in moving extracardiac structures. The same is true for intracycle constancy of the center of mass motion for that exact reason [26,27]. In a 1993 study [27], CMR demonstrated that 4/10 patients prior to hemi-Fontan had total heart volumes vary by >5% and the center of mass of the entire heart significantly moved in
indicate that the volume loaded single ventricle not only performs more volume work but also "wastes" energy by unnecessarily displacing extracardiac structures. Only 1/8 patients (13%) after hemi-Fontan (but prior to Fontan) exceeded the 5% limit of total volume change throughout the cardiac cycle and the hemi-Fontan group had the least center of mass motion than the other two surgical subgroups.

As the single ventricle patient progresses through staged surgical reconstruction, different physiological stresses are placed on the ventricle as noted at the beginning of this chapter, which in theory, should lead to altered regional strain and wall motion. Seventy one percent of Fontan patients also did not have constant total heart volume, and although not volume loaded, it is presumed that this law was disrupted in part by baffle placement. When the heart’s center of mass motion was broken down into orthogonal components, correlations existed between the lateral plane and the anteroposterior and superoinferior planes ($r^2 = 0.51-0.91$), presumably because these planes are linked by the lateral wall tunnel baffle sewn into the lateral and posterior walls of the atria. These findings indicated that the Fontan group of patients along with patients prior to hemi-Fontan reconstruction all

Figure 16.12 3-plane velocity mapping in the systemic venous pathway of a Fontan patient. The upper right amplitude image depicts the anatomy of the Fontan baffle (B). The upper left, lower left and lower right are velocity maps encoded foot-head (F-H), right-left (R-L) and through plane (Thru) of the baffle respectively. F-H, R-L and Thru encodes increased signal intensity toward the head, left and posterior respectively. Note the flow patterns. Inn: innominate artery; SVC: superior vena cava.
motion. When this was studied by myocardial tagging using CMR [28], the highest regional compressive strains were found to occur in the ventricles prior to the hemi-Fontan procedure and in those after the Fontan procedure. This may be due to volume loading and disruption of the normal strain patterns by the intracardiac baffle respectively.

Regardless of the stage of surgical reconstruction or ventricular morphology, 31/33 of single ventricles in that same study twisted in the short axis counterclockwise in one region, clockwise in another and met at a “transition zone” of no twist. By comparison, the normal human left ventricle twists uniformly in a short axis. This “transition zone” had the highest strains of all regions. In the Fontan group, it was demonstrated that the inferior walls moved paradoxically in early systole.

**Surgical planning**

Using 3-dimensional velocity mapping as well as static SSFP imaging from CMR, many studies of the single ventricle patient have surfaced to demonstrate that, using computation fluid mechanics, power loss in the systemic venous pathway can be calculated, flows can be visualized and relative contributions of caval flows to the branch pulmonary arteries can be accurately measured [23,29–33]. This approach not only can assess what is presently the status of the circulation in vivo but has the potential to assess various structural modifications of the cardiovascular system. For example, one study assessed the effect of left pulmonary artery stenosis on flow and power loss in the single ventricle patient; this was followed by a “virtual angioplasty” on the workstation and a reassessment of flow and power loss when the stenosis was removed [33].

With this notion, the idea that a surgeon, cardiologist or engineer can create virtual systemic venous baffles and assess the effect of flow and power loss has been entertained and implemented in select patients. One example is the single ventricle heterotaxy patient with an interrupted inferior vena cava with azygous continuation. A Kawashima operation in this type of patient connects the superior vena cava to the pulmonary arteries thereby diverting nearly all systemic blood flow except for the hepatic venous flow to into the lungs. Without hepatic venous blood, pulmonary arterio-venous malformations typically develop; however, when the hepatic veins are subsequently baffled to the superior vena cava–pulmonary artery connection, flow phenomena may occur which would preferentially stream hepatic flow to one lung; this allows pulmonary arterio-venous malformations to occur in the contralateral lung. Figure 16.13 demonstrates one such patient prior to hepatic venous baffle construction with bilateral pulmonary arteriovenous malformations on cardiac catheterization. Hepatic venous baffles are virtually created on a workstation for a number of patient specific options. One is intra-atrial and one is extracardiac with two different geometries each; note how the power loss appears approximately the same for both yet the flow distribution of hepatic venous blood is very different. It is clear given that there are pulmonary arteriovenous malformations in both lungs that the nearly equal flow splits of hepatic venous blood using the intra-atrial approach is preferred. This example demonstrates how this approach can avoid unilateral pulmonary arteriovenous malformations and subsequent hepatic venous baffle revision.

An example of a single ventricle patient with heterotaxy syndrome and azygous continuation to a superior vena cava who had a hemi-Fontan procedure as well as hepatic venous baffle creation is shown in Figure 16.14. This case and the methodology was recently published [34]. The patient was proven to have left lung arteriovenous malformations on cardiac catheterization and the CMR was performed not only to understand the hepatic venous flow splits to both lungs but for patient specific surgical planning as well. From the geometry and flow characteristics determined by CMR and computational fluid mechanics, it was shown that the large “pouch-like” hemi-Fontan reconstruction along with the azygous vein and superior vena cava flow emptying into it created a “vortex” in the hemi-Fontan reconstruction. This “vortex” acted as a wall which prevented hepatic flow from the baffle entering the left pulmonary artery and deflecting this flow into the right pulmonary artery.
Flow from the azygous and superior vena cava were directed into the left pulmonary artery and hence, the development of left pulmonary arteriovenous malformations. Three options were entertained and connection of the azygous vein to the hepatic venous baffle was chosen; after the surgery, the patient’s systemic arterial saturation which preoperatively was 76% was now 94%.

**Conclusion**

It is clear from both a clinical and research standpoint, that the advent of CMR has benefited the surgical and medical management of the single ventricle patient. The CMR imager must be familiar with the basic protocol to image these patients along with the wide variety of anatomy, physiology, function and surgical reconstructive techniques which are found in these patients to successfully assess and contribute to their overall care. CMR is a versatile modality to assess patients with single ventricle, utilizing 2- and 3-dimensional imaging to assess anatomy, morphology, function and flow (Figure 16.15). With the continuing advances in CMR, the next 25 years hold even greater promise of progress in the non-invasive assessment of these patients than the past 25.

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**Pre-Operative Anatomy**

- Single right ventricle with heterotaxy syndrome
- Interrupted IVC - azygous continuation to RSVC
- After Stage I and Bidirectional Glenn operations

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*Figure 16.13 (a) Example of surgical planning in a single ventricle patient with heterotaxy syndrome and an interrupted inferior vena cava (IVC) with azygous (AZ) continuation to the right superior vena cava (RSVC). (a) Pre-operative anatomy after Stage I Norwood operation and bidirectional Glenn procedure (Kawashima operation in this case). IVC here refers to hepatic venous flow for ease of nomenclature. The bottom 3 panels demonstrate the 3-dimensional anatomy with the atria, ventricles and aorta included (in blue) from anterior, right and superior views (from left to right). Systemic venous anatomy is in red. The right upper panel is an anterior view with the atria, ventricles and aorta removed. LPA: left pulmonary artery; RPA: right pulmonary artery; SVC: superior vena cava.*

(continued on next page)
(b) Intra-atrial hepatic venous baffle without (top) and with (bottom) a “flare” into the pulmonary arteries is virtually created. The diagrams demonstrate, in color coded fashion, the pressure drop across the baffle (left), the global flow structure (all blood flow, middle) and hepatic venous distribution to each lung (right). Power loss (PL) is slightly higher without a “flare” than with a “flare;” hepatic venous flow distribution to both lungs for each geometry is approximately the same with the right pulmonary artery (RPA) getting slightly more hepatic venous flow than the left pulmonary artery (LPA).

(c) Extracardiac hepatic venous baffle with two geometries leading into the pulmonary arteries is virtually created. Demonstration of all parameters are similar to (b). Note that PL are in the same range as (b) yet hepatic venous flow distribution is markedly different, favoring the RPA. This may lead to left lung arteriovenous malformations (in conjunction with the bioengineering department at Georgia Tech).
Figure 16.14 Example of surgical planning in a single ventricle patient with heterotaxy syndrome and an interrupted inferior vena cava (IVC) with azygous (AZ) continuation to the right superior vena cava (RSVC) after hepatic venous baffle (B) placement. (a) The top panel is a frame from a coronal cine CMR to demonstrate the B connection to the right pulmonary artery (RPA) and RSVC. The bottom three panels are 3-dimensional reconstructions of the systemic venous pathway from the posterior, anterior and lateral views (going left to right); note the hemi-Fontan “pouch” (P).

(continued on next page)
Figure 16.14 Continued (b) The left upper and right lower images depict the flow streams in the 3-dimensional reconstruction; note the "vortex" (V) formation in the hemi-Fontan P. Yellow arrows depict how blood is channeled from hepatic venous baffle to the RPA and from the AZ and RSVC to the left pulmonary artery (LPA). The upper right graph plots the percentage of hepatic venous flow coursing to the LPA as a function of the cardiac cycle.

(b)

(c) Three virtually created surgical options for this patient. The AZ can be connected to the RPA (left), can be connected to the hepatic venous baffle (middle) or the hepatic venous baffle can use a "Y" graft to split flow to RPA and LPA. Top and bottom panels of each virtual reconstruction are views from different angles (in conjunction with the bioengineering department at Georgia Tech). SVC: superior vena cava.
Figure 16.15 The world of CMR in single ventricles: This summary figure is intended to show the versatility of CMR in assessing single ventricles. The upper right and middle panels are 3-dimensional reconstructions highlighting the native aortic (nAo) to native pulmonary artery (nPA) anastomosis (right) and the Fontan baffle (B) (middle). Phase encoded velocity mapping (upper left) can assess flows in 3-dimensions; the example is an in-plane velocity map of the Fontan B. The lower right panel is a 2-dimensional steady state free precession image of the nAo to nPA anastomosis. The 2nd from the right is a short-axis view of a supero-inferior ventricle whereas the 2nd from left is a steady state free precession “3-chamber” view of a patient with double inlet left ventricle after Fontan. Arrow points to the fenestration flow shunting right to left. The lower left panel demonstrates the morphology of the semilunar valves of the outlets of the double inlet left ventricle with a bicuspid pulmonary valve (BV) and a smaller trileafl et aortic valve (TV); the fenestration flow can be seen here as well (arrow). SVC: superior vena cava.

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CHAPTER 17

Baffles and conduits

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Introduction

Reconstructive surgery of complex congenital heart disease may require redirection of blood flow in the case of an incorrect connection between cardiac segments, or “bridging” of blood flow to lead it across an obstacle as presented by the nature of a specific defect. To achieve this goal, heart surgeons and more recently cardiac interventionists have developed a variety of different techniques involving the use of conduits and baffles to optimize blood flow. A conduit is used for extracardiac redirection of blood flow, usually (but not exclusively) to connect cardiac chambers with arteries. Due to its extracardiac nature, in nearly all (but not all) cases, no viable tissue from the patient’s heart or vessel wall can be used as part of this tunnel, and hence a conduit is composed of circular artificial material resulting in potential lack of growth. Conduits are frequently but not always valved given they usually aim to replace an obstructed outflow tract or semilunar valve. In contrast, a baffle is used for intracardiac redirection of blood flow. This allows any resultant tunnel to be composed, in part, of viable growing tissue, from endogenous heart wall, septal wall or vessel wall tissue, while only the remainder to be composed of extraneous material (the “baffle”) which can be synthetic or fashioned from autologous pericardial tissue. Some baffle operations do not need any foreign material, but rather utilize septal tissue to redirect blood flow [1,2]. Hence, compared to a conduit, an intracardiac baffle is much less likely to become obstructed with growth. Conduits may need replacement due to somatic growth particularly if smaller sizes are implanted at an early age [3].

The ideal baffle or conduit used for surgical repair should resemble the biological properties of native blood vessels in order to provide optimal functional integrity with minimal flow resistance and turbulence. Its material should be fully bio-compatible without thrombogenic properties. There should be no major mismatch in compliance between autologous vessel and implanted scaffold, adequate suture retention and tensile strength at high intraluminal pressures, resistance to infection and, ultimately, the potential to grow with the child [4]. Furthermore, in the case of a valved conduit, preserved valvular competence, lack of stenosis and/or calcification are desirable. However, no such ideal material is currently available though research in the field is very active [5,6], particularly for tissue-engineered valves [7,8] and valved pulmonary conduits [7,9] where early experience in both animal models [10] and patients [11] appears promising. Important problems encountered with available materials include lack of growth, thrombogenicity (due to incomplete endothelial coverage, particularly at low-velocity flow conditions), neointima formation and calcification resulting in obstruction and loss of compliance. The problems are particularly prevalent in valved conduits depending on the materials used and may eventu-
ally lead to sufficient conduit failure to necessitate replacement [3,12]. The issue of limited durability is particularly important in the pediatric population where surgical or transcatheter interventions to treat conduit or baffle failure may be multiple over a lifetime.

In this context the role of cardiac imaging is twofold. First, the choice of the optimal reconstructive approach involving the use of baffles and conduits is largely based on diagnostic imaging techniques displaying the precise nature of the anatomical problem. Second, once the surgical repair has been successfully achieved, both the complexity of the cardiac defect and the composition and size of the graft material will determine the need for and frequency of careful individual follow-up investigations designed to detect graft failure and its effect on cardiac function. Cardiac magnetic resonance imaging (MRI) is a powerful imaging method for these patients providing not only unique three-dimensional structural detail but also hemodynamic information regarding flow rates, flow velocities and ventricular function [13]. Cardiac MRI (CMR) has become the reference standard for the quantitative assessment of ventricular dimensions, function and mass against which other imaging modalities are currently validated [14]. CMR is usually performed following, and in conjunction with, transthoracic echocardiography in neonates and infants. In contrast, CMR often becomes the first line technique in adolescents or adults with more complex anatomy. In these patients, body habitus and interposition of scar tissue and lungs become an increasing problem for transthoracic echocardiography; and conduits which often lie directly retrosternal may be very difficult to visualize. Hence, CMR has largely replaced diagnostic cardiac catheterization in many institutions. In turn, interventional catheter procedures can be more targeted and hence their duration and risks minimized by prior use of CMR.

The purpose of this chapter is to describe in detail current CMR indications and strategies used when planning intervention and follow-up in patients with complex congenital heart defects where baffles and conduits play a key role in the surgical repair. A brief review of present materials and techniques will be made. This will be followed by a summary of CMR imaging principles and then by a more detailed discussion of specific CMR applications in the field. Schematic diagrams will be used where appropriate and corresponding clinical case examples both pre- and post-repair.

Available materials for baffles and conduits

The search for an “ideal” material for conduits and baffles has been a continuous challenge throughout the development of congenital heart surgery including latter day advances in the field of “tissue-engineering” [4,7,11] and transcatheter valve replacement [15]. Grafts may be composed of either autograft material, such as a flap created from the patient’s own pericardium or atrial septum, or allograft material, such as human aortic or pulmonary homograft, or even xenograft material, such as bovine jugular venous valve grafts used for pulmonary valve replacement in infants. Moreover, a variety of artificial prosthetic materials are commonly used in reconstructive congenital heart surgery.

Autologous materials

Autologous materials (genetically identical donor and recipient) certainly have the great advantage of no immunologic reaction against the graft, and there is growth potential. Atrial septal tissue has been successfully used to create baffles for redirection of blood flow [1,2,16]. Pericardial patches may be reconstructed to serve as autologous tissue forming both baffles [17] and ventriculo-pulmonary conduits [18,19]. Fresh autologous pericardial conduits seem to compare favorably with porcine xenograft valved conduits [18,19] although rapid deterioration of fresh autologous pericardial leaflets placed in the blood stream was occasionally observed, leaving a pericardial conduit with an incompetent valve [18,20]. Treatment of autologous pericardial tissue with glutaraldehyde increases the durability of valvular leaflets [21]. Recently, an interesting modification of an intraoperatively self-constructed pericardial pulmonary valved conduit was introduced using a valved pericardial conduit with single point attached commissures [22]. The importance of this alternative to pulmonary homografts or bovine jugular venous grafts lies with the fact that these may not be readily available in all parts of the world.
Allograft materials
Allograft materials are defined as a tissue graft from a donor of the same species as the recipient but not genetically identical with the recipient. In congenital heart surgery these are usually cryopreserved aortic or pulmonary homografts. Aortic homografts may be more difficult to remove and revise due to their propensity for calcification and adherence to surrounding structures. In contrast, pulmonary homografts seem to have shown fewer propensities to obstruction and calcification, and consequently they have been favored over aortic homografts in some centers [23]. Several surgical techniques have been introduced to improve homograft durability. They are largely devoted to decrease any compliance mismatch between either the graft and the native artery distally, or between the graft and the ventricular outflow tract proximally. Moreover, addition of prosthetic or pericardial skirts on the proximal end may be necessary to facilitate connection to the ventricle. It has been has been observed for both aortic and pulmonary homograft conduits that smaller graft sizes and young age at repair are associated with a much higher likelihood for early conduit failure, perhaps secondary to host immunologic response to residual antigenic tissue [3,24]. A further disadvantage is the limited availability of cryopreserved homografts, particularly for smaller sizes.

Xenograft materials
Xenograft material is defined as tissue from a donor of a different species. Examples include the porcine glutaraldehyde-preserved pulmonary xenograft (e.g., Hancock® valve) which is less regularly used as degeneration has been observed to occur more rapidly than with homograft valves [12]. As an alternative for right ventricular outflow tract (RVOT) reconstruction, the bovine jugular valved venous graft (Contegra®) was introduced 1999 as an option for pulmonary valve replacement, hoping to overcome disadvantages of porcine xenografts but also of homografts with their lack of availability and durability in smaller children. Early excellent results reported in animal studies [25,26] were initially confirmed in human clinical trials [27]. However, more recent mid-term reports describe formation of neo-intima proliferation at the distal anastomosis. This may cause severe stenosis and may be associated with proximal conduit dilatation and valve insufficiency, particularly in smaller infants and when there are residual high right ventricular pressures [28,29].

Prosthetic materials
Prosthetic materials of non-biological artificial origin are created to serve as conduits or baffles when biological tissues are unavailable for the purpose, or when the necessary shape or size is not adequate. The two most commonly used materials are Dacron® (Polyethylene Terephthalate), which is widely used in the textile industry, and PTFE (Poly-Tetra-Fluoro-Ethylene). The first clinical application of Dacron® was reported by Dr DeBakey in 1952, who created a Dacron tube graft for aortic reconstruction using his wife’s sewing machine. PTFE was first developed in 1938 by Dr Roy Punkett, and improved years later after the discovery of expanded PTFE which is more compliant and porous. In pediatric heart surgery, PTFE-based materials are used, for example, to create aorto-pulmonary shunt connections such as the modified Black-Taussig shunt [30]; for atrial baffles in the lateral tunnel Fontan operation (Goretex®); and for non-valved ventricular-pulmonary conduits such as with the "Sano" modification of the Norwood operation for hypoplastic left heart syndrome (HLHS) [31].

Bi-leaflet metallic prosthetic heart valves may be integrated into a composite conduit to reconstruct the right ventricular outflow tract or the aortic valve and ascending aorta. This is considered an important alternative to biological valve replacement in adult patients with congenital heart disease, but much less popular in the pediatric age group as management of anticoagulation is a major concern in these patients. Aspects of CMR imaging of metallic valve prostheses are discussed elsewhere in this book.

CMR for baffles and conduits
For both planning purpose and post-repair follow-up, Table 17.1 summarizes our institutional CMR practice, which is to use 3D imaging early during the anatomical CMR investigation and supplement this with appropriate 2D single- or multi-slice
Baffles are not difficult to manage. Cautious and thorough post-repair follow-up is highly reliable. This volume scan can be acquired either as single-phase (end-diastole or end-systole) [32] or dual-phase data set (end-diastole and end-systole) [33]. Respiratory motion correction is achieved by use of a prospective navigator to monitor diaphragm motion. Advanced post-processing tools such as a virtual incision-making, based on reconstruction of 3D CMR images, may be helpful to visualize complex anatomy pre-operatively. The intracardiac morphology can be inspected from any desired surgical view as outlined in Figure 17.1. Coronary artery anatomy as displayed by 3D-SSFP [34] can be integrated into such models. Alternatively, casts may be printed based on such 3D-SSFP data and these can then be used for both planning of operations or interventions as well as for teaching (Figure 17.2) [35].

Table 17.1 General CMR protocol for complex congenital heart disease.

<table>
<thead>
<tr>
<th>Task</th>
<th>Details</th>
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<tbody>
<tr>
<td>1 Scouts and reference scans (parallel imaging)</td>
<td></td>
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<tr>
<td>2 Interactive imaging for acquisition of scan plane geometries (four-chamber, short-axis, RVOT, LVOT, pulmonary trunk through-plane, aorta through-plane, aortic arch, branch pulmonary arteries, 2 orthogonal in-plane views and through-plane)</td>
<td></td>
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<tr>
<td>3 3D contrast-enhanced MRA of the thoracic great vessels (1–3 dynamics)</td>
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<tr>
<td>4 Balanced-SSFP (b-SSFP) 3D “whole-heart” volume scan (or: conventional black-blood axial imaging if not available)</td>
<td></td>
</tr>
<tr>
<td>5 2D multi-slice b-SSFP cine images for volumetry</td>
<td></td>
</tr>
<tr>
<td>5.1 Whole ventricle in short-axis plane</td>
<td></td>
</tr>
<tr>
<td>5.2 Alternatively, in complex conditions, to cover the whole heart in a transverse plane</td>
<td></td>
</tr>
<tr>
<td>6 2D single-slice b-SSFP cine images</td>
<td></td>
</tr>
<tr>
<td>6.1 Four-chamber view</td>
<td></td>
</tr>
<tr>
<td>6.2 RVOT (1–2 planes)</td>
<td></td>
</tr>
<tr>
<td>6.3 LVOT (1–2 planes)</td>
<td></td>
</tr>
<tr>
<td>6.4 Branch pulmonary arteries</td>
<td></td>
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<tr>
<td>6.5 Aortic arch</td>
<td></td>
</tr>
<tr>
<td>6.6 Targeted slices to address vascular stenoses</td>
<td></td>
</tr>
<tr>
<td>7 Phase-contrast velocity mapping</td>
<td></td>
</tr>
<tr>
<td>7.1 Main PA through-plane</td>
<td></td>
</tr>
<tr>
<td>7.2 Ascending aorta through-plane</td>
<td></td>
</tr>
<tr>
<td>7.3 Vascular stenosis in-plane or through-plane</td>
<td></td>
</tr>
<tr>
<td>8 2D multi-slice black-blood imaging (if previous scanning was unsatisfactory)</td>
<td></td>
</tr>
<tr>
<td>8.1 Branch pulmonary arteries</td>
<td></td>
</tr>
<tr>
<td>8.2 Aortic arch</td>
<td></td>
</tr>
<tr>
<td>8.3 Vascular stenosis</td>
<td></td>
</tr>
</tbody>
</table>

LVOT: left ventricular outflow tract; MRA: magnetic resonance angiography; PA: pulmonary artery; RVOT: right ventricular outflow tract; SSFP: static steady state free precession.

steady state free precession (SSFP) or black-blood scans as needed. Flow studies in the pulmonary trunk and ascending aorta are routinely performed to screen for or quantify any shunting, and a short-axis cine 2D-SSFP stack for volumetric quantitative ventricular function is generally required. In very complex cardiac malformations, it may be wise to add a stack of cine 2D-SSFP slices covering the whole heart, thus providing detailed morphology and quantitative ventricular function. Although there may be variation in exact scan preferences among different centers, the goals of the examination will likely be the same.

Pre-repair planning
Not uncommonly, patients with complex lesions present beyond infancy when the initial management had been surgical palliation in order to avoid a high mortality risk from early repair. These patients may then not only have more limited acoustic windows but also additional associated complex pulmonary or aortic vascular malformations and/or vascular obstructions related to the initial palliation.

Here, CMR has become an invaluable planning tool and has replaced diagnostic cardiac catheterization in many centers. In particular, stacked cine 2D SSFP slices across the heart in transverse and/or coronal orientation as well as 3-dimensional CMR imaging and volume rendering models based on this information are very helpful. 3D contrast-enhanced CMR angiography will allow delineation of thoracic vascular territories such as the pulmonary vessels, the aorta and the systemic veins. For additional information on intracardiac morphology, 3-dimensional “whole-heart” steady-state free-precession CMR imaging (3D-SSFP) covering the whole mediastinum has been shown to be highly reliable. This volume scan can be acquired either as single-phase (end-diastole or end-systole) [32] or dual-phase data set (end-diastole and end-systole) [33]. Respiratory motion correction is achieved by use of a prospective navigator to monitor diaphragm motion. Advanced post-processing tools such as a virtual incision-making, based on reconstruction of 3D CMR images, may be helpful to visualize complex anatomy pre-operatively. The intracardiac morphology can be inspected from any desired surgical view as outlined in Figure 17.1. Coronary artery anatomy as displayed by 3D-SSFP [34] can be integrated into such models. Alternatively, casts may be printed based on such 3D-SSFP data and these can then be used for both planning of operations or interventions as well as for teaching (Figure 17.2) [35].

Post-repair follow-up
As described above, conduits and baffles are not exempt of complications although materials and surgical techniques are being improved. The congenital heart defects that need such repair tend to be complex and thus more difficult to manage. Hence, cautious and thorough post-repair follow-
up in these patients is mandatory. Visualization of conduits and baffles by echocardiography may suffer from limited acoustic windows as baffles may be placed remotely from the transducer and conduits may run behind the sternum and thus be inaccessible to the ultrasound beam. Cardiac catheterization including cine-cardioangiography has been the traditional gold standard method in this scenario, but it is invasive and also exposes the patient to ionizing radiation. The latter is considered particularly harmful in children who have a higher associated tumor risk and who often need repeated investigations for baffle/conduit performance during their life time. Moreover, both echocardiography and cine-cardioangiography are limited in terms of providing reproducible quantitative information on ventricular function.

Figure 17.1 Planning double outlet right ventricle (DORV) repair – Virtual incision tool. Double outlet right ventricle (DORV) with one subaortic and another large muscular ventricular septal defect (VSD) plus pulmonary stenosis and a persisting left superior vena cava (LSVC). Reformatted 2D-plane from a 3D-SSFP data set showing the relation of the subaortic VSD to the aorta (a); corresponding plane showing the segmented myocardium (b), green color. Myocardial tissue properties are added and a virtual incision tool is created, allowing the surgeon to expose selected surgical views through interactively defined incisions. The virtual incision tool allows visualization of the spatial relation of both VSD to the aorta (c) as well as the pulmonary stenosis (d) through dedicated cuts into the model. (Courtesy: Thomas Sangild Sørensen, MSc, PhD, Department of Computer Science and Institute of Clinical Medicine, Aarhus University, Denmark.) Ao: aorta; LV: left ventricle; MPA: main pulmonary artery; RA: right atrium; RV: right ventricle; SVC: superior vena cava.
In this context, the unique properties of CMR render the method an indispensable complementary imaging tool in patients with baffles and conduits [13,36]. The main aims are to detect graft failure such as residual single or multi-level narrowing, leakage involving residual shunting or incompetence in the case of a valved conduit, and to quantify the hemodynamic impact of such findings including quantification of ventricular function, mass and flows. Thus, CMR can help to decide not only on the type of re-intervention, but also, most importantly, on the optimal timing [37,38]. One of the few limitations of CMR in follow-up evaluation is image degradation due to ferromagnetic components in the supporting framework of conduit valves which appear as a loss of signal. This loss of signal is evident in spin echo images, but even more accentuated in gradient-echo based sequences, including cine phase-contrast MR flow. However, with a combination of imaging sequences clinically interpretable scans are possible. As a result, CMR has replaced cardiac catheterization for follow-up purposes in many tertiary centers. Specific examples and suggested approaches to imaging assessment with CMR of various conditions are discussed below. Of course, there can be variation in surgical practice and access to surgical operative notes together with discussion with clinical colleagues is invaluable in image planning and interpretation in such cases.
Baffles

“Veno-atrial” baffles: anomalous pulmonary venous connections

Patients with partial anomalous pulmonary venous connections (PAPVC) may require a veno-atrial baffle to redirect blood flow to the left atrium. This is the case in PAPVC from the right lung to the superior vena cava (SVC) as outlined in Figure 17.3a. In Scimitar’s syndrome there is anomalous pulmonary venous connection of the right lung usually through the diaphragm to the inferior vena cava. This condition is associated with various degrees of pulmonary sequestrations and right pulmonary hypoplasia. For these defects, the surgical treatment consists of forming a patch tunnel, usually from autologous pericardium, to redirect the pulmonary venous blood across an atrial septal defect towards the left atrium; alternatively, an ASD will be created for this purpose. Occasionally, in PAPVC to the SVC, a patch plasty of the SVC will help to widen the lumen where the patch tunnel passes below.

CMR evaluation includes assessment of systemic venous and baffle patency, quantitative ventricular function and exclusion of residual left-to-right shunting across a baffle leak or residual PAPVC as outlined in Table 17.2. Should there be evidence for pulmonary venous congestion due to baffle obstruction or obliteration, then the differential pulmonary perfusion should be quantified using phase-contrast through-plane velocity mapping separately in each branch pulmonary artery and may demonstrate reduced flow into the affected lung. Figure 17.3 gives an example of a patient with baffle narrowing repair of anomalous pulmonary venous connection of the right upper lobe pulmonary vein to the superior vena cava.

“Intra-atrial” baffles: transposition and heterotaxy syndromes

Intra-atrial baffles are used to redirect blood flow in a number of important cardiac defects.

- The most well-known condition is d-transposition of the great arteries (d-TGA), where there is atrioventricular concordance but ventriculo-arterial discordance (Figure 17.4a). The aorta generally arises anterior and rightward from the right ventricle and the pulmonary artery posterior and leftward from the left ventricle and the great vessels run parallel to one another. Until the mid-1980s, surgical repair was performed with an atrial level switch (Senning or Mustard procedure), in order to direct oxygenated pulmonary venous blood to the right ventricle in systemic position and de-oxygenated blood to the subpulmonary left ventricle (Figure 17.4b). The Mustard operation uses mostly autologous pericardium whereas the Senning operation utilizes the atrial septum itself to create the baffle.
Another important condition where atrial baffles are utilized is the repair of associated defects in congenitally corrected transposition of the aorta (CC-TGA) where there is atrioventricular discordance combined with ventriculo-arterial discordance which result in normal physiologic flow, however, the right ventricle (RV) pumps to the aorta and the left ventricle (LV) pumps to the subaortic morphological right ventricle which supports the systemic circulation.

### Table 17.2 Veno-atrial and intra-atrial baffles.

<table>
<thead>
<tr>
<th>Goals of the MRI examination</th>
<th>Suggested MRI protocol</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Systemic venous obstruction</td>
<td>1. Transverse stack of multi-slice 2D cine SSFP slices to evaluate quantitative RV and LV volumes, AV valve function and screen for baffle patency (or: 4-chamber view + short-axis ventricular volumes)</td>
</tr>
<tr>
<td>• Pulmonary venous obstruction</td>
<td>2. Gadolinium-enhanced 3D MRA in 2 subsequent dynamics, tailored (1) to pulmonary venous return through baffle plus aortogram, and (2) to systemic venous return plus pulmonary arteries</td>
</tr>
<tr>
<td>• Baffle leak</td>
<td>3. Whole-heart 3D SSFP volume scan (isotropic resolution)</td>
</tr>
<tr>
<td>• Tricuspid valve regurgitation</td>
<td>4. Consider: Late gadolinium-enhancement (LGE) studies for evaluation of the baffle and the ventricles (fibrosis/scarring)</td>
</tr>
<tr>
<td>• VSD</td>
<td>5. Phase-contrast quantitative flow (through-plane) of the aorta and the pulmonary artery to screen for residual left-to-right shunt across a baffle-leak (or associated/residual VSD)</td>
</tr>
<tr>
<td>• Subarterial obstruction</td>
<td>6. Review available anatomical information and decide if more scanning is necessary: Angulated single 2D SSFP or black-blood slices across suspicious parts of the baffle</td>
</tr>
<tr>
<td>• Bi-ventricular analysis: volumes, mass and function</td>
<td></td>
</tr>
</tbody>
</table>

AV: atrioventricular; LV: left ventricle; MRA: magnetic resonance angiography; RV: right ventricle; SSFP: static steady state free precession; VSD: ventricular septal defect.

[2,17]. Although atrial redirection surgery has been gradually replaced by the arterial switch operation to avoid leaving the right ventricle in a systemic position, there is now a considerable adult population with need for regular follow-up including serial CMR assessment of venous pathway patency and right ventricular function utilizing dark blood and cine imaging.

**Figure 17.4** D-TGA and atrial baffle repair. (a): Atrioventricular concordance and ventriculo-arterial discordance with transposition of the great arteries (d-TGA) leading to cyanosis. (b): Surgical repair (Mustard, Senning) using an atrial baffle to re-direct (green arrows) the de-oxygenated blood from the systemic veins to the subpulmonary morphological left ventricle and oxygenated blood from the pulmonary veins to the subaortic morphological right ventricle which supports the systemic circulation.
or pulmonary venous anomalies, there may be the need for atrial baffles to separate systemic venous, hepatic venous and pulmonary venous connections at atrial level in order to overcome cyanosis either for biventricular repair or single-ventricle Fontan-type palliation, as displayed in Figure 17.6 (see below under “Baffle or conduit in Fontan surgery: variants of TCPC”, and the chapter in this book on “The single ventricle”).

The goals of the post-operative CMR evaluation of intra-atrial and veno-atrial baffles are summarized in Table 17.2. In all these examples, CMR provides excellent information on quantitative right ventricular function, atrioventricular valve regurgitation, systemic venous or pulmonary venous pathway patency and baffle leaks [39].

From a practical point of view, the following imaging strategy may be considered:

1. Start with a stack of transverse 2D cine SSFP slices covering the whole heart, in order to look
for atrial anatomy, possible thrombi, tunnel patency, AV valvular function and quantitative ventricular function. Alternatively: “4-chamber” view and short-axis 2D cine-SSFP stack for ventricular volumetry, combined with transverse and coronal cine-SSFP stacks to image the baffle.

2. Perform a contrast-enhanced 3D MR angiogram with two subsequent dynamics (sets of 3D images) of <10–12 sec each. Tailor the first dynamic to display the pulmonary venous return through the atrial baffle (Figure 17.7d). This dynamic will usually also display the aorta. If the duration of each dynamic is kept short enough this will usually give excellent subsequent delineation of the systemic venous baffle return to the subpulmonary ventricle and the pulmonary arteries.

3. Consider if late gadolinium enhancement viability imaging of the myocardium or baffle would be useful (see below).

4. Perform a free-breathing isotropic 3D whole-heart SSFP [32] as this is highly valuable to identify baffle obstructions (Figure 17.7c,e). The scan is easy to plan and usually runs for 6–10 minutes depending on coverage, resolution, heart rate and navigator efficiency. This technique has the advantage of easier scanning and free image reformatting in any desired plane during post-processing, but does not provide cine information such as flow turbulences from obstructions.

5. Determine the quantitative flow rate in the great arteries to screen for residual shunting due to baffle leak or associated/residual ventricular septal defect (VSD). It is tempting to acquire a single slice as both arteries are somewhat parallel and flow in each great artery in the same timeframe can be compared, but separate scans may still give better alignment perpendicular to flow and furthermore allow for individualized setting of VENC in the case of outflow tract obstructions.

6. At this point, while running the flow studies, transfer the 3D data to a post-processing workstation and review for pathology. Decide if the anatomical information obtained so far is good enough or if subsequent 2D slices to further delineate a problem anatomically are necessary.

   Consider:
   1. 2D cine-SSFP slices for visualization of dynamic narrowing and turbulences in baffles or outflow tracts.
   2. 2D black-blood imaging for additional anatomy (rarely necessary after the above scans).
   3. 2D velocity mapping slices may be used in areas of obstruction both in-plane or through-plane to assess peak jet velocity and estimate the

---

**Figure 17.6** Heterotaxy syndrome (left-atrial isomerism) – Fontan palliation involving an intraatrial tunnel directing hepatic venous blood to the left pulmonary artery.
(a): 3D reconstruction of the heart confirming situs inversus, dextrocardia and heterotaxy syndrome with no inferior vena cava (IVC) visualized and azygos continuity to the superior vena cava (SVC). The hepatic veins were tunneled towards the left pulmonary artery (LPA). The right sided azygos vein drains into the right superior vena cava (RSVC), which had been previously anastomosed to the right pulmonary artery (RPA) by means of a bidirectional Glenn shunt (source images: 3D CE-MRA).
(b): This second 3D reconstruction illustrates the RSVC and azygos vein (blue) being connected to the RPA and the hepatic venous tunnel to the LPA forming the Fontan-type circulation in this patient (source images: 3D CE-MRA). (c): Image reformatted from 3D CE-MRA data set, demonstrating the intracardiac course of the hepatic-venous to LPA intracardiac tunnel (arrow). Hepat.: hepatic; PA: pulmonary artery.
Figure 17.7 D-TGA after atrial baffle redirection surgery. 
(a) and (b): 3D segmentation demonstrating d-TGA (= atrioventricular concordance and ventriculo-arterial discordance) after atrial redirection repair. 
(c) and (d): Systemic venous baffle pathway (c), (3D SSFP reformat) and pulmonary venous baffle pathway (d), (3D CE-MRA reformat). 
(e) and (f): Obstruction (arrow) of the upper SVC baffle limb (c), (3D SSFP reformat) and venous bypass through the azygos vein caudally (d), (3D CE-MRA reformat). 
Ao: aorta; Az: azygous; IVC: inferior vena cava; LV: left ventricle; PA: pulmonary artery; PVAC: pulmonary venous atrial chamber; RA: right atrium; RV: right ventricle; SVC: superior vena cava.
CHAPTER 17 Baffles and conduits

pressure gradient using the modified Bernoulli equation [40].

4. Separate branch pulmonary artery quantitative flow studies in case of unilateral pulmonary venous baffle obstruction, to further assess its hemodynamic impact.

5. Review if the azygos vein was imaged appropriately by one of the flow scans and check if there is flow reversal. Consider to repeat a dedicated flow scan to screen for this if above information is unsatisfactory.

Systemic venous pathway obstruction seems more common with the Mustard operation and is mostly seen in the superior vena cava (SVC) limb (Figure 17.7e), whereas pulmonary venous obstruction appears more commonly after the Senning repair. Significant atrial baffle obstruction should be diagnosed when one of the following criteria is found: Apparent narrowing of the baffle pathway on contiguous images associated with downstream signal loss from turbulent flow towards the atrioventricular valve on 2D cine SSFP images and a peak jet velocity of 1.5 m/sec or greater in the direction of the atrioventricular valve at cine velocity mapping [41]. Upstream venous dilatation due to congestion may be particularly easy to demonstrate when using contrast-enhanced 3D MR angiography (3D CE-MRA) or 3D-SSFP, including bypass run-off flow to the inferior vena cava (IVC) territory over the azygos vein (Figure 17.7f). Dedicated flow mapping scans with VENC set at 80 centimeters/second (cm/sec) will prove such flow reversal in the dilated azygos vein. However, dilatation of the azygos–hemazygos venous system may also be observed in patients with azygos continuation of an interrupted IVC, congestive/restrictive heart failure or portal hypertension. Late-gadolinium enhancement of the myocardium (Figure 17.8) was described in adult patients with d-TGA after atrial redirection surgery and related to adverse outcome variables [42,43]. Such viability imaging may be considered on an individual basis.

"Intra-ventricular baffles": Rastelli and Rastelli-like patch tunnel to connect LV with the aorta

A number of complex cardiac defects with conotruncal abnormalities of the origin of the great arteries need redirection surgery at ventricular level to connect the morphological left ventricle with the aorta.

- Double outlet right ventricle (DORV), is a complex congenital heart disease where both great arteries arise from the right ventricle, with at least one semilunar valve not in continuity with an atrioventricular valve, and a ventricular septal defect is the only outlet of the left ventricle. Pulmonary or subpulmonary stenosis may be present or absent (Figure 17.9a). Hemodynamics and surgical management plan depend on:
  - the position of the great arteries to each other
  - the position of the great arteries in relation to the position of the ventricular septal defect (VSD) which represents the outlet out of the left ventricle (Figures 17.1 and 17.2)
  - the presence and severity of outflow tract obstructions towards the aorta or the pulmonary artery
  - the presence of associated defects.

In the subgroup of DORV with subaortic VSD and side-by-side position of the great arteries (aorta right-lateral to the pulmonary artery) with or without pulmonary stenosis, repair may be performed in infancy patch-tunneling the subaortic VSD with the subaortic outflow tract (Figure 17.9b and 17.10). This tunnel may be subject to obstruction at either end, and in some patients the VSD needs to be enlarged. Additionally, relief of a pulmonary obstruction by resection and right ventricular outflow tract (RVOT) patch or a valved conduit may be necessary. Even if there is no pulmonary stenosis, the VSD patch tunnel may encroach on the subpulmonary outflow tract resulting in RVOT obstruction which needs to be addressed during follow up. In other forms of DORV where the VSD is in a subpulmonary position (Taussig–Bing hearts) or remote from the great arteries such as with a posterior muscular or inlet VSD, a patch tunnel cannot be performed.

- An important example is d-TGA with large outlet VSD and subpulmonary and/or pulmonary valve obstruction. In this constellation an arterial switch operation may not be possible, and simple VSD patch closure with relief of the pulmonary obstruction would leave the RV in the systemic position. In the Rastelli procedure, a Dacron baffle is formed to create a patch-
In congenital corrected transposition of the great arteries (CC-TGA) with large outlet VSD and subpulmonary and/or pulmonary valve obstruction there is not only a need to redirect flow at atrial level with baffle surgery (see above) but also to connect the morphological left ventricle to the aorta, and a conduit is inserted to connect the right ventricle to the pulmonary artery (Figure 17.10). A more recent development for repair is the Nikaidoh procedure where there is aortic translocation.

Figure 17.8 Late gadolinium enhancement in the systemic RV after atrial redirection surgery (Mustard or Senning). (a): Image showing thinning of free wall of the systemic RV (dotted arrow) which is hypertrophied and dilated with impaired systolic function decades following Mustard surgery for transposition of the great arteries. The jet of mild AR (block arrow) is also visible from this still frame taken from SSFP cine imaging. (b): The corresponding late gadolinium image shows full thickness late enhancement in the free wall of the RV (black arrow) and also endocardial enhancement extending over the systemic RV myocardium (white arrows). (c): In a different patient there is also free wall RV late enhancement (dotted arrow) and in addition, there is late enhancement associated with previous VSD patch closure (block arrow). (d): There is late enhancement in the RV free wall (dotted arrow). The presence of such small and or patchy late enhancement must be confirmed in phase swaps, cross cuts and different slices and compared with corresponding cines to ensure that artifacts are not over-reported. The insertion points where the RV inserts into the LV (block arrows) are a common finding in late gadolinium studies of systemic RV and do not appear to correlate with adverse clinical outcomes. Abbreviations are as previously stated in the chapter. Ao: aorta; LV: left ventricle; RA: right atrium; RV: right ventricle.
Figure 17.9 Diagram of double outlet right ventricle with ventricular septal defect (VSD) – intraventricular tunnel repair. (a): Double-outlet right ventricle with (subaortic) large VSD. (b): Surgical repair using an intraventricular baffle to create a tunnel (large green arrow) from the left ventricle through the VSD (being the only outlet of the left ventricle) to the aorta.

Figure 17.10 DORV with VSD and pulmonary stenosis after Rastelli repair. (a): Still frame from SSFP cine after Rastelli-type repair of DORV with VSD and pulmonary stenosis. The conduit is narrowed to 13 millimeters (mm) proximally and 8 mm where most narrowed. The peak recorded velocity was 3.5 meters/second (m/sec). The associated signal loss from the conduit jet is represented by the arrow in this still frame from SSFP cine imaging. There is moderate pulmonary regurgitation with a pulmonary regurgitant fraction 33%. (b): This illustrates the patch (arrows) VSD closure connecting the LV to the aorta. Abbreviations are as previously stated in the chapter.
tricle with the aorta (Figure 17.5). An arterial switch operation may then be impossible, and simple VSD patch closure with relief of the pulmonary obstruction would leave the RV in the systemic position, which is not the preferred long-term option. The desired additional redirection on ventricular level may be successfully achieved by a Rastelli-type operation. As mentioned above this includes an intraventricular patch-tunnel to close the VSD which will baffle the morphological left ventricle to the aorta, and a right ventricle to pulmonary artery conduit (as discussed below) is inserted allowing the RV to be subpulmonary and the systemic ventricle to be the morphological left ventricle.

CMR with its inherent 3-dimensional nature can be very helpful for planning of the procedure. CMR protocols may be chosen from Table 17.1. In particular, this is the case in DORV as it is often difficult to appreciate the 3-dimensional anatomy using 2-dimensional imaging (Figure 17.2). The main issue here often is the size, shape and position of the VSD in relation to the great arteries in order to decide whether an intracardiac repair is feasible in an individual patient. Models created from 3D CMR data can be very helpful in these rare but demanding patients. Both casts printed from 3D CMR data [35] (Figure 17.2) and computer-segmented models together with advanced visualization such as virtual incision tools [44] (Figure 17.1) can assist the surgeon to better plan the optimal approach well in advance of the procedure rather than subjecting the patient to on-table decisions based on inspection of the anatomy. This is particularly appreciated when there are additional issues complicating the picture such as cardiac situs anomalies or unusual spatial relations such as dextrocardia. Another important aspect is that 3D-SSFP “whole heart” morphological imaging can also display coronary anomalies [34]. This may be relevant if ventricular incisions are necessary to reconstruct the RVOT in associated outflow tract obstructions (Figure 17.11).

In the post-repair follow-up, CMR is a valuable tool to assess the precise nature and localization of a tunnel complication and to quantify its hemodynamic effect. To visualize the patch tunnel, angulated 2D cine SSFP slices are suited (Figure 17.10) and will demonstrate turbulence due to fixed or dynamic obstructions, which represent subaortic obstructions after the repair. 3D SSFP scanning performed as single- or dual phase acquisition may help in understanding the tunnel geometry based on segmentation. Velocity mapping using targeted single slices can determine the jet velocity and hence the significance of a given tunnel obstruction. Moreover, biventricular quantitative function and mass by CMR can be followed serially and may constitute important information regarding the timing of a re-intervention. Through-plane flow imaging in the great arteries can quantify a residual shunt across a tunnel leakage. Flow and volumetry data can be combined to quantify atrioventricular (AV) valve incompetence. The CMR protocols to achieve these goals may be used as outlined in Tables 17.1 and 17.2.

Baffle or conduit in Fontan surgery: variants of the total cavo-pulmonary connection (TCPC)

Univentricular hearts represent a wide variety of structural cardiac abnormalities associated with a “functional single ventricular chamber” [45]. The term “univentricular heart” should include double inlet left and right ventricles, absence of one AV connection (Figure 17.12a), common AV valves with only one well developed ventricle and complex conditions with heterotaxy syndromes and ventricular imbalance [46]. For more details and pathophysiology please refer to the chapter in this textbook which deals with the single ventricle. Whatever the underlying pathology and initial palliation to achieve sufficient systemic or pulmonary perfusion, the surgical management finally aims to convert a parallel to a serial circulation. The final stage of such conversion is a Fontan-type reconstruction where all systemic venous blood is separated from the heart and directed towards the pulmonary arteries. In contemporary practice, the generally accepted approach is to achieve this in two steps, namely connecting the superior vena cava (SVC) and subsequently the inferior vena cava (IVC) to the undivided right pulmonary artery (RPA) in the typical case, thus creating a “total cavo-pulmonary connection” (TCPC) [47]. In most patients, a bidirectional Glenn or hemi-Fontan operation will be performed as the first step towards a Fontan circulation. Whereas the Glenn procedure simply connects the SVC end-to-side to the RPA, the hemi-Fontan operation is more complex as it involves a connection between the
In the lateral tunnel Fontan operation (Figure 17.13), a previous hemi-Fontan procedure is preferred by some surgeons as this makes surgery for the lateral tunnel easier. The intracardiac baffle (usually PTFE) used in the lateral tunnel operation retains growth potential as the posterior wall of the tunnel from the IVC to the RPA is formed of atrial and septal wall tissue. In most upper roof of the right atrium and the RPA together with a patch sealing off the SVC flow from the remainder of the right atrium. For the final stage, two major TCPC-modifications exist where either an intra-atrial baffle (Figure 17.12b) [47] or an extracardiac conduit (Figure 17.12c) [48] is used to direct the systemic venous blood from the IVC and hepatic veins towards the RPA.

Figure 17.11 DORV with large subarterial VSD and side-by-side position of the great arteries. (a): Reformatted 3D-SSFP image demonstrating side by side relationship of the great arteries, subpulmonary VSD and subpulmonary stenosis. (b): Reformatted 3D-SSFP image showing the left-anterior descending coronary artery (LAD) originating from the right coronary artery to cross over the right ventricular outflow tract. (c) and (d): Pre-operation planning revealing the relationship of the VSD to the great arteries. AAo: ascending aorta; Ao: aorta; LV: left ventricle; PA: main pulmonary artery; RV: right ventricle; RVOT: right ventricular outflow tract; VSD: ventricular septal defect.
cases, a fenestration will be placed in the baffle to allow shunting of deoxygenated blood to the pulmonary venous atrium and hence the systemic circulation which has been found to improve clinical outcomes (maintaining cardiac output at the expense of cyanosis) [49].

- In extracardiac conduit Fontan repair (Figure 17.14), a conduit is formed usually of a PTFE (Goretex®) tube or more rarely equine or autologous pericardium to connect the IVC and hepatic venous blood with the right pulmonary artery, rather than using an intraatrial baffle. A fenestration which is a connection between the systemic and pulmonary venous pathways allowing blood to shunt from right to left is also performed. In this scenario, a prior hemi-Fontan
operation has no advantages and a bidirectional Glenn is usually preferred. Potential advantages of this variant of TCPC include avoidance of extensive suturing within the atrium as necessary for the intracardiac lateral tunnel. Moreover, the procedure is thought to have improved hemodynamic performance as there may be less irregularity of tunnel geometry [50] which is hoped to result in better flow dynamics. The main concern is lack of conduit growth with the available material. Hence this ultimate step of the Fontan palliation can only be performed when the patient size allows for insertion of a large enough conduit to avoid replacement surgery in adolescence. Another potential problem is conduit stenosis due to peel formation and thrombosis in con-

**Figure 17.13** “Lateral tunnel” (LT) Fontan operation using an intra-atrial baffle. (a), (b) and (c): Sagittal (a), (c) and frontal (b) views reformatted from a 3D SSFP data set to demonstrate the intra-atrial baffle connecting the inferior venous blood to the right pulmonary artery (RPA) in a lateral tunnel Fontan operation. Note the irregular configuration of the lateral tunnel. (d): Transverse view reformatted from the same 3D SSFP data set to demonstrate the unobstructed branch pulmonary arteries in this patient. Ao: aorta; IVC: inferior vena cava; LPA: left pulmonary artery; PA: pulmonary artery.
to the pulmonary arteries is a simple and effective way to address various issues in a single scan:
• Atrial anatomy, including possible thrombi
• Conduit/baffle patency and fenestration, including possible thrombi
• Pulmonary venous obstructions and possible restrictive atrial septal defect, particularly the intracardiac lateral tunnel Fontans when the pulmonary venous blood is required to travel from the left side towards a right-sided AV valve inlet to the single ventricle (such as with classical hypoplastic left heart syndrome (HLHS)-Fontan)
• AV valvular function
• Ventricular geometry and quantitative ventricular function.

2. After this, 3D CE-MRA (Figure 17.6) and subsequently 3D whole-heart SSFP (Figure 17.13) will be advantageous as a large volume can be covered in a relatively short space of time. For 3D CE-MRA, a good approach is to plan 2–3 subsequent dynamics (volume data sets) of <10–12 sec each. The contrast injection should occur preferably through an arm vein as a foot vein approach may not be large enough.

Figure 17.14 Extra-cardiac conduit Fontan operation. (a) and (b): Frontal (a) and transverse (b) views from angulated 2D SSFP cine slices to demonstrate patency of the extra-cardiac Fontan tunnel (arrows) from the inferior vena cava to the right pulmonary artery. The underlying anatomy is double-inlet left ventricle (DILV) with ventriculo-arterial discordance and L-transposition of the aorta. The ventricular septal defect (VSD) is very large (*) and does not present a subaortic obstruction. Ao: ascending aorta; ASD: atrial septal defect; Des Ao: descending aorta; PA: native (ligated) main pulmonary artery; RV: right ventricular outlet chamber.
Table 17.3 “Lateral tunnel” and “Extracardiac conduit” TCPC (Fontan).

<table>
<thead>
<tr>
<th>Goals of the MRI examination</th>
<th>Suggested MRI protocol</th>
</tr>
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<tr>
<td>• Assessment of the pathways from the systemic veins to the pulmonary arteries for obstruction, dilatation and thrombus</td>
<td>• Transverse stack of multi-slice 2D cine SSFP slices to evaluate quantitative RV and LV volumes and screen for baffle patency (or: 4-chamber view + short-axis ventricular volumes + separate transverse scan for baffle patency)</td>
</tr>
<tr>
<td>• Detection of hemi-Fontan or tunnel baffle fenestration or leaks</td>
<td>• Gadolinium-enhanced 3D MRA</td>
</tr>
<tr>
<td>• Evaluation of the pulmonary veins for compression</td>
<td>• 3D SSFP whole-heart volume scan (isotropic resolution)</td>
</tr>
<tr>
<td>• Bi-ventricular analysis: volumes, mass and function</td>
<td>• Additional angulated single 2D SSFP and 2D flow slices across the lateral tunnel</td>
</tr>
<tr>
<td>• Ventricular outflow tract obstructions</td>
<td>• Angulated flow velocity mapping (2D cine phase-contrast) in a plane aligned with the expected location of a jet</td>
</tr>
<tr>
<td>• Quantitative analysis of valve regurgitation</td>
<td>• Phase-contrast quantitative flow (through-plane) of the aorta and all Fontan pathways, i.e. branch pulmonary arteries, SVC and IVC/baffle to screen for residual left-to-right shunt across a baffle-leak</td>
</tr>
<tr>
<td>• Assessment of aortic arch: obstructions and aneurysm.</td>
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<tr>
<td>• Collateral vessels: aorto-pulmonary, systemic venous or systemic to pulmonary venous collaterals</td>
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Abbreviations as defined previously in this chapter.

result in dilution of the contrast bolus in the IVC resulting in impaired contrast-to-noise on venous imaging. The first dynamic is tailored to display the aorta for Daymus-Kaye-Stansell (DKS) connection, arch patency and aorto-pulmonary collaterals. The second dynamic will usually show the systemic venous return (patency, collaterals, and thrombi), intra-atrial baffle and the pulmonary arteries well if the dynamic duration is short enough. Only rarely two separate injections in an arm vein and a foot vein may be needed to image the baffle-tunnel and the pulmonary arteries. Alternatively, time-resolved CE-MRA has been suggested using parallel imaging and keyhole undersampling to image both the Fontan pathways and also qualitatively assess the pulmonary perfusion, using consecutive injections in arm and leg veins, depending on the number and site of the veno-pulmonary connections [52].

3. Free-breathing isotropic 3D whole-heart SSFP [32] is an excellent technique to identify lateral tunnel and veno-pulmonary pathway obstructions and thrombi. The scan is easy to plan and usually runs for 6–10 minutes depending on coverage, resolution, heart rate and navigator efficiency. This technique has the advantage of simple scanning and free image reformating in any desired plane during post-processing, but does not provide cine information such as flow turbulences from obstructions. Alternatively, spin-echo and double-inversion-recovery “black-blood” fast spin-echo sequences are less favorable for baffle anatomy as intraluminal saturation due to slow flow may render the baffle and veno-pulmonary pathway images sometimes difficult to analyze.

4. Additional single coronal or sagittal 2D SSFP cine slices (Figure 17.14) may show swirling and turbulence of blood flow in patients with an obstructed or dilated Fontan connection and may also detect intracardiac baffle leaks.

5. 2D phase-contrast velocity mapping studies are of key importance in the assessment of TCPC hemodynamics as they quantitate flow rate in both pulmonary arteries [53], the SVC, the IVC (caudal end of the baffle tunnel) and the ascending aorta (cardiac index) [54]. For flow studies, it is generally advisable to use free-breathing scanning in cooperative patients but breath-hold scans in ventilated children, because any increase in intrathoracic pressure will compromise the non-pulsatile flow in these conditions and may produce biased results.

Beyond quantification of volume flow rate, 3-dimensional patterns of flow are particularly interesting in Fontan pathways. This can be assessed more reliably by use of novel free-breathing volume velocity mapping techniques, with flow encoded in all three directions of space and sampled over time (“4D flow”). It has been demonstrated in an elegant study that blood flow pat-
terns are more organized and uniform in TCPC than in atrio-pulmonary Fontan pathways and are significantly influenced by pathway diameter suggesting that TCPC may result in a more hemodynamic-efficient circulation [55]. Please see chapter on single ventricles regarding the numerous studies published regarding 4-dimensional flows in the systemic venous pathway by the collaboration of the engineers at the Georgia Institute of Technology and the CMR cardiologists at The Children’s Hospital of Philadelphia. These support initial reports from hydrodynamic data provided by de Leval and co-workers in the paper introducing TCPC as a new concept [47]. Disadvantageous flow characteristics increase with pathway dilatation. Factors that influence the flow dynamics of Fontan circulations to consider include: type of Fontan operation, offset between cavopulmonary connections, flaring of the cavopulmonary connection, conduit and vessel stenosis, pulmonary vascular resistance, conduit and vessel compliance, fenestration, and pulmonary artery curvature. It can be expected that multidimensional quantitative CMR flow mapping will add important information on baffle physiology. It may become possible to identify unfavorable flow patterns such as turbulences in dilated baffles that may result in energy losses and increased resistance to net forward flow. Please see the end of the chapter on single ventricles regarding the studies of this possibility. The accuracy of 4D flow may be further improved when using novel techniques for respiratory motion compensation [56].

Conduits

Ventricular to pulmonary artery conduits

Two different clinical situations need consideration when discussing conduit applications in congenital heart disease.

1. Implantation of a valved or non-valved conduit may be needed for initial corrective surgery in young children. This is necessary in various forms of congenital heart defects where there is no natural ventriculo-pulmonary connection such as with an atretic pulmonary valve, or when there is no main pulmonary artery at all. Moreover, a ventriculo-pulmonary conduit may be needed in lesions with severe subpulmonary/pulmonary valve stenosis, where simple resection and RVOT patch is impossible due to an anomalous coronary artery traversing across the RVOT (Figures 17.11 and 17.15d). It is usually the goal to connect the RV to the pulmonary artery, but this may occasionally also involve connecting the morphological LV with the pulmonary artery such as with CC-TGA, VSD and pulmonary stenosis when a double switch/Rastelli-type operation is considered unfeasible (Figure 17.16).

2. Valved conduits may simply serve as pulmonary valve replacement when a biological valve is preferred later in life, often as a re-intervention after initial repair. Conduit failure from these types of conduits in later follow-up may be amenable to percutaneous approaches to relief of homograft/conduit stenosis and restoration of pulmonary competence. In percutaneous pulmonary valve implantation (PPVI), a stent with an integrated bovine jugular venous valve graft is used to treat RVOT and pulmonary valve obstruction and/or regurgitation. (Figure 17.15a,b) This procedure needs careful patient selection to avoid device embolization from dilated outflow tracts [15].

Important examples of corrective surgery needing implantation of a right ventricular-to-pulmonary conduit are listed below.

- In pulmonary atresia with intact ventricular septum, antegrade flow into the main pulmonary artery may be established using a homograft or bovine jugular venous graft when the right ventricle is only moderately hypoplastic and the coronary perfusion is not right ventricular-dependent; often an additional aorto-pulmonary shunt is necessary as well.

- Pulmonary atresia with VSD (tetralogy of Fallot-type) and confluent central pulmonary arteries is a classical constellation for a RV-to-PA conduit for which most centers will favor a pulmonary homograft. When there are no confluent central pulmonary arteries, a unifocalization procedure may be needed as a first step.

- Truncus arteriosus variants need an artificial conduit as there is no pulmonary valve available.

- A number of defects with transposition or malposition of the aorta, VSD and severe pulmonary stenosis or atresia will finally undergo a Rastelli or Rastelli-type operation for definitive repair to baffle LV to the transposed aorta and
Figure 17.15 Right-ventricular to pulmonary valved conduits in the management of tetralogy of Fallot.
(a) and (b): The patient had repair of tetralogy of Fallot using a conduit and underwent percutaneous pulmonary valve implantation in the main PA for pulmonary regurgitation with residual pulmonary stenosis in late follow-up. Ferromagnetic artifact from the stent of the valve is seen in (a) (white arrow). Thinning of the myocardium in the RVOT is associated with regional wall motion abnormality. The akinetic RVOT area corresponds to a large area of late gadolinium enhancement seen in (b) and, as is often the case, there is also late enhancement in the area of VSD patch closure (*). The undesirable inherent properties of the unoperated RVOT are, of course, not addressed by percutaneous valve insertion. (c): An RV-to-PA conduit has been inserted to bypass native pulmonary stenosis (PS) in this repair of tetralogy of Fallot. The pulmonary valve (white arrow) can be seen and this is competent. The through plane velocity map of flow (upper right insert in (c)) in the conduit (black arrow) had peak velocity less than 3 m/sec. (d): Due to a large right coronary artery (RCA) branch crossing in front of the RVOT to supply the anterior descending coronary artery, an RV-PA conduit (white arrow) was inserted after patch RVOT enlargement. The native outflow tract (black arrow) is also seen in the view. Both are severely narrowed and peak velocity in the 6 × 7 mm conduit is more than 4.5 m/sec. The patient is unsuitable for PPVI due to its attempt resulting in coronary compression and redo surgery was offered.
Ao: aorta; LV: left ventricle; PA: pulmonary artery; RA: right atrium; RV: right ventricle.
to establish a right ventricle-to-pulmonary continuity by a conduit. As discussed above, the operation was introduced for repair of d-transposition of the great arteries with large outlet VSD and subpulmonary outflow obstruction, but is now also used for DORV variants and for congenitally-corrected transposition of the great arteries (CC-TGA) with VSD and pulmonary stenosis, the latter combined with atrial redirection by Senning or Mustard procedure (see above).
In the group of lesions summarized under the term “hypoplastic left heart syndrome,” there is ductal-dependency of the systemic circulation due to anatomical obstructions or hypoplasia of the subaortic left ventricle. These patients are usually offered the Norwood operation in order to connect the dominant right ventricle to a reconstructed neo-aorta. Reconstruction of the ventricular outflow and the aortic arch is needed. For this, the main pulmonary is anastomosed with the ascending hypoplastic aorta and hence disconnected from the pulmonary bifurcation. Blood flow to the pulmonary arteries is established through a modified Blalock–Taussig shunt (aorto-pulmonary shunt) or some surgeons prefer the “Sano” modification (PTFE-conduit from the RV to the confluence of the pulmonary arteries) instead of a shunt [31]. Adequate mixing at atrial level is ensured by atrial septectomy. See chapter on the single ventricle for more details.

Not uncommonly, CMR is used prior to repair to obtain 3D data sets allowing subsequent 3D segmentations in order to plan a more complex operation involving a conduit. This is the case particularly if it is unclear whether or not a conduit would fit behind the sternum, for instance, in severe pulmonary stenosis in complex lesions, associated situs anomalies and dextrocardia.

Careful post-repair follow-up is mandatory in all these patients as the mid and long-term durability is limited. Please refer to Table 17.4 which summarizes the goals and useful CMR sequences in more detail in order to achieve a thorough investigation after ventriculo-pulmonary conduit surgery [57].

In the more recent management of RV-to-PA conduit failure using percutaneous pulmonary valve implantation (PPVI) techniques, 3D CMR imaging has great value not only in the assessment of the morphology of the right ventricular outflow tract and pulmonary arteries but also in the delineation of the coronary anatomy to avoid coronary compression during PPVI [58]. Cine imaging of the RVOT and PA (Figure 17.16a,c) gives complementary information on the degree of contractility of native RVOT and PA which may also be relevant in case selection. Though suitability and indications may widen, currently available PPVI is most suited to relief of previous conduit failure.

<table>
<thead>
<tr>
<th>Table 17.4 Ventriculo-pulmonary conduits.</th>
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<tr>
<td><strong>Goals of the MRI examination</strong></td>
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<tr>
<td>• Bi-ventricular analysis: volumes, mass and function</td>
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<tr>
<td>• AV valve and aortic valve competence, assessment of cardiac output</td>
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<tr>
<td>• Ventricular outflow tract obstructions</td>
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<tr>
<td>• Detection of conduit stenosis</td>
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<tr>
<td>• Quantitative analysis of conduit regurgitation</td>
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<tr>
<td>• Assessment of pulmonary artery size and possible branch pulmonary artery obstructions</td>
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<tr>
<td>• Pulmonary differential pulmonary blood flow</td>
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<tr>
<td>• Exclusion of dilation of aortic root and ascending aorta and arch anomalies</td>
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Abbreviations are defined previously in this chapter.
Left ventricular “apico-aortic” conduit

Apico-aortic conduits were used to overcome otherwise untreatable complex left ventricular outflow tract obstruction, in order to create a second outlet for the left ventricle by anatomizing a valved conduit from the left ventricular (LV) apex usually to the descending (or rarely ascending) aorta, with or without cardiopulmonary bypass. Material used included Dacron tubes containing glutaraldehyde-preserved porcine pulmonary valves or aortic homograft valves, or a mechanical valve. Frequent problems include conduit obstruction [59], [36], conduit valve dysfunction [60] thrombosis and infective endocarditis [59]. These patients are now only rarely seen as the Ross–Konno procedure has largely replaced this approach, either as primary repair or to treat apico-aortic conduit failure.

Survivors will benefit from CMR as an ideal imaging method to assess conduit complications and quantitative ventricular function and mass. Follow-up of these patients by CMR have been published [36]. Protocols may be chosen from Table 17.1. Stacked axial black-blood imaging and/or reformattting of 3D-SSFP and 3D-CE-MRA volume data sets covering the whole extracardiac course allows the description of the course and patency of the conduit between the left ventricular apex and the descending aorta. Angulated cine imaging (2D-SSFP) will visualize any conduit valve stenosis or regurgitation that may then be quantified by in-plane or through-plane velocity mapping, and also allow imaging of the geometry of the narrowed native left-ventricular outflow tract and ascending aorta when a Ross–Konno operation is considered in case of conduit failure.

Ascending to descending aortic conduit: interrupted aortic arch

Interrupted aortic arch is defined as complete luminal and anatomic discontinuity between two segments of the aortic arch; the thoracic aorta aortic arch may be interrupted at one of three sites: (Type A) interruption located just distal to left subclavian artery; (Type B) interruption located between left subclavian and left common carotid arteries; and (Type C) interruption located between left common carotid and innominate artery. Surgical opinion differs as to whether a one-stage or staged repair is superior. There is preference towards an end-to-side anastomosis avoiding the atretic segment. Alternative methods of arch repair are indicated when a primary ascending to descending aortic anastomosis is not advisable. In 1980, Vijayanagar and colleagues [61] published the surgical approach of treating complex coarctation or atretic aortic segments with an extra-anatomic ascending-to-descending aortic bypass (Figure 17.17). Surgical technique is performed with an end-to-side anastomosis between the descending aorta and a ring reinforced expanded PTFE (= polytetra-fluoro-ethylene) graft. The graft is passed along the diaphragmatic aspect of the right ventricle, anterior to the inferior vena cava, around the right atrium and brought to the ascending aorta.

CMR is the first-line non-invasive method for follow-up imaging as it will be difficult to visualize the full extent adequately by echocardiography, and repeated CT angiograms carry obvious disadvantages from ionizing radiation exposure. The size of the surface coil needs consideration as in many adult patients, a cardiac surface coil may not provide enough coverage. Large multi-element abdominal surface coils for parallel imaging are most useful in these patients as they may cover the whole thoracic and abdominal aorta. CMR can easily follow the pathway of the conduit using 3D contrast-enhanced MRA and/or large volume 3D SSFP imaging as this allows for reformattting in any desired plane, thus appreciating the often multi-tortuous course of these conduits. High-resolution black-blood images gated to systole for maximal wash-out effect can provide important vessel wall anatomy locally where a problem such as thrombus or neo-intima formation has been identified by 3D imaging. Local wall aneurysm and dissection may be assessed by dedicated black-blood imaging and cine SSFP slices. Cine 2D sequences may then provide further detail regarding narrowing (turbulence from cine 2D SSFP) or jet flow velocity (phase-contrast in-plane or through-plane velocity mapping). Also, the relation of the distal conduit-aortic anastomosis to the single branch abdominal arteries and the renal arteries is of interest.

Summary and perspectives

In complex congenital heart disease requiring baffles and/or conduits for flow redirection in their
respective repair strategy, CMR has proved itself to be a useful tool not only for pre-repair planning, but also for post-repair follow up. A simplified protocol to anatomically image conduits and baffles by CMR appeared a number of years ago [13]. This is of great patient benefit as the “ideal” conduit or baffle with minimal resistance, no thrombogenic risk, no degradation with auto-growth and auto-repair remains elusive. The use of CMR is supported by a large body of literature and is considered class I or II in these patients [62]. As such, CMR is the ideal investigation, providing additional benefits to echocardiography in terms of detailed graft anatomy and function. Moreover, serial quantification of ventricular function and mass will help timing re-do procedures, particularly for primary or secondary pulmonary valve replacement.

In the future, development of novel CMR sequences and new intravascular contrast agents are likely to further improve the assessment of anatomy and physiology of conduits and baffles. Multi-phase cine 3D CMR imaging and 4D flow are likely to allow easier and reliable hemodynamic assessment. Such CMR techniques may also form an excellent basis for building patient-specific biophysical cardiovascular models of baffle and/or conduit performance that couple mechanical and computational fluid dynamics. In the nearer future, to assist with planning of complex surgical repair, advanced post-processing tools may be further extended towards realization of virtual surgery, when myocardial tissue characteristics are added as properties in a multi-dimensional detailed cardiac model (see chapter dealing with the single ventricle for further details). CMR data from rapid prototyping may be used to make individually designed surgical or transcatheter apparatus. CMR is already an invaluable tool for planning and follow-up of percutaneous pulmonary valve implantation, a procedure which may benefit from CMR X-ray image fusion in the catheterization laboratory. Moreover, there still is an active vision of such a procedure to treat RV-PA conduit failure to be performed partly or solely under CMR-guidance as newer stents and devices will become CMR compatible. Such advances in CMR-related imaging sciences are expected while there are ongoing advances in the field of tissue engineering, with the potential to create improved and more durable
replacement materials that can grow to meet the physiological demands on blood flow of the developing child.

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52 Goo HW, Yang DH, Park IS, et al. Time-resolved threedimensional contrast-enhanced magnetic resonance angiography in patients who have undergone a Fontan operation or bidirectional cavo-pulmonary connection:


Cardiac tumors in the pediatric age group are rare and frequently benign. They can arise from and involve almost any part of the heart and its surrounding pericardial space. The true population-based incidence of cardiac tumors in children is not known. However, cardiac tumors are found in 0.07–0.2% of children seen at major teaching hospitals [1–3]. In recent years, the incidence of cardiac tumors appears to be rising. This is likely related to the availability of better diagnostic modalities [3,4].

Clinical presentation and diagnosis

Children with cardiac tumors may be asymptomatic at diagnosis or may present with symptoms of congestive heart failure or low cardiac output resulting from obstruction to cardiac inflow or outflow, arrhythmias, heart murmurs, abnormal findings on EKG or chest X-ray and sometimes with sudden cardiac death. A significant proportion of tumors are now diagnosed incidentally on imaging studies performed either for prenatal screening or for the evaluation of unrelated symptoms during childhood [5]. Echocardiography is limited in its ability to differentiate between various types of tumors and to accurately delineate the relationship of the tumor to surrounding structures. Echocardiographic imaging may be especially limited in adolescents and young adults due to poor acoustic windows. Because of these limitations, the use of magnetic resonance imaging (MRI) is increasing in the evaluation of children with cardiac tumors, especially when surgical excision is being planned. Referral for MRI is usually made in order to confirm the diagnosis, predict the histologic type of tumor, define its extent and relationship with surrounding cardiac and thoracic structures, to assess ventricular function/blood flow and to assess the vascularity of the tumor.

Types of tumor

The vast majority of cardiac tumors seen in the pediatric age group are benign with rhabdomyomas (45–65%), myxomas (10–15%), fibromas (8–25%) and intrapericardial teratomas (2–10%) being the most common [2–5]. Other less common tumors include hemangiomas, lipomas, hamartomas, fibroelastomas, papillary tumors, lymphangiomas, paragangliomas, Purkinje cell tumors and primary or metastatic malignant tumors of the heart.

MRI evaluation for cardiac tumors

Use of cardiac MRI in the evaluation of cardiac tumors was first described in the 1980s mostly in adult patients [6–9]. Since then, its usefulness in children has also been reported, although in only a small number of patients [10]. Diagnostic questions at the time of referral for MRI usually pertain
to characterization of the tumor type, its degree of vascularity, the effect of the tumor on ventricular function/blood flow (for example left ventricular outflow tract obstruction) and better delineation of the relationship and attachment of the tumor to surrounding structures such as the great vessels and the coronary arteries.

**MRI technique**

A suggested scanning protocol that we use for children with known or suspected cardiac tumors is shown in Table 18.1. Scanning is usually commenced with anatomic imaging using fast spin echo black-blood and cine steady-state free precession (SSFP) techniques. Static SSFP can also be used. These allow delineation of the size and extent of the tumor, along with its relationship to surrounding structures. T1 and T2 weighted spin echo imaging with and without saturation of signal from fat is performed to identify the signal characteristics of the tumor; this is used in determining what type of tumor it may be. Gradient echo cine imaging is performed if there is a question as to whether the mass is thrombus or not. A form of gradient echo cine imaging, myocardial tagging (for example spatial modulation of magnetization or SPAMM) can be used to determine if the mass is contracting (for example is it just localized hypertrophied muscle) and what the mass’s effects may be on regional myocardial contraction. STIR imaging can also be used to visualize edema and confirm that the mass does not contain fat (fat saturation is usually a part of the STIR sequence). Occasionally T2 prepared SSFP sequences using the navigator technique may be used to visualize the extent the tumor has encased the coronary arteries (see chapter on coronary arteries in this textbook for more details). First-pass perfusion imaging during injection of contrast is used to define the vascularity of the tumor. Contrast enhancement is assessed by repeating T1 weighted spin echo imaging with fat saturation. Finally, myocardial delayed enhancement imaging is performed approximately 10 minutes after contrast injection to examine kinetics of the contrast agent within tumor tissue.

**Table 18.1 Scanning protocol.**

| Optional: static steady state free precession (multiple planes) |
| Cine steady state free precession (multiple planes) |
| T1 weighted black-blood fast or spin echo with and without fat saturation |
| T2 weighted black-blood fast spin echo with fat saturation |
| Optional: Gradient echo cine imaging if thrombus is suspected |
| Optional: Myocardial tagging if mass is suspected to be muscle or to evaluate regional myocardial contraction. |
| Optional: STIR imaging |
| Optional: T2 prepared SSFP to visualize involvement by the coronaries |
| First-pass perfusion imaging during contrast injection (0.1–0.2 mmol/kg) |
| T1 weighted black-blood fast spin echo with fat saturation immediately post-contrast |
| Myocardial delayed enhancement imaging 10–20 minutes post-contrast |

**Tissue characterization**

Due to the rarity of cardiac tumors in the pediatric age group, limited data are available on the image characteristics of the different types of cardiac tumors and the accuracy of such characterization compared to pathologic diagnosis, especially for the rarer tumor types [10]. Based on available data, imaging characteristics of common cardiac tumors seen in the pediatric age group are shown in Table 18.2 and representative images are shown in Figures 18.1–18.7. In predicting tumor type, in addition to imaging characteristics, consideration should be given to the clinical history. As shown, most cardiac tumors in children are benign. Primary malignant tumors of the heart are extremely rare in the pediatric age group, the most type being sarcoma. However, malignancies involving the mediastinum such as non-Hodgkin lymphoma or leukemia may directly invade nearby great vessels or cardiac chambers and Wilms’ tumor of the kidney may invade the inferior vena cava. Rarely, metastatic involvement of the heart by distant tumors such as a melanoma may occur. Imaging features that might suggest a malignant tumor include a heterogeneous appearance, infiltration of adjacent tissues, edema in the region, presence of necrosis or calcification along with avid first-pass perfusion and contrast enhancement.
<table>
<thead>
<tr>
<th>Clinical correlates</th>
<th>Number</th>
<th>Site</th>
<th>Appearance</th>
<th>Mobile</th>
<th>Pericardial effusion</th>
<th>T1w</th>
<th>T2w</th>
<th>First-pass perfusion</th>
<th>Contrast enhancement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rhabdomyoma</td>
<td>Usually multiple, rarely single</td>
<td>Ventricles atria, intracavitary or intramural</td>
<td>Nodular masses</td>
<td>Sometimes</td>
<td>Rare</td>
<td>Isointense or slightly hyperintense</td>
<td>Mildly hyperintense</td>
<td>Poor</td>
<td>No</td>
</tr>
<tr>
<td>Tuberous Sclerosis</td>
<td>Single, rarely multiple</td>
<td>Left (more common) or right atrial. Attached to fossa ovalis, atrial free wall or mitral valve leaflets. May arise from other chambers</td>
<td>Pedunculated, lobular mass, commonly cause valvular obstruction</td>
<td>Yes</td>
<td>No</td>
<td>Isointense or hypointense, heterogeneous</td>
<td>Hyperintense</td>
<td>Poor</td>
<td>Yes</td>
</tr>
<tr>
<td>Myxoma</td>
<td>Single</td>
<td>Ventricular septum or left ventricular free wall, rarely atrial wall</td>
<td>Large intramural masses</td>
<td>No</td>
<td>No</td>
<td>Isointense or hypointense</td>
<td>Poor</td>
<td>Hypointense. On MDE, enhancing center with unenhanced outer shell</td>
<td></td>
</tr>
<tr>
<td>Myxoma</td>
<td>Single</td>
<td>Attached to the surface of the base of the heart (to the root of the aorta or pulmonary artery)</td>
<td>Large, bosselated, irregular mass with solid and cystic areas</td>
<td>No</td>
<td>Common</td>
<td>Heterogeneous, hyperintense areas</td>
<td>Heterogeneous Heterogeneous</td>
<td>Heterogeneous enhancement</td>
<td></td>
</tr>
<tr>
<td>Fibroma</td>
<td>Single</td>
<td>Ventricular septum or left ventricular free wall, rarely atrial wall</td>
<td>Large intramural masses</td>
<td>No</td>
<td>No</td>
<td>Isointense or hypointense</td>
<td>Poor</td>
<td>Hypointense. On MDE, enhancing center with unenhanced outer shell</td>
<td></td>
</tr>
<tr>
<td>Teratoma</td>
<td>Single</td>
<td>Attached to the surface of the base of the heart (to the root of the aorta or pulmonary artery)</td>
<td>Large, bosselated, irregular mass with solid and cystic areas</td>
<td>No</td>
<td>Common</td>
<td>Heterogeneous, hyperintense areas</td>
<td>Heterogeneous Heterogeneous</td>
<td>Heterogeneous enhancement</td>
<td></td>
</tr>
<tr>
<td>Clinical correlates</td>
<td>Number</td>
<td>Site</td>
<td>Appearance</td>
<td>Mobile</td>
<td>Pericardial effusion</td>
<td>T1w</td>
<td>T2w</td>
<td>First-pass perfusion</td>
<td>Contrast enhancement</td>
</tr>
<tr>
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</tr>
<tr>
<td>Hemangioma</td>
<td>—</td>
<td>Single</td>
<td>Atria or ventricle, Intramural or endocardial</td>
<td>Intramural masses are poorly circumscribed spongy masses, Endocardial masses are well circumscribed</td>
<td>Variable</td>
<td>Common</td>
<td>Slightly hyperintense</td>
<td>Hyperintense</td>
<td>Well perfused</td>
</tr>
<tr>
<td>Lipoma</td>
<td>—</td>
<td>Single</td>
<td>Commonly right atrium or atrial septum, may involve any chamber. May arise from endocardium, epicardium or myocardium</td>
<td>Circumscribed spherical or elliptical masses</td>
<td>No</td>
<td>No</td>
<td>Very hyperintense, intensity reduced on fat saturation</td>
<td>Iso or hypointense</td>
<td>Poor</td>
</tr>
<tr>
<td>Purkinje cell tumor</td>
<td>Ventricular arrhythmias</td>
<td>Single or multiple</td>
<td>Ventricles</td>
<td>?</td>
<td>No</td>
<td>No</td>
<td>Very Hyperintense, lower intensity on fat saturation</td>
<td>?</td>
<td>?</td>
</tr>
<tr>
<td>Fibroelastoma</td>
<td>—</td>
<td>Single</td>
<td>Cardiac valves (aortic or mitral) or endocardium</td>
<td>Pedunculated endocardial or valvular mass</td>
<td>Yes</td>
<td>No</td>
<td>Iso or hyperintense</td>
<td>?</td>
<td>?</td>
</tr>
<tr>
<td>Thrombus</td>
<td>—</td>
<td>Variable</td>
<td>Variable</td>
<td>Variable</td>
<td>Variable</td>
<td>No</td>
<td>Hyperintense early, iso or hypointense early, iso or hypointense late</td>
<td>Hyperintense on organized</td>
<td>Poor</td>
</tr>
<tr>
<td>Pericardial cyst</td>
<td>—</td>
<td>Single</td>
<td>Right cardiophrenic angle</td>
<td>Smooth walled, well circumscribed</td>
<td>No</td>
<td>No</td>
<td>Hypointense</td>
<td>Hyperintense</td>
<td>Poor</td>
</tr>
</tbody>
</table>
Figure 18.1 Large solitary intramural biopsy-confirmed cardiac fibroma involving the anterior left ventricular wall and the ventricular septum. The tumor was hypointense compared to normal myocardial on steady state free precession imaging (a), isointense on T1 weighted spin echo (b), hypointense on T2 weighted spin echo (c), showed poor first-pass perfusion (d) and was hyperenhanced on MDE imaging (e) with an unenhanced “shell” of normal myocardium.

Figure 18.2 While rhabdomyomas are frequently small and multiple, this large solitary biopsy-confirmed rhabdomyoma arose from the ventricular septum and “mushroomed” to the anterior inter-ventricular groove with an associated pericardial effusion. The tumor was slightly hyperintense compared to normal myocardial on steady state free precession (a), iso to slightly hyperintense on T1 weighted spin echo (b), hyperintense on T2 weighted spin echo (c), showed poor first-pass perfusion (d) and did not enhance on MDE imaging (e).
Figure 18.3 Biopsy-confirmed hemangioma arising from the right atrium near the atrioventricular groove (arrowheads). The mass was poorly circumscribed with extensive attachment to the atrial wall on steady state free precession imaging (a). It was isointense compared to the myocardium on T1 weighted spin echo (b), hyperintense on T2 weighted spin echo (c), showed avid first-pass perfusion (d and e) and was hyperenhanced on T1 weighted contrast imaging with fat saturation after injection of contrast (f).

Figure 18.4 Biopsy-confirmed pericardial teratoma in a newborn infant. The tumor was a large, bosselated mass with heterogeneous appearance and attachment to the anterior surface of the ascending aorta and superior vena cava along with a pericardial effusion on steady state free precession imaging (a). The tumor showed heterogeneous signal intensity on T1 weighted spin echo (b), was heterogeneously hyperintense on T2 weighted imaging (c), showed poor first-pass perfusion (d) and showed mild heterogeneous enhancement on MDE imaging (e).
Figure 18.5 Biopsy-confirmed right ventricular myxoma. The tumor was a pedunculated lobulated mass attached to the interventricular septum on steady state free precession imaging (a), was isointense compared to normal myocardium on T1 weighted spin echo (b), hyperintense on T2 weighted spin echo (c), showed poor first-pass perfusion (d) and was hyperenhanced on MDE imaging (e).

Figure 18.6 Metastatic melanoma invading the myocardium. The tumor was a poorly circumscribed mass involving the inferior wall of the right ventricle and the interventricular septum on steady state free precession imaging (a), showed heterogeneous signal intensity on T1 weighted spin echo (b), was hyperintense on T2 weighted spin echo (c) and showed heterogeneous enhancement after contrast injection on T1 weighted spin echo (d).
Figure 18.7 Fibroma affecting left coronary circulation. (a) Left coronary artery. The upper left and lower right images are two orthogonal projections using T2 prepared steady state free precession of the fibroma displacing the left coronary artery. (b) Left circumflex coronary artery. The upper left and lower right images are also two orthogonal projections using T2 prepared steady state free precession of the fibroma growing around the left circumflex coronary artery. (c) Three dimensional volume rendered image utilizing T2 prepared steady state free precession of the left coronary and circumflex coronary arteries as they are displaced. Note the gaping hole in the heart which represents the tumor which does not give off signal in the T2 prepared steady state free precession. (Courtesy of Dr. Mark Fogel, The Children’s Hospital of Philadelphia.)
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References
Considerations in the post-operative patient

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Introduction

The evaluation of post-operative patients is a major part of the practice of pediatric and congenital cardiovascular magnetic resonance imaging in developed countries. This importance is a necessary consequence of the demographics of congenital heart disease. In regions with advanced health care systems, the overwhelming majority of serious congenital cardiovascular disease is diagnosed in infancy or early childhood, and receives prompt treatment. These treatments, although often corrective or effectively palliative, are only rarely curative. However, the treatments are usually very successful in prolonging life and relieving symptoms. Indeed, the longevity of patients with many serious but relatively common defects such as tetralogy of Fallot or coarctation of the aorta is now approaching that of the general population [1].

It follows that a very high percentage of the patients with congenital heart disease in any given locale will be post-operative. In most cases there are potential problems related to the disease process or the operative interventions that will require periodic follow-up for the lifetimes of these patients. Thus, the majority of patients that most practitioners of pediatric cardiovascular magnetic resonance encounter will be post-operative patients. Magnetic resonance imaging is already a powerful and widely utilized tool for functional and anatomical characterization of the cardiovascular system in post-operative patients and its future appears even more promising. This chapter will outline some of the applications of magnetic resonance imaging to the evaluation of post-operative patients with congenital cardiovascular disease.

There are now a plethora of techniques that can be applied to the study of the cardiovascular system. The good clinician will want to achieve the optimum value for the patient in utilizing these multiple methods, which should be viewed as augmenting and being synergistic with each other rather than as sole solutions. The goal is to employ methods that yield the most information for the least cost in terms of patient risk, discomfort, and financial burden. We want to steer an intermediate course between the Scylla of inadequate diagnosis and the Charybdis of overutilization.

In comparison with other imaging methods, magnetic resonance imaging fares well. With the exception of some specialized applications, magnetic resonance imaging is non-invasive, and therefore does not incur the risk of catheter-related complications. Magnetic resonance imaging (MRI) does not subject patients to ionizing radiation. Cardiac catheterization and X-ray cineangiography, X-ray computed tomography, and nuclear medicine techniques are all associated with large radiation burdens [2,3]. Ionizing radiation is of particular concern in the population of patients with congenital cardiovascular disease. Many of
There have been multiple recent advances in magnetic resonance imaging software and hardware. The techniques made possible by these advances have substantially increased the information that can be effectively derived from magnetic resonance imaging in post-operative patients. A comprehensive treatment of these innovations is beyond the scope of this chapter, but we will list several of the more important ones with which practitioners examining post-operative patients should be familiar, although only a subset of these techniques will be employed in any given examination [6,7].

1. Cardiac synchronization:
   - Prospective triggering and retrospective gating.
   - Utilization of electrocardiogram and peripheral pulse for a cardiac synchronization signal.
   - "Self-gating", using magnetic resonance signal created by the cardiac motion itself for a cardiac synchronization signal.
   - "Real-time" magnetic resonance imaging techniques that effectively freeze cardiovascular motion.

2. respiratory synchronization/compensation methods:
   - Breath-holding.
   - Respiratory triggering.
   - Respiratory navigator technique for coronary imaging and isotropic three-dimensional steady state free precession imaging of the thorax.
   - Multiple average acquisitions.

3. Parallel acquisition of MRI signal (sensitivity encoding or SENSE, etc).

4. Spin echo imaging.

5. Cine or multiphasic cardiovascular imaging:
   - Standard segmented gradient echo cine imaging with and without increasing TE to enhance flow signal.
   - Steady-state free precession cine imaging.

6. Flow quantitation with phase contrast velocity encoding MRI.

7. Contrast-enhanced three-dimensional angiography.

8. Myocardial viability imaging (myocardial delayed gadolinium enhancement).


10. Analysis of ventricular function with MRI myocardial tagging.

11. Dobutamine and adenosine stress MRI.
Specific post-operative conditions

This section will discuss the post-operative evaluation of several common congenital cardiovascular malformations with emphasis on how magnetic resonance imaging can provide useful information in their follow-up. The propensity of patients for residual and recurrent post-operative cardiovascular problems and the nature of these problems can be predicted to a large extent based on the natural history of the malformation. Emphasis of the examination can then be placed on the most frequently affected cardiovascular subsystems.

Coarctation of the aorta

It is important to recognize that the term “coarctation of the aorta” includes several different pathological entities [8]. These abnormalities include: (1) an obstructive shelf localized to the posterior and internal region of the aorta at the aortic isthmus or juxtaductal region just distal the left subclavian artery; (2) tubular hypoplasia and elongation of the aortic arch; and (3) waist lesions that are introflexions of the aorta, encircling it at the site of narrowing. Any of these lesions may occur in isolation or in combination with the other two. Most patients with a shelf or waist malformation have some degree of hypoplasia of the transverse aortic arch, particularly the distal transverse aortic arch which lies between the left common carotid artery and the left subclavian artery. A wide spectrum of severity is seen in the lesions, ranging from minimal abnormalities that require no treatment to near complete interruption of the aorta.

A variety of surgical and interventional cardiology interventions have been employed to treat coarctation. Most of these have yielded good immediate results in the majority of cases. Obviously, very severe coarctation anomalies are more challenging to treat than mild types of coarctation. However, several problems are commonly found in patients who have undergone surgical or interventional catheter therapy for coarctation of the aorta. Recurrent or residual obstruction is a common post-operative difficulty. This obstruction may be either discrete or diffuse. Differentiation between discrete and diffuse obstruction is very important for guiding therapeutic intervention for residual or recurrent coarctation.

Upper and lower extremity blood pressures and Doppler echocardiography can often identify the presence of aortic obstruction. However, poor acoustic penetration in the mid-thorax frequently precludes confident delineation of the anatomy of the mid-portion of the descending aorta by echocardiographic imaging, particularly in older patients. Magnetic resonance imaging can provide exquisite anatomical images of the entire aorta, and clearly determine the location of the obstruction and whether obstruction is discrete or diffuse. Furthermore, magnetic resonance imaging can identify aortic collaterals that are usually poorly defined by echocardiography. The presence of substantial collateral formation can confound estimates of coarctation severity based purely on blood pressure and Doppler gradients. Phase contrast magnetic resonance imaging methods can quantify the significance of collateral formation and the significance of aortic obstruction [9] (Figure 19.1).

Aortic aneurysm formation can also occur in post-operative coarctation patients. Aneurysms have been shown to sometimes form subsequent to surgical patch repair of aortic coarctation (Figure 19.2). Likewise, aortic aneurysms sometimes form after balloon angioplasty of coarctation (Figure 19.3). If residual aortic obstruction is present, aneurysms can form where the flow jet from the obstruction strikes the aortic wall (Figure 19.4). In patients with residual tubular hypoplasia of the aortic arch, the ascending aorta may dilate as a consequence of the distal obstruction. Many patients with coarctation of the aorta also have bicuspid aortic valves which can readily be diagnosed with cine cardiac magnetic resonance. These patients are at increased risk for aneurysmal dilation of the ascending aorta. No clear guidelines are available regarding when to repair aortic aneurysms in post-operative coarctation patients. When the patient with a small aortic aneurysm is asymptomatic with no evidence of aortic dissection, our approach has been to obtain multiple evaluations at different times with magnetic resonance imaging and assess the rate of growth of the aneurysm.

Aortic valve, subvalve and/or mitral valve stenosis are sometimes seen in conjunction with coarctation of the aorta. Magnetic resonance imaging can clearly identify these lesions anatomically and
Figure 19.1 Status post balloon dilation of coarctation of aorta with mild residual coarctation. Parasagittal section obtained with phase contrast magnetic resonance imaging technique. (a) Magnitude image. (b) Phase image. Arrow: Flow jet through residual coarctation.

Figure 19.2 Status post surgical repair for treatment of coarctation of aorta. (a) Posterior view of contrast enhanced magnetic resonance angiogram. (b) Posterior view of rendered three-dimensional reconstruction of contrast enhanced magnetic resonance angiogram. L: Left; R: right; P: Posterior; H: Head; F: Foot. Arrow points to large aortic aneurysm.
can provide quantitative flow information with phase contrast techniques.

Persistent or recurrent left ventricular hypertrophy is frequently present in post-operative patients with coarctation of the aorta. Magnetic resonance imaging has been shown to be a more precise tool than echocardiography for accurately determining left ventricular mass. Therefore, when magnetic resonance imaging is performed for evaluation of post-operative aortic anatomy subsequent to coarctation repair, it is usually desirable to also obtain cardiac magnetic resonance images that will facilitate quantitative evaluation for left ventricular hypertrophy.

A substantial incidence of cerebral aneurysms has been reported in post-operative patients with coarctation of the aorta. Magnetic resonance imaging can clearly delineate such aneurysms [10]. In the future, screening of post-operative patients with coarctation for cerebral aneurysms with magnetic resonance imaging may become standard medical practice. The optimal time for performing such screening examinations remains to be defined.

**Interrupted aortic arch**

Interrupted aortic arch malformations can be successfully repaired surgically, with good early results in a high percentage of cases with appropriate early diagnosis. Current surgical practice is to perform corrective single stage direct anastomosis to repair the aortic arch in the neonatal period. Interrupted aortic arch is almost always associated with a ventricular septal defect, which is closed at the time of surgery. Recurrent subaortic obstruction of the left ventricular outflow tract in the late post-operative period is a frequent finding in these patients. Magnetic resonance imaging examination for definitive evaluation of the state of the aortic arch repair is useful especially prior to surgical re-intervention to repair the recurrent subaortic obstruction. Patients with interrupted aortic arch who were repaired with techniques other than direct anastomosis have a high incidence of late post-operative

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**Figure 19.3 Status post balloon dilation for treatment of coarctation of aorta.**

(a) Lateral view of contrast enhanced magnetic resonance angiogram. (b) Lateral view of rendered three-dimensional reconstruction of contrast enhanced magnetic resonance angiogram. A: Anterior; P: Posterior; L: Left; H: Head; F: Foot. Arrow points to small aortic aneurysm.
Figure 19.4 Status post surgical repair for treatment of coarctation of aorta with both recurrent proximal stenosis and moderate size aneurysm just distal to site of stenosis. (a) Parasagittal section showing magnitude image from phase contrast study. Arrow points to aortic aneurysm of moderate size. (b) Parasagittal section of colorized phase image from phase contrast magnetic resonance study. Arrow indicates flow jet through proximal stenosis. (c) Parasagittal section showing turbo spin-echo black blood double inversion image through descending aorta. Arrow points to aortic aneurysm of moderate size. (d) Lateral view of contrast enhanced magnetic resonance angiogram. Arrow points to aortic aneurysm of moderate size. (e) Lateral view of rendered three-dimensional reconstruction of contrast enhanced magnetic resonance angiogram. Arrow points to aortic aneurysm of moderate size. (f) Posterior view of rendered three-dimensional reconstruction of contrast enhanced magnetic resonance angiogram. Arrow points to aortic aneurysm of moderate size.
Tetralogy of Fallot or pulmonary valve atresia with ventricular septal defect status post systemic to pulmonary artery shunt

Young infants with severe tetralogy of Fallot or tetralogy of Fallot with pulmonary valve atresia are often treated with palliative aortopulmonary shunts in the neonatal period when therapy is required for severe cyanosis [11] (Figure 19.5). However, aortopulmonary shunts cause substantial ventricular volume loading. This ventricular volume loading has been demonstrated to have deleterious effects on ventricular function in many patients when sustained over a long period of time. Therefore, these shunts are typically taken down when the patient grows to a size and age at which either corrective surgery or a more physiologic cavopulmonary shunt, which in theory does not volume load the ventricle, is feasible. Thus, with rare exceptions, aortopulmonary shunts are not now intended to be permanent. Historically, they were intended to be permanent palliations, and occasionally adult patients will still present for follow-up examination having only been treated with an aortopulmonary shunt.

These patients require post-operative follow-up examinations because complications can happen subsequent to shunt placement. Sometimes the aortopulmonary shunt is too large, resulting in pulmonary overcirculation and pulmonary artery hypertension. Aortopulmonary shunts are now typically synthetic tubes which do not grow in diameter so that excessive pulmonary flow will be evident in the immediate post-operative period. However, shunts that are composed of entirely native artery, such as the classic Blalock–Taussig shunt, may enlarge over time. Shunts may be of adequate size shortly after surgery and then gradually become insufficient as a source of pulmonary blood flow because of patient growth and sometimes because of neointimal buildup in the shunt. Thrombi can form in shunts causing partial or complete obstruction to flow through the shunt. Aortopulmonary shunts can distort the pulmonary arteries, causing branch pulmonary artery stenosis with reduced pulmonary flow to one pulmonary artery and overperfusion of the contralateral pulmonary artery. The direct ascending aorta to right pulmonary artery shunt (Waterston) and the direct descending aorta to left pulmonary artery shunt (Potts), which are primarily of historical interest, were particularly prone to produce such distortions with patient growth (Figure 19.6). As has been noted, ventricular dilation and dysfunction can ensue secondary to long-term volume loading. Patients with such cardiac dilation and dysfunction are often prone to arrhythmias and atrioventricular valve regurgitation.

In a substantial number of cases, echocardiographic delineation of pulmonary artery anatomy and of the shunt is suboptimal; magnetic resonance imaging is an obvious choice and affords assessment of the shunt and volume load on the ventricle. Besides demonstrating the patency of the shunts, magnetic resonance imaging can also clearly depict the native pulmonary arteries, identify the presence of any substantial distortion to the arteries by the shunt, and evaluate for significant aortopulmonary collateral vessels, which may have been missed on pre-operative echocardiographic imaging.
Considerations in the post-operative patient

CHAPTER 19

with a small right ventricular to pulmonary artery conduit. This initial surgical palliation of hypoplastic left heart syndrome usually works reasonably well, but a number of post-operative problems can occur.

Right ventricular dysfunction is a common difficulty. Magnetic resonance imaging can provide high quality quantitative measures of right ventricular volume and ejection fraction as such, it is the preferred imaging method.

Norwood stage 1 (see Chapter 16)

Patients with hypoplastic left heart syndrome typically undergo a procedure in the neonatal period that establishes the proximal main pulmonary artery as the ascending aorta (neoaorta), repairs the aortic arch, and isolates the left and right branch pulmonary arteries from the proximal main pulmonary artery. A large atrial septal defect is surgically created. The branch pulmonary arteries are typically perfused with a modified right Blalock–Taussig shunt, although a modification of the procedure, introduced by Sano, perfuses them with a small right ventricular to pulmonary artery conduit. This initial surgical palliation of hypoplastic left heart syndrome usually works reasonably well, but a number of post-operative problems can occur.

Right ventricular dysfunction is a common difficulty. Magnetic resonance imaging can provide high quality quantitative measures of right ventricular volume and ejection fraction. Tricuspid valve and/or neoaortic valve regurgitation also may occur. Determination of tricuspid and neoaortic regurgitant fraction is straightforward with phase contrast magnetic resonance imaging. Obstruction of the neoaortic repair in the upper descending aorta can develop (Figure 19.7). Magnetic resonance imaging is an excellent tool for delineating such obstruction. The left and right branch pulmonary arteries can be distorted by the modified Blalock–Taussig shunt or Sano conduit, and such
Figure 19.7 Status post Norwood Stage I procedure for hypoplastic left heart syndrome with recurrent severe coarctation of aorta (arrows). Note severe dilation of neoaorta proximal to stenosis. (a) Sagittal view of contrast enhanced magnetic resonance angiogram. (b), (c) and (d), Angulated views of view of rendered three-dimensional reconstruction of contrast enhanced magnetic resonance angiogram.
distortion can readily be identified by magnetic resonance imaging [12]. Regrowth of the atrial septum can occur in the late post-operative period, so that the initial surgically created atrial septal defect becomes obstructed. Atrial septal defect restriction can also be clearly demonstrated by magnetic resonance imaging as can the pulmonary venous collateral vessels that may form with gradual restriction to atrial septal defect flow. A recent prospective randomized single center trial has indicated that magnetic resonance imaging can be an effective and less costly alternative to routine pre-operative cardiac catheterization prior to second stage bidirectional Glenn or hemi-Fontan procedures in many of these patients [13].

**Functional single ventricle after bidirectional Glenn shunt (see Chapter 16)**

Functional single ventricle patients commonly receive a bidirectional Glenn or hemi-Fontan shunt either as an intermediate palliative procedure if they require an aortopulmonary shunt for cyanosis in the neonatal period, or as an initial palliative procedure if they do not require an aortopulmonary shunt. Surgical results from these procedures are usually good, and they serve to partially unload the single ventricle, but post-operative problems can occur.

Occasionally branch pulmonary artery stenosis can develop. This problem can be clearly characterized with magnetic resonance imaging. Magnetic resonance imaging can also quantify the perfusion to the left and right lungs, data which can be important in judging the clinical significance of such branch pulmonary artery stenosis.

Intrapulmonary arteriovenous shunting is commonly present subsequent to the bidirectional Glenn or hemi-Fontan procedure and can cause substantial systemic arterial desaturation, which tends to increase in severity over time. This intrapulmonary shunting has been found to be primarily due to absence of pulmonary perfusion with hepatic venous blood. The liver apparently produces signaling factors that inhibit pulmonary angiogenesis. In the absence of these factors, intrapulmonary artery to vein fistulas form that bypass the alveoli, making gas exchange ineffective. Much of this shunting occurs through microscopic pulmonary arteriovenous communications that are too small to visualize with angiographic techniques, even X-ray contrast cineangiography or X-ray computed tomographic angiography. Three-dimensional contrast-enhanced magnetic resonance imaging can define pulmonary arteriovenous malformations approximately 1–3 mm in diameter, which is below the 5 mm minimum size commonly recommended for catheter coil occlusion. Time-resolved approaches with three-dimensional contrast-enhanced magnetic resonance imaging have recently been introduced and may prove to have substantial advantages [14].

In any event, the best intervention for pulmonary arteriovenous malformation development (see Chapter 16) subsequent to the bidirectional Glenn or hemi-Fontan procedure is usually Fontan completion [15]. As long as the hepatic venous blood flow is directed to the systemic venous side of the Fontan baffle and as long as this flow is equitably distributed to both lungs, regression of pulmonary venous malformations typically ensues without the need for further procedures. Magnetic resonance imaging can provide useful information for the Fontan by demonstrating the anatomical location of the hepatic veins so that they can be properly incorporated into the repair.

Pre-operatively unrecognized partial anomalous systemic to pulmonary venous communications, such as a vestigial levocardiac vein, can be detrimental in the post-operative bidirectional Glenn or hemi-Fontan patient. The increased venous pressure in the post-operative state can cause these communications to substantially enlarge. Enlargement of normally occurring small systemic venous collaterals between the upper and lower body may also occur under the influence of increased upper body systemic venous pressure. Any significant degree of pulmonary vascular obstruction will tend to make this tendency to systemic venous collateral enlargement and shunting worse. Magnetic resonance imaging can assist in identifying the significance of such collateral formation and in guiding therapeutic decisions.

Post-operative ventricular function and atrioventricular valve regurgitation are always of concern in these patients, and can be very effectively evaluated with magnetic resonance imaging. Patients
with atresia or stenosis of either the tricuspid or mitral valves require a widely patent atrial septal defect to ensure good systemic flow. Magnetic resonance imaging is effective in evaluating the adequacy of the atrial septal defect in these patients.

The combination of echocardiography and magnetic resonance imaging can reveal so much information in patients that have undergone the bidirectional Glenn or hemi-Fontan procedure that some investigators now question the need to perform routine cardiac catheterization prior to surgical Fontan completion in every patient, although others disagree [16,17]. The utility of catheterization in selected high-risk patients has not been disputed. Rather, as non-invasive techniques progress, there is a need to justify the significant population morbidity that attends the policy of catheterization of all such patients. Additional studies are clearly needed in this regard. A retrospective investigation suggests that pre-Fontan catheterization can be dispensed with in a substantial subset of patients [18].

**Functional single ventricle after Fontan procedure (see Chapter 16)**

Multiple post-operative problems can occur subsequent to the Fontan procedure and many are amenable to characterization with magnetic resonance imaging [19] (Figure 19.8). Obstruction to flow in the Fontan circuit is a common difficulty. Such obstruction is most frequent in the pulmonary arteries, but can occur in the inferior vena cava to pulmonary artery baffle. Obstruction can also occur in the pulmonary veins. Sometimes pulmonary venous obstruction occurs subsequent to repair of anomalous pulmonary venous connections. Such anomalous connections are often found in association with functional single ventricle. On other occasions, pulmonary venous obstruction may occur as a secondary consequence of the Fontan repair itself. One example is right pulmonary venous obstruction due to severe right atrial dilatation subsequent to a classical atrio-pulmonary Fontan repair. Another example is pulmonary venous obstruction by an extracardiac Fontan conduit.

Anatomic narrowing can be effectively delineated with both black-blood spin echo methods and bright-blood gradient echo techniques. Turbulent flow can be qualitatively identified by cine gradient echo employing relatively long TE. Estimates of pressure gradients across regions of obstruction can be made by phase contrast magnetic resonance imaging and evaluation of volumetric blood flow can also be made by phase contrast MRI in the inferior and superior vena cava, left and right branch pulmonary arteries, and ascending aorta [20,21]. These quantitative data can be of help in assessing the functional significance of an obstruction. Appropriately low velocity encoding parameters should be utilized by practitioners employing phase contrast techniques in the venous side of the Fontan circulation. Because the flow velocities are low, high velocity encoding settings will necessarily lead to inadequate dynamic data range and inaccurate measurements. The goal is to use velocity encoding levels that are just high enough to avoid exceeding the maximum velocity that can be encoded.

Subaortic or outlet foramen obstruction can occur following Fontan surgery in patients in whom the aorta arises from a small anterior outlet chamber. Such obstruction may not be present prior to surgery. However, when ventricular loading changes occur subsequent to surgery and the ventricular volume shrinks, the opening between the outlet chamber and the main body of the ventricle may become relatively restrictive. Any subaortic obstruction is deleterious in the Fontan circuit since such obstruction tends to raise the left ventricular end diastolic pressures and hence the central venous pressures. Evaluation of the subaortic region should be part of any comprehensive assessment of a Fontan patient with magnetic resonance imaging.

Aortic, neoaortic and/or systemic atrioventricular valve regurgitation can also occur in postoperative Fontan patients. Qualitative identification of valve regurgitation is readily made with cine gradient echo methods using a prolonged TE. Aortic and neoaortic valve regurgitation can be directly quantified by phase contrast magnetic resonance imaging. Systemic atrioventricular valve regurgitant volume is usually estimated by determining the stroke volume of the single ventricle from anatomical cine imaging studies and then subtracting the aortic stroke volume measured by phase contrast techniques.
Ventricular dysfunction is often the limiting factor in the longevity of Fontan patients. Magnetic resonance imaging is generally accepted as the “gold standard” for estimation of ventricular volumes, ejection fraction, and mass. Because magnetic resonance imaging can perform temporally resolved volumetric imaging, there is no need to utilize geometric models. These models frequently do not accurately approximate the non-standard ventricular shapes frequent in Fontan patients. Cine imaging of the ventricles can be quickly accomplished with cine balanced gradient echo sequences. The technique is reliable and lends itself readily to serial comparisons. Sophisticated analysis of ventricular function with magnetic resonance tagging methods may enable investiga-
tors to understand better the pathophysiological determinants of ventricular dysfunction in these patients [22,23].

Blood flow through the Fontan circuit is relatively slow. Patients with poor ventricular function are at high risk for thrombus formation. Thrombi of substantial size can be clearly visualized with magnetic resonance imaging techniques.

Pleural and pericardial effusions often occur subsequent to the Fontan procedure, and magnetic resonance imaging can easily and accurately delineate these. Partial diaphragmatic paralysis is sometimes seen in these patients and can also be readily demonstrated with real-time magnetic resonance imaging.

**Transposition of the great arteries after arterial switch procedure**

Most patients with complete transposition of the great arteries and without significant pulmonary valve or subpulmonic stenosis now undergo an arterial switch procedure. Experienced centers report excellent operative mortality for this corrective surgery, and studies suggest a better long-term prognosis for patients receiving the arterial switch than was previously the case for patients receiving atrial switch procedures. Despite these generally good results with the arterial switch procedure, significant post-operative problems can occur. In particular, long-term studies have demonstrated a significant incidence of sudden death in these patients, a finding which has prompted reevaluation of post-operative screening strategies.

During the repair, the pulmonary arteries are displaced anterior to the aorta by the Le Compte maneuver. Pulmonary artery branch stenosis can develop near the origin of the left and or right pulmonary arteries secondary to resultant tension on these arteries or because of differential growth patterns of the great arteries. Magnetic resonance imaging is an excellent tool for detecting such branch pulmonary artery stenosis.

Substantial dilation of the neoaortic root can occur following the arterial switch procedure. The patients at highest risk for this dilation are those who have had ventricular septal defects in addition to transposition. Magnetic resonance imaging is an excellent imaging tool for sequential follow-up of such dilation and for quantitation of any associated neoaortic valve regurgitation.

The arterial switch procedure necessitates translocation of the coronary arteries to the neoaorta (Figure 19.9). Acute coronary artery insufficiency can occur in the immediate post-operative period if the coronary anastomoses become obstructed. However, coronary artery insufficiency can also occur in the late post-operative period if differential growth patterns cause tension on the coronary anastomoses or kinking or stretching of the coronary arteries. Late sudden death following the arterial switch procedure for transposition of the great arteries is thought to be primarily secondary to coronary artery malfunction [24].

Magnetic resonance imaging can identify coronary artery obstruction and the effects of coronary artery obstruction, such as ventricular dysfunction and infarction. Taylor and colleagues demonstrated in a preliminary study the feasibility of coronary artery and viability imaging with magnetic resonance in this patient population [25]. Further investigations are needed to characterize the sensitivity and specificity of magnetic resonance magnetic resonance coronary angiography for identification of post-operative arterial switch patients at risk for sudden death and in need of surgical or catheter re-intervention.

**Transposition of the great arteries after atrial switch procedure**

Atrial switch procedures for transposition of the great arteries have been largely superseded by the arterial switch operation. Exceptions include rare cases of isolated ventricular inversion, for which
atrial switch is the corrective procedure of choice, and patients with congenitally corrected transposition of the great arteries in whom both atrial and arterial switches are sometimes performed. However, many older patients with Senning or Mustard repairs for typical complete transposition of the great arteries survive and require continued follow-up [26] (Figure 19.10).

Multiple late onset complications of the Senning and Mustard repairs have been described. Most of these problems can be delineated by magnetic resonance imaging. It should be noted that many older patients with atrial switch repairs have arrhythmias, particularly sinus node disorders, presumably secondary to their extensive atrial surgery. A sizable proportion of these patients will have cardiac pacemakers or defibrillators, which have historically been a relative contraindication to magnetic resonance imaging.

Dysfunction of the systemic right ventricle occurs in a substantial number of older patients with atrial switch repairs for complete transposition of the great arteries. Magnetic resonance imaging is an excellent tool for providing quantitative data on right ventricular volume, mass, and ejection fraction in these patients.

Hemodynamically significant tricuspid valve regurgitation may also develop, frequently secondary to the tricuspid annular dilation caused by right ventricular dysfunction. Magnetic resonance imaging can provide quantitative determination of regurgitant fraction and right ventricular volumes; MRI has internal checks (for example net flow in the aorta must equal net flow in the pulmonary arteries in the absence of intracardiac shunting) and is therefore the preferred method.

Obstruction of the pulmonary or systemic venous return at the intracardiac baffle is another common complication. Both pulmonary and systemic venous pathways can be clearly delineated anatomically by magnetic resonance imaging (Figure 19.11). Phase contrast imaging can define blood flow velocities in these pathways and characterize obstruction to flow.

**Atrioventricular septal defects**

Surgical repair of atrioventricular septal defects is now routine at major centers for congenital heart surgery, and overall results in experienced hands are excellent. However, the grossly malformed atrioventricular valve tissue present in some patients may preclude the creation of competent and non-stenotic atrioventricular valves by the surgeon. Magnetic resonance imaging can accurately quantify atrioventricular valve regurgitation. Magnetic resonance imaging can also quantify residual left to right shunts that may be present post-operatively. Patients with atrioventricular canal defects may have associated left or right ventricular hypoplasia, left or right ventricular outflow tract obstruction, pulmonary artery branch stenosis, or coarctation. Magnetic resonance imaging can clearly delineate these associated anomalies when present and define how effectively they were addressed by the surgery.

**Secundum atrial septal defects**

Secundum atrial septal defects can be effectively corrected with either surgical closure or catheter device occlusion. Most patients are free from significant post-operative problems, although late onset arrhythmias, persistent pulmonary hypertension, and rare device-related complications can occur, although pericardial and pleural effusions can occasionally cause difficulty in the immediate post-operative period. Magnetic resonance imaging can clearly delineate the substantial reduction in right atrial and right ventricular volumes sizes that accompany successful elimination of substantial atrial left to right shunts. Furthermore, magnetic resonance imaging can be very helpful in atypical post-operative cases with persistent cardiovascular symptoms. Such symptoms can occasionally be secondary to incomplete pre-operative diagnosis, such as the failure to detect anomalous pulmonary venous connections. Rarely there can be misidentification of structures at surgery with consequent improper repair. For example, inadvertent closure or even misdirection into the left atrium of the connection between the inferior vena cava and right atrium has been described. Implanted atrial septal occluder devices of commonly employed types are not generally considered contraindications to magnetic resonance imaging with standard magnetic resonance systems and techniques. However, responsible physicians should be aware of the device type and any specific recommendations that may pertain to it regarding
Figure 19.10 Transposition of the great arteries after Senning procedure. Rendered three-dimensional reconstruction of contrast enhanced magnetic resonance angiogram of the right ventricle and pulmonary arteries. (a) Frontal view. (b) Posterior view. (c) Contiguous diastolic short axis sections through the left and right ventricles obtained with steady state free precession technique. Note the dilated right ventricle.
Considerations in the post-operative patient

Most patients undergoing surgical closure of ventricular septal defects have excellent overall results. However, late post-operative problems can occur. Right bundle branch block with resultant asynchrony of ventricular contraction and flattened or paradoxical motion of the interventricular septum occur frequently. However, these anomalies typically have minimal effect on overall ventricular function.

Ventricular dysfunction, dilation, and residual hypertrophy in the late post-operative period subsequent to ventricular septal defect closure can be seen. Repair after the first year of life is thought to be a risk factor for such ventricular dysfunction and dilation. Magnetic resonance imaging can readily identify and quantify these problems.

Sinus venosus atrial septal defects

Sinus venosus atrial septal defects require surgical closure. Operative results are typically excellent, but post-operative problems infrequently occur. One potential problem is sinus node dysfunction which can occur subsequent to surgery near the sinus node [27]. Anomalous pulmonary venous connections are nearly always present with sinus venosus atrial septal defects. Corrective surgery baffles the anomalous pulmonary veins into the left atrium. Occasionally the baffling procedure can inadvertently result in obstruction of vena caval flow into the right atrium. Alternatively, the baffle can be inadequately sized and the rerouted pulmonary veins can become obstructed. Magnetic resonance imaging can provide clear delineation of both systemic and pulmonary venous pathways post-operatively and is an excellent tool to identify or rule out suspected systemic or pulmonary venous obstruction.

Ventricular septal defects

Most patients undergoing surgical closure of ventricular septal defects have excellent overall results. However, late post-operative problems can occur. Right bundle branch block with resultant asynchrony of ventricular contraction and flattened or paradoxical motion of the interventricular septum occur frequently. However, these anomalies typically have minimal effect on overall ventricular function.

Ventricular dysfunction, dilation, and residual hypertrophy in the late post-operative period subsequent to ventricular septal defect closure can be seen. Repair after the first year of life is thought to be a risk factor for such ventricular dysfunction and dilation. Magnetic resonance imaging can readily identify and quantify these problems.

Figure 19.11 Transposition of the great arteries after Senning procedure. Contiguous coronal sections from rapidly acquired magnetic resonance imaging volume obtained subsequent to left arm intravenous injection of gadolinium. S: superior limb of systemic venous baffle. I: inferior limb of systemic venous baffle.
Aortic regurgitation can be seen in the late post-operative period in these patients. Although aortic regurgitation is readily detected by flow sensitive magnetic resonance cine sequences, magnetic resonance imaging is most helpful when accurate quantitative estimates of aortic regurgitant fraction and left ventricular volume are required using phase-contrast techniques.

Residual ventricular shunts can also be present subsequent to surgery. Most of these shunts are small and hemodynamically insignificant. Rarely, however, large residual shunts may be present and necessitate reoperation. Flow sensitive cine magnetic resonance sequences can readily detect these shunts. However, phase contrast magnetic resonance imaging of the aorta and pulmonary artery can also quantify the shunt volume and the ratio of pulmonary to systemic flow. These data can be very helpful for making decisions regarding possible reoperation.

Occasionally pulmonary hypertension can persist subsequent to surgery for ventricular septal defect and even progress in severity in the post-operative period. Magnetic resonance imaging can play an important role in assessment of patients with suspected post-operative pulmonary hypertension by quantifying right ventricular function and persistent hypertrophy and by delineating unrecognized distal pulmonary artery branch stenosis (see chapter on pulmonary circulation).

**Total anomalous pulmonary venous connection**
Surgical results for patients with total anomalous pulmonary venous connection are usually excellent [28]. However, a subset of patients will develop pulmonary venous obstruction at either the anastomotic site between the pulmonary venous confluence and the main body of the left atrium or in the individual pulmonary veins. Magnetic resonance imaging can contribute to the evaluation by documenting the location of pulmonary venous obstruction (Figure 19.12), quantifying the resultant effect on right ventricular function, and by determining differential pulmonary artery and pulmonary venous blood flows.

**Partial anomalous pulmonary venous connection**
Partial anomalous pulmonary venous connection is usually effectively corrected by surgery. However, late post-operative problems can and do occur. The rerouted pulmonary veins can become obstructed. When the repair necessitates placement of a baffle in the superior or the inferior vena cava, these systemic veins can become

![Figure 19.12](image-url)
obstructed. The anatomy and hemodynamic effects of both types of obstruction can be well characterized by magnetic resonance imaging using cine MRI, gadolinium techniques and phase contrast measures.

**Anomalous coronary arteries**

Coronary arteries may originate anomalously from the pulmonary artery. When the left main coronary artery originates from the pulmonary artery, left ventricular ischemia and resultant heart failure generally ensue in early infancy as soon as the pulmonary vascular resistance drops. There is a high rate of recovery of these patients following reimplantation of the left coronary artery into the aorta. Unfortunately, those that recover will often have residual myocardial infarctions. Substantial mitral valve regurgitation can also be present in survivors of surgery, often secondary to pre-operative left ventricular papillary muscle ischemia. Magnetic resonance imaging can define the extent of infarction in post-operative patients, and in quantifying their ventricular function and mitral regurgitation.

Patients with origin of the right coronary artery from the main pulmonary artery are typically asymptomatic in early life, although coronary artery dilation can be striking. Surgical repair is usually recommended because ischemia and sudden death can occur in older patients. Surgical results of coronary reimplantation are good. Post-operative magnetic resonance imaging can quantify ventricular size and function and document successful reimplantation using navigator coronary sequences and absence of infarction utilizing delayed enhancement.

Anomalous origin of the left coronary artery from the right sinus of Valsalva, and anomalous origin of the right coronary artery from the left sinus of Valsalva are frequent autopsy findings in young athletes dying suddenly. When these defects are identified clinically, surgery is often recommended. Magnetic resonance imaging is an excellent tool for quantifying post-operative ventricular function in these patients using cine MRI, for delineating both pre and post-operative coronary anatomy utilizing Navigator directed, T2 prepared steady state free precession MRI, and ruling out infarction using myocardial viability imaging.

**Tetralogy of Fallot after corrective surgery**

Tetralogy of Fallot is a common congenital cardiovascular defect, which usually receives corrective surgery during infancy, and is often seen in follow-up by practitioners of cardiovascular magnetic resonance imaging. Tetralogy of Fallot is repaired by closure of the ventricular septal defect, resection of infundibular muscle, and usually by some type of pulmonary valve resection, annular patch enlargement, or valvuloplasty. Sometimes the right ventricular outflow is so obstructed that a patch across the entire outflow is necessary, completely disrupting pulmonary valve diastolic function. Since most patients with tetralogy of Fallot have significant pulmonary valve stenosis, the surgical procedures on the pulmonary valve are usually unavoidable if post-operative right ventricular hypertension is to be prevented. However, these procedures usually result in substantial pulmonary valve regurgitation.

Although most patients with tetralogy of Fallot have good initial results, a number of problems can develop, particularly in the late post-operative period. Recurrent or residual right ventricular outflow, pulmonary valve, or main pulmonary artery obstruction may occur. Pulmonary artery branch stenosis is an even more frequent difficulty. Such residual obstruction will cause right ventricular hypertrophy, which, depending on its severity, will adversely affect long-term prognosis. Precise localization of stenoses, particularly distal stenoses, and precise anatomical delineation of whether or not stenoses are discrete or long segment is often not possible with ultrasound because of poor acoustic penetration of regions of interest. Magnetic resonance imaging can usually provide clear depiction of these problems using cine and phase contrast techniques along with 3D gadolinium imaging so that clinical decision making can be facilitated (Figures 19.13 and 19.14).

Right ventricular dilation and right ventricular or biventricular dysfunction also occur in a significant number of post-operative tetralogy of Fallot patients. Quantitative evaluation is most accurately performed with magnetic resonance imaging (Figure 19.15). Steady state free precession sequences are highly advantageous for volumetric analysis with magnetic resonance imaging because
Figure 19.13 Tetralogy of Fallot after repair. (a) and (b) Residual pulmonary valve stenosis. (a) Magnitude image from phase contrast study. (b) Color flow map superimposed on magnitude image from phase contrast study, with scale on left side of image. Pulmonary valve regurgitation. (c) Magnitude image. (d) Color flow map superimposed on magnitude image, with scale on left side of image. PS: pulmonary stenosis jet; PR: pulmonary regurgitation jet.
there is an inherent high contrast between the blood pool and the ventricular walls. Image quality is typically best with breath-holding techniques. However, respiratory gating or multiple averaging can be effectively employed when breath-holding is not feasible.

Pulmonary valve regurgitation is another common problem seen in post-operative tetralogy of Fallot patients. In the short term, pulmonary valve regurgitation is generally well tolerated. However, deleterious effects from long-standing pulmonary valve regurgitation can and do occur [29]. Volumetric quantification of such regurgitation is best made with phase contrast magnetic resonance angiography (Figure 19.16). If no residual shunts are present, and if regurgitation from other cardiac valves is insignificant, pulmonary regurgitant fraction can also be estimated by subtracting the left ventricular stroke volume from the right ventricular stroke volume and dividing by the right ventricular stroke volume.

Multiple magnetic resonance imaging techniques are often utilized for investigation of obstruction of the right ventricular outflow tract and pulmonary arteries in patients with tetralogy of Fallot. The standard cardiac synchronized multislice spin echo method has been successfully used in this application for many years. Modification of the spin echo method to obtain multiple k-space lines on each acquired image with each heartbeat

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**Figure 19.14** (a) and (b) Rendered three-dimensional reconstruction of contrast enhanced magnetic resonance angiogram of the right ventricle and pulmonary arteries in a patient after repair of tetralogy of Fallot. Note that both distal right and left pulmonary arteries are severely narrowed. RPA: Right pulmonary artery; LPA: Left pulmonary artery.

**Figure 19.15** Tetralogy of Fallot after repair. Contiguous diastolic short axis sections through the left and right ventricles obtained with steady state free precession technique. Note the severely dilated right ventricle.
Figure 19.16 Tetralogy of Fallot after repair. (a) Sagittal section showing magnitude image from phase contrast study. (b) Sagittal section of colorized phase image from phase contrast magnetic resonance study. Arrow indicates diastolic flow jet of pulmonary regurgitation. (c) Axial section showing magnitude image from phase contrast study. (d) Axial section of colorized phase image from phase contrast magnetic resonance study. Arrow indicates diastolic flow jet of pulmonary regurgitation. (e) Flow profile of the pulmonary regurgitation jet. Note the irregularity of the profile. (f) Time-dependent main pulmonary artery flow volume calculated from phase contrast magnetic resonance imaging in this patient.
has greatly expedited spin echo imaging. Inversion methods for nulling the signal from the blood pool are now also frequently employed to good effect. Spin echo images are usually static. However spin echo images typically have high quality and relatively low sensitivity to susceptibility artifacts from ferromagnetic materials such as sternal wires commonly found in post-operative patients. Thus, a set of spin echo images covering the right ventricular outflow tract and pulmonary arteries is typically obtained in these patients.

Multiphasic gradient echo images are also widely employed to evaluate the right ventricular outflow tract and pulmonary arteries. The multiphasic images are helpful because the dynamic nature of right ventricular outflow tract contraction and pulmonary artery movement with the cardiac cycle can be visualized. There are several methods for acquiring multiphasic gradient echo images. We generally employ a standard segmented gradient echo technique that allows us to obtain multiple lines of k-space per phasic image per cardiac cycle. We have found that the off-resonance artifacts common with the new steady state free precession sequences are accentuated by flow abnormalities in the right ventricular outflow tract and pulmonary arteries. Thus, balanced gradient echo sequences are less satisfactory than standard segmented gradient echo sequences for this application in our experience. Often we use a relative long effective TE with our multiphasic gradient echo sequences to make them flow sensitive and to thus facilitate qualitative assessment for regions of obstruction to blood flow.

We also routinely employ three-dimensional contrast enhanced magnetic resonance angiography for these cases. The three-dimensional coverage, high spatial resolution and excellent contrast-to-noise of contrast enhanced magnetic resonance angiograms is a great help for anatomic characterization of small tortuous blood vessels in the pulmonary vasculature. Three-dimensional volume rendering software can create realistic renditions of gadolinium enhanced three-dimensional magnetic resonance angiograms that can greatly facilitate presentation of anatomical details to clinicians with limited experience in viewing tomographic images.

With parallel acquisition techniques such as SENSE and its variants, substantial temporal resolution of the magnetic resonance angiograms can be achieved. When sequential volume acquisitions are employed with volume acquisition times of 8 seconds or less, it is usually possible to separate the pulmonary and systemic arterial enhancement phases of the magnetic resonance angiogram. The pulmonary arteries can then be readily differentiated from adjacent pulmonary veins and are unobscured by the overlying aorta. Further reduction of volume acquisition time to the subsecond range is achievable if signal to noise or spatial resolution is reduced. Such extremely rapid scans have great utility for qualitative assessment of pulmonary perfusion in these patients.

A significant number of patients with tetralogy of Fallot will require reintervention at some point after their corrective surgery. Magnetic resonance imaging is often utilized to guide clinical decision-making in this regard. The most common reinterventions are catheter or surgical arterioplasty and/or stenting for persistent or recurrent pulmonary artery branch stenosis, surgical revision of recurrent or persistent right ventricular outflow tract obstruction, and valved conduit placement for relief of pulmonary valve regurgitation. Criteria for recommending the first two interventions are relatively straightforward. Right ventricular outflow obstruction that is of a moderate or severe degree has a poor long-term prognosis without intervention and is usually successfully relieved by repeat surgery when the obstruction is discrete. Most clinicians will recommend repeat surgery upon the identification of such discrete right ventricular outflow tract obstruction. Similarly, discrete proximal stenosis of one or both pulmonary artery branches can cause long-term problems for these patients and can usually be effectively relieved by surgery or by interventional catheter stenting. Thus, therapy is usually advised when such stenosis is identified.

The proper timing of valved conduit replacement for alleviation of pulmonary valve regurgitation is a much more difficult clinical decision. This difficulty arises primarily because of the limitations of available valved conduits. These conduits do not grow and can become relatively stenotic on that basis alone when they are placed in young patients who subsequently increase in body size and cardiac output. If a conduit is implanted at a young age,
patient growth alone will ensure multiple reoperations for conduit replacement. Furthermore, tissue reaction to such conduits may itself cause stenosis in the mid-portion the conduit, at its valve, or at its insertion sites. When such stenosis is substantial, conduit replacement is required. Finally, conduit valves may degenerate over time and become regurgitant, negating the positive hemodynamic effect of the conduit implantation. On the other hand, there is evidence suggesting that prolonged severe pulmonary regurgitation can eventually result in irreversible right ventricular damage that cannot be corrected by pulmonary valve replacement. Mortality from sudden death and heart failure increase during the third decade following repair, suggesting that chronic pulmonary regurgitation is not benign. It follows that the goal of most clinicians is to delay conduit replacement for as long as possible to allow for patient growth, but to implant conduits, when necessary, prior to the development of substantial deterioration of the right ventricle.

Several published studies now provide rough guidelines for the timing of pulmonary valve replacement to alleviate chronic pulmonary valve regurgitation. Therrien and colleagues performed a retrospective evaluation of 17 consecutive adult patients with tetralogy of Fallot who underwent pulmonary valve replacement [30]. Magnetic resonance imaging studies before and after the procedures revealed a decrease in right ventricular end diastolic and end systolic volume for all patients. However, no patient with a right ventricular end diastolic volume of greater than 170 ml/m$^2$ or a right ventricular end systolic volume of greater than 85 ml/m$^2$ achieved normal right ventricular end diastolic (less than 108 ml/m$^2$) or end systolic volumes (less than 47 ml/m$^2$) on the post-operative study. This study in a limited number of older patients suggests that right ventricular dilatation can become irreversible and that pulmonary valve implantation can reverse this dilatation if it is performed before the right ventricular dilatation becomes too severe.

Geva and colleagues correlated magnetic resonance imaging findings with clinical status in 100 consecutive tetralogy of Fallot patients who had undergone corrective surgery at a median follow-up time of 21 years [31]. They found that left ventricular and right ventricular dysfunction, as evidenced by low left ventricular and/or right ventricular ejection fractions, were the best imaging predictors of poor clinical status. Interestingly, pulmonary regurgitant fraction and right ventricular diastolic dimension were not independently associated with clinical status.

Substantive right ventricular myocardial fibrosis has been noted in pathological specimens of post-operative patients with tetralogy of Fallot who have died suddenly. Late gadolinium enhancement is a sensitive method for detecting myocardial fibrosis in vivo. Multiple investigative groups have employed this technique to identify right ventricular myocardial fibrosis in tetralogy of Fallot. Babu-Narayan and colleagues quantitatively correlated the extent of right ventricular myocardial fibrosis measured with late gadolinium enhancement magnetic resonance imaging to other markers, including right ventricular function, exercise tolerance, neurohumoral activation, and arrhythmia incidence and found a strongly positive association [32]. Longitudinal studies are needed to further define the role of late gadolinium enhancement as a prognostic tool in these patients.

**Systemic venous obstruction and systemic arterial obstructive syndromes subsequent to therapy for congenital cardiovascular disease**

Systemic venous obstruction is sometimes seen in patients who have undergone therapy for congenital cardiovascular disease. Flow through the superior vena cava, the inferior vena cava or both may be blocked. Obstruction to superior vena caval flow may cause in elevation of cerebral venous pressure and result in hydrocephalus. Systemic venous obstruction can preclude implementation of the Glenn procedure and/or Fontan completion and thus can have major prognostic implications. Systemic venous obstruction can often be suspected by physical examination. With two-dimensional and Doppler echocardiography, visualization of the systemic veins is often incomplete, and false negative results can occur. Magnetic resonance imaging can clearly delineate systemic venous obstruction or effectively rule it out by using cine, gadolinium and dark-blood techniques, and is helpful in determining suitability of such obstruction for catheter intervention. Systemic venous obstruction that is discrete can often be
effectively relieved by catheter techniques, whereas long segment obstruction is much less amenable to such treatment. It is important to aggressively pursue definitive diagnosis of systemic venous obstruction as soon as it is suspected, since treatment is much more effective when the obstruction is of recent onset than when obstruction is long standing.

Systemic arterial obstruction can also be seen subsequent to catheter and operative therapy for congenital cardiovascular disease. The relatively small femoral arteries of infants and young children are particularly prone to such obstruction after instrumentation. Magnetic resonance imaging with cine gradient echo and with contrast-enhanced gadolinium angiographic techniques can clearly delineate the presence of systemic arterial obstruction or effectively exclude it. Systemic arterial obstruction should be investigated aggressively as soon as it is clinically suspected, since, as with systemic venous obstruction, therapeutic results are much better with recently developed obstruction than with obstruction of long duration.

**Truncus arteriosus**
Corrective surgery for truncus arteriosus is usually performed in early infancy. The procedure involves ventricular septal defect closure with routing of the left ventricular outflow into the truncus arteriosus, disconnection of the left and right pulmonary arteries from the truncus arteriosus, and placement of a right ventricular to pulmonary artery conduit. The right ventricular to pulmonary artery conduit placed at the initial surgery will typically become stenotic in early childhood, necessitating repeat surgery for conduit replacement. Magnetic resonance imaging can accurately determine the site or sites of such conduit stenosis and characterize the anatomy of the distal pulmonary arteries. Regurgitation through the right ventricular to pulmonary artery conduit is frequently present and magnetic resonance imaging can provide accurate quantitative estimates of regurgitation severity using phase contrast techniques and cine MRI of the left and right ventricles. Furthermore, magnetic resonance imaging can quantitate any resultant effects of conduit stenosis and/or regurgitation on right ventricular volume and ejection fraction [33].

Many truncal valves are abnormal, and truncal valve regurgitation is a frequent finding. Phase contrast magnetic resonance imaging can provide quantitative estimates of truncal valve regurgitation. The post-operative neo ascending aorta often is dilated after truncus arteriosus repair. Magnetic resonance imaging using cine, gadolinium and dark-blood techniques is an excellent technique for identifying such dilation and tracking its growth.

**Aortic valve and subvalve stenosis**
Magnetic resonance imaging can identify and quantify the severity of valvar and subvalvar aortic stenosis. Most congenital aortic valve stenosis is amenable to catheter balloon angioplasty. This treatment, while often effective in relieving the gradient across the aortic valve, usually results in some degree of aortic valve regurgitation. The aortic regurgitation typically increases over time, and eventually may necessitate aortic valve replacement. Magnetic resonance imaging is an excellent tool for quantifying regurgitation severity using phase contrast mapping, left ventricular diastolic and systolic cavity sizes using cine MRI, and left ventricular ejection fraction. MRI can thus be of substantial assistance in guiding decisions regarding operative intervention (Figure 19.17).

**Post-operative neurological function**
Neurological function in post-operative patients with congenital cardiovascular disease has become a matter of increasing concern. Mortality from surgery and interventional catheter treatment of these patients has steadily dropped. Institutions with advanced levels of expertise report high survival rates, even with severe lesions. However, significant morbidity continues to be associated with these treatments. Neurological morbidity is a particularly important problem. Even small neurological impairments can reduce the ability of these young people to independently function in society. Major neurological impairments are, of course, tragedies for everyone concerned. Most patients with congenital cardiovascular disease are treated at an early age, long before their cognitive function can be definitively evaluated by psychometric testing. Neurological outcomes assessment that depends on such testing requires many years for completion. There is a great need for suitable surrogate markers that are identifiable in infancy and early childhood to be correlated with outcome. If such markers can be identified,
they then can be utilized to define the neurological effects of cardiovascular care modifications expeditiously and so facilitate time efficient technique optimization.

Detailed neurological anatomical and functional information is readily obtainable with magnetic resonance imaging methods. Several early studies of brain function and structure in patients with post-operative congenital cardiovascular disease have suggested that magnetic resonance imaging has great promise for providing surrogate markers of neurological outcome.

Miller and colleagues performed magnetic resonance imaging on 23 children after open heart surgery for congenital cardiovascular disease [34]. They obtained abnormal studies that showed

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Figure 19.17 Subaortic stenosis treated by homograft aortic root replacement. Stenosis through homograft aortic root replacement. (a) Magnitude image from phase contrast study. (b) Color flow map superimposed on magnitude image from phase contrast study with scale on left side of image. Regurgitation through homograft aortic root replacement is present. (c) Magnitude image from phase contrast study. (d) Color flow map from phase contrast study superimposed on magnitude image, with scale on left side of image. R: Right; L: Left; H: Head; F: Foot.
evidence of hypoxic-ischemic encephalopathy in 17 (74%). Mahle and colleagues prospectively evaluated 24 term neonates with congenital cardiovascular disease with pre-operative magnetic resonance imaging and subsequent post-operative studies in the early post-operative period and at several months after surgery [35]. They identified mild ischemic lesions, primarily periventricular leukomalacia, in several pre-operative subjects and in the majority of post-operative patients. In 19 of their patients, pre-operative magnetic resonance spectroscopy was performed. Elevated brain lactate was detected in 10 cases, and the presence of elevated lactate correlated significantly with the presence of pre-operative lesions on magnetic resonance imaging. Galli and colleagues performed magnetic resonance imaging studies on 105 neonates and infants less than or equal to 6 months of age with congenital heart disease who had undergone cardiopulmonary bypass. They found a 54% (44 of 82) incidence of periventricular leukomalacia in the neonates compared with only a 4% (1 of 23) incidence of periventricular leukomalacia in the older infants. They observed that periventricular leukomalacia is common after neonatal cardiac surgery. They noted that periventricular leukomalacia seen on magnetic resonance imaging is associated with developmental delay and hyperactivity in infants without congenital heart disease. However, they cautioned that further study was required to determine the long-term significance of the early post-operative magnetic resonance imaging findings in this patient population.

McQuillen and colleagues employed magnetic resonance imaging to study 29 consecutive term infants with transposition of the great arteries prior to corrective surgery [36]. They identified 12 patients (41%) with pre-operative brain injury. All of these patients had undergone balloon atrial septostomy. In an earlier study, this group had utilized magnetic resonance spectroscopy to demonstrate the presence of abnormal brain metabolism in pre-operative patients with transposition [37]. An extended version of this brain metabolism investigation was recently reported [38]. These studies have demonstrated that magnetic resonance imaging can detect significant brain abnormalities in patients with congenital cardiovascular disease. They make clear the important point that pre-operative neurological abnormalities can be present in these patients. It follows that comparative pre-operative assessments are necessary before adverse neurological outcomes can be fairly ascribed to an intervention.

In a more recent study McQuillen and colleagues evaluated pre-operative magnetic resonance imaging studies in 62 neonates with congenital cardiovascular disease and post-operative magnetic resonance imaging studies in 53 of these patients [39]. They identified pre-operative brain injury in 39% of patients, most commonly stroke, and post-operative brain injury in 35% of patients, most commonly white matter injury. Infants with single ventricle physiology and aortic arch obstruction were particularly prone to exhibit post-operative brain injury. Risk factors for post-operative brain injury also included low intra-operative cerebral hemoglobin oxygen saturation during the myocardial ischemic period of cardiopulmonary bypass and a low mean blood pressure on the first post-operative day.

Partridge and colleagues have recently applied a new technique, diffusion weighted magnetic resonance imaging, to the study of young infants with congenital heart disease before and after surgery [40]. Diffusion weighted magnetic resonance imaging facilitates visualization and quantitative characterization of white matter pathways in vivo. They demonstrated that tractography-based methods facilitated by diffusion tensor imaging detected significant differences in pyramidal tract development among injured and non-injured infants that were not observed on conventional magnetic resonance images. These preliminary data suggest that advanced structural imaging of the brain with magnetic resonance has great promise.

There is evidence that altered cerebral blood flow patterns occur in many post-operative patients with congenital cardiovascular disease. In addition to providing anatomical and metabolic assessments of the brain in these patients, magnetic resonance imaging techniques can quantify their cerebral blood flow. Several recent studies have demonstrated the potential of magnetic resonance imaging in this regard.

Fogel and colleagues noted that the bidirectional Glenn procedure, which is commonly used in the
staged repair of functional single ventricles, and is incorporated into the Fontan circulation, places the cerebral circulation in a series connection with the pulmonary circulation [41]. Since the autoregulatory mechanisms of the cerebral and pulmonary vasculature respond in opposing fashions to hypercarbia and hyperoxia, they questioned which autoregulatory effect would predominate in patients receiving the procedure. They used magnetic resonance velocity mapping to study 12 intubated and ventilated patients who had undergone bidirectional Glenn procedures. Patients were evaluated with 100% inspired oxygen, on room air, and with 3% inspired carbon dioxide. The magnetic resonance flow measurements under these conditions demonstrated that the cerebral response to carbon dioxide overrode the pulmonary response, since total cardiac index, total jugular flow, and the percentage of cardiac index flowing to the brain and lung all substantially increased with hypercarbia. The response to hyperoxia was balanced, with no significant change in cerebral blood flow, pulmonary blood flow, or cardiac index. These findings potentially have important implications for the management of patients with single ventricle physiology.

Phase contrast magnetic resonance imaging can measure blood flow in large vessels. Tissue perfusion can also be quantified with magnetic resonance with the arterial spin labeling method. Licht and colleagues applied this method to the study of 25 term infants with congenital heart defects prior to surgery [42]. They found that pre-operative cerebral blood flow was low compared to accepted normal values. Cerebral blood flow response to hypercarbia was tested in the patients and also found to be relatively low. Some technical details remain to be resolved in very small patients, but the concept of employing arterial spin labeling to characterize post-operative cerebral perfusion is obviously of great interest.

It is clear that neurological magnetic resonance imaging will play an important role in optimizing interventions for congenital cardiovascular disease so that these interventions are not only effective in treating the cardiovascular disorders, but also avoid precipitating nervous system damage. Much additional investigation is needed in this area.

References


Introduction

The last 20 years have been witness to phenomenal advances in the field of magnetic resonance imaging (MRI) which, in turn, have enabled practical developments using this imaging modality to guide interventions [1]. Conventional X-ray fluoroscopy-guided cardiac catheterization and interventions carry a substantial risk of exposure to ionizing radiation for both patients and staff [2,3]. This is currently particularly relevant in younger patients who often need multiple procedures. The need for an imaging modality which offers multiplanar or even three-dimensional (3D) imaging, superior structural delineation of complex cardiac anatomy and additional physiological information without the risk of ionizing radiation has made MRI a very attractive technique to use, particularly in young patients. However, it is only during the last few years that clinical programs using MRI guidance for cardiac catheterization have started and are now showing promising results [4].

After the first MRI images demonstrating live human anatomy were produced [5,6], this technique evolved to enable a variety of clinical applications of MRI. Over the years, improvements in signal detection, data-handling, pulse sequences, artifact suppression and understanding of spin systems have resulted in much faster examination times, considerable improvements in image resolution and have enabled three- [7], or even time-resolved four-dimensional imaging [8,9]. These ultra-fast imaging techniques form the basis of real-time cardiac MRI imaging used for magnetic resonance (MR)-guided cardiac catheterization. However, the first important step in making MRI cardiac catheterization a clinical reality is the design of a suitable interventional MRI system and MRI-compatible catheter material [10,11].

Interventional MRI systems

In designing an interventional MR suite, one has to retain the full capabilities of a high specification diagnostic scanner without encumbering the interventionalist or creating the risk of high radiofrequency (RF) or switched magnetic field exposure. Open magnet designs allow easier access to the patient but are typically not available in field strengths higher than 1 Tesla. The cylindrical horizontal bore systems can offer higher field strengths and gradient slew rates allowing higher resolution imaging, shorter examination times, higher signal-to-noise-ratios (SNR), reduced image distortion and improved functionality by way of real-time imaging. All these factors are of paramount importance when endovascular interventions are being considered [12]. A trade-off with the traditional cylindrical magnet designs is access to the patient. More recently, magnets with shorter bores and flared margins have been designed, offering better patient access, especially for cardiovascular inter-
ventions, without compromising the advanced MR features of diagnostic scanners.

Rapid improvements in the processing power of computers, multiple channel coil developments [9] and powerful, intuitive software design have allowed researchers to develop novel image data acquisition and reconstruction strategies. Combining all these techniques with contemporary graphics hardware and software, recent parallel imaging techniques and new reconstruction techniques, it is now possible to achieve frame rates as high as 20 images/sec [8,13]. Hence, suitable spatial resolution for interventional applications is provided [14].

Despite the inherent potential and promise of MR-guided interventions and operations, it is still not possible to perform the complete procedure within the MR scanner. Currently there are no dedicated safe MR compatible catheters and devices for cardiovascular interventions in humans commercially available. Therefore the immediate future of interventional MRI lies in exploiting multi-modality imaging such as XMR (X-ray and MRI) or XMR and ultrasound. Such hybrid units are already in existence and allow the separate or combined use of different modalities (Figure 20.1).

With cross-modality image integration, both spatial and temporal information can be provided to the users of these systems, allowing better interrogation of anatomy and pathology and of therapeutic devices. A good example of this is the XMR system which combines X-ray and MRI by having both modalities in the same room with a tabletop design that allows patients to be moved from one modality to the other in a very short time (Figure 20.1) [15,16]. This system additionally

![Figure 20.1: An XMR system combines X-ray and MRI by having both modalities in the same room with a tabletop design that allows patients to be moved from one modality to the other in a very short time. The table position is stored within the system allowing image fusion between the MRI and X-ray system (XMR) or even other imaging modalities (for example echocardiography). This system additionally allows the safe use of electronic devices, such as echocardiography machines and computer equipment, in the scanner room beyond the 5-Gauss line.](image-url)
allows operators to safely use other systems such as echocardiography machines and computer equipment for cardiac ablation procedures.

**Advantages of MR-guided procedures**

**Improved visualization of cardiac and vascular anatomy**

A failing of X-ray fluoroscopy-guided cardiac catheterization is the inherent poor contrast of soft tissues such as the heart and great vessels. This in turn makes it difficult for the cardiologist to position guidewires, catheters, balloons and interventional devices within the heart and the surrounding vessels. A skilled operator usually relies on recognizing anatomical structures from previous experience or on contrast angiographic images acquired earlier in the procedure. The lack of adequate visualization increases the risk of perforating the heart and/or great vessels especially when performing complex interventional procedures.

Certain interventional cardiac procedures involve the selection of an appropriate cardiac device and its successful deployment within the heart or the great arteries. This requires accurate measurement of the size of vessels or cardiac defects and adjacent anatomical structures (for example coronary arteries). Such measurements are possible under X-ray angiography but, as this technique relies on two-dimensional projectional images, this can often be difficult.

A successful interventional cardiac procedure therefore relies heavily on adequate visualization of the heart or vessel. This implies the need for superior imaging methods including 3D imaging, which provides excellent visualization of 3D structures in the patient without increased risk. This role fits MRI very well as it provides exceptional 2D and 3D structural delineation of both the heart and its surrounding vasculature, including the coronary arteries. It therefore allows safe guidance of interventional procedures.

**Reduced ionizing radiation and contrast agent**

There is a pressing need for pediatric cardiac catheterization procedures to be made safer, especially in terms of ionizing radiation [2,3]. According to the National Radiation Protection Board, UK (NRPB) the mean risk of developing a solid tumor as a result of a single cardiac catheterization procedure is approximately 1 in 2500 in adults. In children, this risk increases to 1 in 1000, if exposure occurs at five years of age [17]. Also, the proportion of the body that is irradiated increases as the size of the patient decreases and some procedures on patients with congenital heart disease often require much longer X-ray exposure. The risks are multiplied in children in particular, as they often undergo multiple cardiac catheter procedures. The amount of contrast agent is significantly reduced with cardiac MRI and they are usually less nephrotoxic than iodinated contrast agents. It should also be noted that there is also a significant risk from ionizing radiation to the staff in the catheter laboratory during X-ray fluoroscopy-guided procedures, despite the use of protective shields [18].

**Physiological information**

Cardiac catheterization is used not only to provide anatomical information and perform intervention but also to obtain functional information. Invasive pressures and blood gases are used to calculate systemic and pulmonary blood flow and resistance using the Fick principle. Cine angiography is also used to assess global ventricular function, as well as regional wall motion abnormalities. The functional information obtained at cardiac catheterization is used alongside the anatomical information to assess the suitability for surgery or interventional cardiac catheterization or the need for long-term vasodilator therapy in patients with pulmonary vascular disease.

The Fick principle to quantify flow is dependent on multiple measurements (hemoglobin, aortic/pulmonary artery oxygen saturations, partial pressures, oxygen consumption), which can be a considerable source of inaccuracy. In addition, in patients with large intracardiac shunts and high pulmonary blood flow, the accuracy is further reduced [19,20]. Therefore, there is a need for a method of flow quantification that allows accurate and reproducible measurement of pulmonary vascular resistance (PVR). Velocity encoded phase contrast magnetic resonance (MR) enables non-invasive quantification of blood flow in major vessels. Cardiac output and the pulmonary to sys-
Interventional magnetic resonance imaging

Proposed tracking strategies can be separated into passive, hybrid, and active tracking approaches [29–31]. Passive techniques are comparable to X-ray fluoroscopy and computed tomography (CT), where the device itself is imaged without additional hardware. Active tracking techniques employ small receiver coils, incorporated in the catheter device for signal reception and/or transmission, necessitating hardware modification and extended signal-processing to achieve instrument localization.

Some approaches share both passive and active characteristics and are therefore not clearly distinguishable from the previous mentioned types—therefore called hybrid techniques [32].

**Passive catheter tracking techniques**

With passive tracking techniques, the interventional device can be made visible by creating a localized region of signal attenuation or contrast enhancement leading either to negative or positive contrast. The ideal passive tracking catheter or guidewire needs to be made of a material that provides adequate torque and allows tracking while not obscuring underlying anatomy [11]. Ferromagnetic materials cause large susceptibility artifacts and are therefore not generally suitable for MR-guided procedures. This rules out most metals employed for making cardiac catheter devices. However, certain alloys such as nitinol (nickel and titanium) have magnetic susceptibility close to that of tissue and are therefore best suited for making guidewires and braided catheters that are MRI compatible but these are not necessarily MR safe. The polymeric materials used for making catheters typically have low magnetic susceptibilities and therefore cannot be easily localized in MR images [33]. This implies that if materials with higher susceptibility (positive contrast) can be incorporated into the wall of the catheters or sheath or if the lumen can be filled with a suitable contrast agent, then improved visualization can be achieved [11].

**Negative contrast** is created by a signal void caused by the instrument due to the displacement of signal-creating spins and by taking advantage of susceptibility effects. A large local loss in signal is achieved due to intra-voxel dephasing. Unfortunately, this phenomenon leads to local geometric distortion of the underlying vascular anatomy.
Furthermore, the effect depends on a number of factors, such as field strength, applied pulse sequence parameters and device orientation. This results in inconsistent visualization of the instrument.

In the case of balloon angiographic catheters, if the balloon is inflated with carbon dioxide (as occurs conventionally under X-ray guidance), then the inflated balloon creates a signal void in the MR image thus enabling visualization. This method has been successfully employed to guide catheters in patients under MR \[4,34\]. Although this technique allows easy visualization of the tip, the length is impossible to visualize as the signal void from the catheter length is masked by volume averaging and dephasing effects of thicker slices.

A positive contrast can be achieved by using T1-shortening contrast agents. For instance the catheter can be filled with a paramagnetic contrast agent. Applying a T1-weighted imaging sequence, employing short echo and repetition time and high flip angle, the catheter appears as a bright line on the image. A way to make the device darker than its surroundings but let the blood appear bright in the image can be realized by coating the surface of the catheter \[11\]. Alternatively, exogenous nuclei, i.e. perfluorooctylbromide, were used to visualize catheters with high contrast (CNR) and signal-to-noise (SNR) ratios \[35\]. An advantage of passive tracking techniques is the fact that no hardware adaptation of the device to be tracked has to be considered. Hence, passive tracking can be performed on standard clinical systems, thus minimizing potential patient safety issues. The main challenge of all passive catheter tracking techniques is the concern regarding sufficient contrast between the instrument and its surrounding anatomy. Furthermore, pulse sequences for passive tracking are basically imaging sequences visualizing, at the same time, the underlying anatomy and the catheter location. Hence, visibility of the vascular device and spatial-temporal resolution highly depends on the choice of the imaging sequence.

Besides relatively poor contrast compared to active tracking techniques, passive tracking methods are usually based on single-slice imaging to fulfill real-time requirements of the interventional procedures. The acquired field of view (FOV) should ideally be as small as possible to preserve imaging speed, i.e. covering a volume which just comprises the catheter. Unfortunately, the positioning of the FOV is difficult since the geometric extent of the catheter is unknown and a single-slice may not cover the whole curvature of the catheter/wire.

**Active catheter tracking techniques**

In contrast to passive and hybrid tracking methods which visualize the interventional instrument without any hardware modifications, active tracking techniques employ micro-coils and loopless antennae for signal reception.

One approach uses a miniature solenoidal coil, incorporated into the instrument’s tip, for localiza-

In 2D catheter tracking, a single slice of the surrounding anatomy is acquired. This is referred to as the roadmap, onto which the catheter tip is displayed. The current catheter location is usually used to update automatically the position of the slice. In 3D catheter tracking, the tip is depicted on a high-resolution 3D still image. This overlay technique makes it possible to perform the catheter tracking independently from the imaging step. Hence, the roadmap can be acquired in high resolution without sacrificing high temporal resolution during the tracking sequence, which is typically around 10–30 positions per second.

Unfortunately, this method displays only one single point which is often not suitable for flexible instruments. This drawback can be ameliorated by the incorporation of multiple micro-coils but still, the number of displayable points remains quite limited.

To overcome this shortcoming, loopless antennae have been used to outline the shape of the interventional instrument. MR profiling (i.e. acquiring a conventional MR image of the catheter due to its localized sensitivity) was applied during the tracking procedure, but this resulted in a decreased temporal resolution. This, in turn, necessitated the development of more sophisticated data acquisition methods \[36\]. With loopless antennae, it is even possible to image the immedi-
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Sequences such as steady state free precession with low flip angles [44] can be used [37,38]. Catheters with multiple such resonant coils can be tracked easily when compared to passive catheters and have a relatively better safety profile when compared to some of the active catheter designs.

Safety issues

Bioeffects of magnetic fields

The patient having an MRI examination is exposed typically to three forms of electromagnetic radiation, namely a static magnetic field, a gradient magnetic field and a radiofrequency electromagnetic field. Numerous studies have shown no substantial risks to the patients from the electromagnetic fields employed in clinical MR scanners [45,46]. The risks to the healthcare worker especially in an MRI setting is unknown and as with any unknown, the consensus is that more work needs to be carried out before setting occupational electromagnetic field exposure limits [47]. Furthermore, the bioeffects specifically related to the use of interventional MRI have not yet been fully investigated.

Many of the reports in the literature regarding bioeffects of static magnetic fields are conflicting. There is no strong evidence to suggest that there are any significant cardiac or neurological effects from static magnetic fields under 2 T. Several studies have shown that high static magnetic fields do not significantly alter skin and body temperatures [48–50].

Gradient magnetic fields can induce electrical fields and current in conductive media including biological tissue according to Faraday’s law of induction. The thermal effects of switched magnetic fields are thought to be negligible and not clinically significant. Electrical stimulation of the retina is thought to cause magnetophosphenes, which are completely reversible with no known residual side effects. Some volunteers have also reported a metallic taste and vertigo within 4 T magnets. These bioeffects due to gradient fields are unusual in fields of under 2 T [51].

The exposure limits for radiofrequency radiation are set in terms of specific absorption rate in watts/kilogram (W/kg) which is the mass normalized rate at which RF power is coupled with biological tissue. The main bioeffects associated
with exposure to RF radiation relate to the generation of heat in tissues. These effects are particularly relevant in field strengths above 3 Tesla. Controversially, some researchers have reported that electromagnetic fields cause cancer and developmental abnormalities in animal models. However, the efficiency and absorption pattern of RF radiation is mainly determined by the physical dimensions of the tissue in relation to the incident wavelength. This implies that laboratory animal experiments cannot be simply scaled or extrapolated to humans [52,53].

**Heating and electrical safety of interventional equipment**

The heating of wires, devices, implants and other instruments is an important safety issue which is holding back the rapid advance of interventional MRI. Heating due to radiofrequency radiation occurs by three mechanisms according to Maxwell's theory of electromagnetism [54].

When a conductive device or instrument is moved through a magnetic field, small "rings" of current, "eddy currents," are induced creating internal magnetic fields opposing the change. The kinetic energy that goes into driving the eddy currents inside the metal will give off that energy as heat. Therefore intravascular guidewires or device delivery systems with a metal core are unsafe in the MR environment with documented heating up to 74 degrees Celsius (165 degrees Fahrenheit) of the tip [54,55].

Electromagnetic induction heating has often been blamed for thermal injuries caused by monitoring cables used in MRI. The RF electromagnetic fields and the time-varying gradient magnetic fields can induce voltages in conductive media and cause current to flow. The circulating currents cause power loss by heating, which is referred to as induction heating. A loop in a monitoring cable would therefore increase the circuit's inductance and larger currents would be induced resulting in greater heating of the cable [56,57].

If a circuit is in a resonant state, then there is maximum current induction such that significant electromagnetic induction heating occurs. Lengths of wire, for example, can behave as RF antennas that capture electromagnetic waves to extract power from them. The electromagnetic waves that enter the antenna have electric charges associated with them and corresponding currents. When the antenna is approximately half a wavelength long, resonance occurs and the electrical energy remains confined to the immediate vicinity of a given antinode. Hence, the highest electric field of the antenna is thought to be at the tip. The electrical properties of the media surrounding the antenna and the operating frequency also determine the wavelength [56]. Newer designs of wires/cables aimed at reducing heating are being currently investigated along with novel RF shielding technologies [58].

**Magnetic force and torque**

In addition to the bioeffects of MRI and the heating and electrical safety of interventional devices, a significant risk to interventional procedures also exists in the form of magnetic force and torque exerted by the magnetic field on metallic devices [59,60].

Conventional guidewires made of ferromagnetic materials such as stainless steel and catheters with metallic braiding are inherently unsafe for use in the MR environment. Interventional devices that are ferromagnetic will be subject to both deflection force (translational movement) and torque (rotational movement) thereby precluding their use for procedures within a MR scanner. Hence, all MR imaging facilities must have safeguards to ensure that ferromagnetic objects are not brought into the vicinity of the magnet.

There are, however, certain other metallic alloys such as nitinol that are MR compatible. Not only do these alloys produce minimal susceptibility artifacts but they are also unaffected by the magnetic field in terms of deflection force and torque. However, as conducting materials, they are still susceptible to RF heating. This is an important consideration in the development of suitable catheters and guidewires for use in interventional MRI procedures [10,11].

**XMR guidance**

**XMR facility design**

The room design of a typical XMR facility is outlined in Figure 20.1. There are many design features that make this room different from standard MRI facilities.
A great deal of thought has been given to safety of patients under anesthetic, especially during the transfer between the X-ray and MR tables. All the anesthetic monitoring tubing and lines are designed with extra length and are secured to the movable tabletop to ensure smooth patient transfer.

The ECG and invasive pressure data is sent from the MR-compatible monitoring equipment via an optical network to a computer in the control room, where the cardiac technician is stationed. The appropriate measurement and recording of the data is made in the usual way. The technician has access to monitors which show the appropriate X-ray or MR images of the procedure.

Reliable and accurate ECG synchronization is essential for cardiovascular MRI and in particular, MR-guided cardiac catheterization. When manipulating catheters in the heart, there is the potential to cause arrhythmias (tachyarrhythmia and/or heart block). It is therefore important to perform accurate monitoring of the cardiac rhythm at all times during XMR catheterization. Obtaining a reliable ECG in the magnet particularly during some MR sequences can be difficult. The magneto-hydrodynamic effect and gradient noise can seriously disturb the ECG signal [61]. Vector electrocardiogram (VCG) is a QRS detection algorithm which automatically adjusts to the actual electrical axis of the patient’s heart and the specific multi-dimensional QRS waveform [62]. The VCG produces a reliable R-wave with clearly discernible P and T waves. This allows detection of nearly all arrhythmias. Unfortunately, there are currently no ECG systems that can reliably provide ST segment or T-wave morphological information. In the future, however, using signal processing techniques, it may be possible to obtain an ECG during MRI scanning that provides ST segment and T wave information reliably.

Another complication of performing cardiac catheterization under MR guidance is the noise generated during scanning. There is a headphone and microphone system in the room that reduces the noise and allows staff to communicate with each other between the scanner and control rooms.

Some MR coils have X-ray-visible components and would need to be removed between MR imaging and X-ray imaging of patients. It is there-

Table 20.1 XMR facility-safety features.

<table>
<thead>
<tr>
<th>Feature</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Compulsory safety training of all MR interventional staff</td>
<td>Mandatory training for all staff members.</td>
</tr>
<tr>
<td>2. Specially designed clothes without pockets</td>
<td>Special clothing that does not contain pockets to prevent the introduction of non-ferromagnetic objects.</td>
</tr>
<tr>
<td>3. Safety officer restricting entry to the main room during XMR intervention</td>
<td>Safety officer ensures restricted entry to prevent accidental entry of personnel during XMR intervention.</td>
</tr>
<tr>
<td>4. Clear demarcation of ferromagnetic safe and unsafe areas within the room</td>
<td>Clear demarcation to prevent the movement of ferromagnetic objects.</td>
</tr>
<tr>
<td>5. MR compatible anesthetic and monitoring equipment</td>
<td>Equipment designed to function reliably in the MR environment.</td>
</tr>
<tr>
<td>6. Noise proof headphone systems for all staff within the room</td>
<td>Noise-protected headphone systems to prevent interference from MR noise.</td>
</tr>
<tr>
<td>7. X-ray and RF shielded room</td>
<td>Shielded room to prevent interference from X-ray and RF equipment.</td>
</tr>
<tr>
<td>8. Positive pressure air handling and filtration system</td>
<td>System to maintain a positive pressure in the room to prevent the entry of air.</td>
</tr>
<tr>
<td>9. Tethering of all ferromagnetic equipment to the wall/ floor</td>
<td>Tethering to prevent movement of ferromagnetic equipment.</td>
</tr>
<tr>
<td>10. Safety checks whenever patient is transferred between X-ray and MRI to ensure that metallic instruments used for catheterization are not taken across to the MRI end of the room</td>
<td>Safety checks to ensure metallic instruments are not transferred to the MRI section.</td>
</tr>
<tr>
<td>11. Written log of all safety infringements and regular review of safety procedures</td>
<td>Keeping a record of safety violations and regular review to ensure compliance with safety protocols.</td>
</tr>
</tbody>
</table>

Abbreviations defined previously in the chapter.
fore necessary to have specifically designed coils
that are sufficiently radio translucent to be left in
place during X-ray imaging without any deteriora-
tion of image quality. We use these coils in our
procedures so that the patients do not have to be
disturbed when moving from one imaging modal-
ity to the other [63].

The XMR suite has positive-pressure air han-
dling and filtration appropriate for a catheteriza-
tion laboratory. There is a scrub room which is also
RF and X-ray shielded and can be accessed both
from the XMR suite and control room. This room
acts as an RF lock, allowing access to the XMR suite
during MR scanning.

**Performing XMR procedures**

In general terms, there are two ways in which the
XMR laboratory can be used.

**X-ray imaging as a backup during MRI**

The cardiac catheterization procedure is performed
in the MR scanner after adequate arterial and/or
central venous access has been obtained in the MR
safe area of the XMR laboratory. The MR com-
patible monitoring and anesthetic equipment are
attached. Flexible phase array RF coils are used.
These coils are relatively radiolucent and thus do
not need to be removed between MR and X-ray
imaging. The patient is then placed in the MR
scanner after safety checks, including an operating
theatre-style check of all metallic objects used in
the MR safe area. Imaging is then performed for
assessment of morphology, function and flow
quantification of the heart and great vessels. With
real-time imaging, the end-hole or side-hole
balloon angiographic catheter (4F–7F) is passively
visualized with the balloon inflated with CO₂. It
then can be tracked to the appropriate location for
pressure measurement or deployment of a device.
A duplicate MR control console is positioned next
to the bore of the magnet so that the interactive
window can be easily visualized while the catheter
is being manipulated. Once the catheter is posi-
tioned in the desired vessel or chamber, the appro-
priate pressure data and saturation/blood gas
samples are obtained as in routine cardiac catheter-
ization. If catheter manipulation into a particu-
lar heart chamber or vessel using MR guidance
alone is difficult, the patient is transferred back to
the X-ray end of the room, where the catheteriza-
tion can be continued under X-ray fluoroscopy
(for example to use a guidewire or a braided cath-
ter). The patient can be transferred back to the
MR scanner for further MR measurements once
the catheter is satisfactorily positioned.

**Performing X-ray fused MR-guided interventions**

Interventional procedures or radiofrequency abla-
tion of arrhythmias necessitate part of the proce-
dure to be performed under X-ray fluoroscopy, as
the ablation catheters and delivery devices are not
MR compatible. Therefore, MR imaging is per-
formed at the beginning of the procedure for plan-
ing, during the procedure for guidance and at the
end of the procedure for evaluation.

Recently, image fusion technology has been
developed for fusion between high resolution MR
imaging and electrophysiological models. Several
systems are on the market and are currently being
evaluated for clinical utility [64,65]. Even though
there seems to be an inherent advantage for this
technique, further clinical studies are needed to
prove these systems in the clinical setting.

There are two main methods that have been pre-
sented for the fusion of MRI data to X-ray fluoro-
scopy data in the XMR setting:

1. Optical tracking and calibration of the XMR
   system [15,66].

2. Multimodality markers attached to the patient
   that are both MR and X-ray visible [67].

One advantage of the tracking-based method is
that the calibration procedure is only required
once and remains very stable over the course of
many years. Another advantage is that image
overlay is possible for any anatomical region
regardless of location, as long as it is accessible for
both MRI and X-ray imaging. The tracking-based
method has the disadvantage that it assumes that
the patient remains stationary on the patient table.

The marker-based method has the advantage
that it can track and accommodate for bulk patient
motion. However, it has the disadvantage that it
can only be applied to anatomical regions around
which markers can be placed on the skin in a reli-
able manner. Skin markers are susceptible to
motion that will alter their relative configuration.
Using these techniques a number of clinical cardiac applications are listed (Table 20.2).

**Early experience in humans and future perspectives**

In our center, MR is most frequently used to assess pulmonary vascular resistance in patients as it...
Figure 20.3 A 13-year-old patient was invasively treated for right ventricular outflow tract tachycardia. The geometric model, based on electrophysiology data (a) and the high resolution 3D magnetic resonance (MR) surface-rendered model (b), are presented side by side. Their spatial orientation within the thorax are shown by the thorax model (T). Their spatial coordinates are connected and movement of the electrophysiology model (a) results in the appropriate motion of the corresponding anatomic MR model (b). The 3D MR model (b) shows better anatomical details of the superior vena cava (SVC), inferior vena cava (IVC), right ventricular apex (RVA) and the right ventricular outflow tract (RVOT). The right coronary artery (RCA) can be seen in the 3D MR surface-rendered model (b), but not on the geometric model based on the electrophysiology data (a). The surface (2) and virtual (6 to 10) ECG signal generated from the Ensite Array® catheter (Ens, EnSite System St. Jude Medical) are displayed (c).
Further development of novel catheters and guidewires have been made possible by groups using targeted intra-myocardial injection of progenitor stem cells in animal models of myocardial infarction [85,86]. Also, using real-time MRI and direct apical access in porcine hearts, prosthetic aortic valves have been implanted in the beating heart [87]. This breakthrough application may allow MR guidance of minimally invasive extra-anatomic bypass and beating-heart valve repair. MR guidance of intramyocardial gene therapy is another exciting field [88]. Finally, three-dimensional electromechanical models of the heart have also been created which allow simulation of cardiovascular pathologies in order to test therapeutic strategies and to plan interventions [89].

Conclusion

MR guidance of cardiac catheterization is feasible as outlined in this chapter and has been shown to be safe. The potential benefits from this new technique include a reduction in ionizing radiation exposure, accurate assessment of pulmonary vascular resistance and better visualization of complex anatomy for both diagnostic and interventional (for example stent or other device implantation, radiofrequency ablation of arrhythmia etc.) cardiac catheterization.

However, a number of issues need to be resolved before exclusive interventional cardiovascular MRI becomes routine clinical practice. These include practical issues such as reduction of noise in the MR environment, improved access to the patient and better MR compatible patient monitoring equipment. There is also a pressing need for catheter and device manufacturers to produce tools specifically designed for MR-guided cardiac catheterization. This, combined with the cost of installing XMR suites, is holding back the rapid advance of the interventional CMR. However, for interventional cardiologists, radiologists and surgeons, the prospect of 3D anatomical guidance together with useful additional anatomical and physiological information and the ability to assess tissue response to therapy using MRI makes this remarkable imaging modality unique, offering great promise for safe guidance of complex cardiovascular interventions.

Future perspectives

Several groups have demonstrated the immense potential of interventional cardiac magnetic resonance (CMR) in animal models. The interventions have been shown to be feasible with passive and active catheter techniques ranging from balloon angioplasty of arterial stenoses [75–77] and stenting of vessels [78–80] to atrial septal puncture and atrial septostomy [81]. Device closure of atrial septal defects is another application that has been explored [59]. MRI guided percutaneous pulmonary and aortic valve stent implantation has also been successfully carried out [78,82].

More complex interventions such as percutaneous coronary catheterization and intervention have also been demonstrated in healthy animals using MR [83,84]. Balloon dilation of aortic coarctation in patients under MR guidance has also been performed [77]. Recently, new MR compatible guidewires and catheters have become available and interventions in patients with congenital heart disease under MR guidance should be possible in the near future [11].
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CHAPTER 21

Adult congenital heart disease

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**Introduction**

Progress in surgical techniques, interventional cardiovascular treatments, and medical therapies have allowed more children with congenital heart disease to live well into adulthood. A cardiovascular subspecialty practice has been developed to recognize the growing importance of this group of patients, known as Adult Congenital Heart Disease (ACHD). These patients may have undergone a myriad of repairs or palliative procedures during their lives, and frequently require serial evaluation of their cardiovascular anatomy and function. Often, surgical and medical records are unavailable and families have incomplete databases or recollections regarding previous therapies. Cardiac magnetic resonance (CMR) has evolved as a mature technique for anatomic and functional evaluation of ACHD patients. However, the requirement for, and risks of, radiation and contrast agent exposure limit the applicability of cardiac computed tomography (CCT) in this patient group. Not infrequently, chest wall abnormalities, post-operative changes, anteriorly-placed conduits or other technical limitations lead to inadequate echocardiography evaluations. CMR does not suffer from these limitations, and can image many patients with poor echocardiography windows. Its superior soft tissue depiction and its greater ability to characterize myocardial viability, anatomy, and physiology make it an ideal method for serial assessment of ACHD patients. The purpose of this chapter is to review the current status of CMR in ACHD and to discuss the most common indications for the technique, including pre- and post-operative assessment of this unique patient population.

**General CMR examination principles**

The standard CMR examination in patients with ACHD generally consists of gathering a set of scout or localizer images upon which additional acquisitions are based. Next, single-slice cardiac-gated cine CMR sequences (using a steady-state free precession [SSFP] sequence) in the 4-chamber, 2-chamber, and apical long axis (3-chamber) views are performed in order to optimize the standard orientations for further functional and anatomic evaluation. Turbo (or “fast”) spin echo images (with and without blood suppression) in the axial plane may then be performed to depict the major anatomic features of the ACHD patient. If delayed enhancement imaging is needed, other cine imaging is sometimes performed after contrast injection (and perfusion imaging, if desired), during the 10–15 minute waiting period. This approach improves scan efficiency, and provides greater endocardial definition for later image analysis/segmentation. The multiphase short axis cine images are later used to quantify right (RV) and...
left ventricular (LV) end-diastolic (EDV) and end-systolic (ESV) cavity volumes, and myocardial mass as part of the serial functional evaluation. The cavity volumes are used to calculate RV and LV ejection fraction (EF) by the following relationship: \((EDV - ESV)/EDV = EF \times 100\) (in %). Velocity-encoded cine CMR (VEC) flow measurements are made to measure cardiac output, pulmonary-to-systemic flow ratio \((Qp/Qs)\) in the case of possible shunts, and to quantify valvular regurgitation or stenosis. High-quality magnetic resonance angiographic (MRA) images are acquired using a 3D sequence to depict the size, morphology, and course of vessels in the thorax and/or abdomen. The advantage of this 3D dataset is that operators are permitted to navigate or “fly” through the image space at any oblique angle and produce reformatted images representing the complex vascular anatomy frequently found in ACHD patients. Recently, a 3D SSFP technique with ECG and respiratory navigator gating has been developed that provides high-quality bright-blood images of the vasculature without the use of contrast agents. Parallel imaging methods or sensitivity encoding (SENSE) imaging are frequently used to accelerate image acquisition with the goal of completing a full examination in 30–60 minutes, if possible.

**Tetralogy of Fallot**

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease, and refers to a congenital cardiac malformation combining subpulmonic obstruction, a ventricular septal defect, right ventricular hypertrophy, and a “so-called” overriding aorta (see Chapter 10). These phenomena all derive from a single fundamental abnormality: anterior, cephalad, and leftward deviation of the infundibular or conal septum. As a direct anatomic result, there is subpulmonic stenosis, anterior and cephalad displacement of the aortic valve which sits over the ventricular septum (“overriding aorta”), and a malalignment ventricular septal defect. The subpulmonary stenosis eventually leads to right ventricular hypertrophy. The classic presentation of TOF is cyanosis, often paroxysmal (“Tet spells”), related to abrupt decreases in pulmonary blood flow. The first effective surgical approaches involved increasing pulmonary flow by means of systemic-to-pulmonary arterial shunts. Techniques for complete repair of TOF were later developed, and recent surgical therapies have followed an early definitive repair strategy without preceding palliative shunt. Nevertheless, many current adult patients with repaired TOF have had prior shunts. TOF spans a wide anatomic spectrum, from very mild malalignment with absent pulmonary valve to pulmonary atresia.

**Variations in RV outflow tract obstruction**

There is a wide variety in the clinical severity of outflow tract obstruction. TOF with absent pulmonary valve is a relatively rare variant that incorporates the criteria for diagnosis of TOF, but includes the absence of a functional pulmonic valve, and often the presence of large branch pulmonary artery (PA) aneurysms with the potential for compression of the bronchi, pulmonary veins, and coronary arteries. These patients usually present early in life, and have a worse prognosis. On the other hand, TOF with pulmonary atresia is an extreme form of TOF, with complete obstruction to RV outflow to the pulmonary artery. Pulmonary blood flow frequently depends on a patent ductus arteriosus and there are often multiple aorto-pulmonary collateral vessels. Pulmonary arteries are often small in patients presenting with TOF, likely as a result of the decreased pulmonary blood flow. Maneuvers to augment pulmonary blood flow including shunts and total repair stimulate growth of the PAs. Nevertheless, many adults with repaired TOF have branch PA stenosis. In addition to the decreased blood flow during development, PAs appear prone to stenosis at sites of attachment of systemic-PA shunts. These lesions can have significant hemodynamic consequence both via pressure overload of the right ventricle and by increasing the severity of pulmonary valvular regurgitation. CMR evaluation should include description of the anatomy and physiology of these lesions. Velocity-encoded phase contrast imaging across the lesion can define the pressure gradient and directly provide an estimate of antegrade and retrograde flows. Differential blood flow to the right and left lungs should also be described [1].
Associated abnormalities
A right aortic arch is present in approximately 25% of patients with TOF. Other abnormalities of aortic, head and upper extremity vessels are less common, but are associated with 22q11 deletion [2]. These findings have clinical significance and the aortic arch anatomy should be described in detail. Approximately 5% of patients will exhibit an aberrant left anterior descending (LAD) coronary artery arising from the right coronary artery and coursing anteriorly across the right ventricular outflow tract (RVOT). This anatomy precludes incision of the RVOT, an approach commonly used to relieve severe infundibular stenosis. Identification of this course, especially for primary repair but also for repeat surgery, is crucial to avoid transection of the LAD and consequent myocardial infarction. A small subset of adult patients will have CMR evidence of anterior myocardial scar as a result of surviving this potentially catastrophic event.

Role of CMR in pre-operative assessment
The majority of adult patients with TOF who present for evaluation will be after palliative or definitive repair. However, a brief description of the surgical approach to unrepaired TOF patients is useful. The major goal in evaluating unrepaired TOF patients for surgical treatment is to define the location and severity of RVOT obstruction, the relative size of the ventricular septal defect (VSD), degree of left to right shunting, and the sources of pulmonary blood flow (pulmonary artery sizes and presence of stenoses, aortopulmonary collaterals, patent ductus arteriosus).

Surgical repairs – shunts
The purpose of these systemic-to-PA shunts is to increase pulmonary blood flow and to increase the size of the PAs for later definitive repair. The contemporary approach to children with TOF is usually primary total repair, but most adult patients with TOF have had a shunt prior to intracardiac repair. The palliative shunt is usually interrupted during the total repair procedure, but may still be present and should be documented by CMR studies. A minority of patients have only a palliative shunt, and evaluation of patency and flow is important in these patients. The various types of shunt are outlined in Table 21.1.

<table>
<thead>
<tr>
<th>Name</th>
<th>Anatomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Classic Blalock–Taussig</td>
<td>Subclavian artery is ligated and connected to a branch pulmonary artery (PA), usually on the opposite side of the aortic arch</td>
</tr>
<tr>
<td>Modified Blalock–Taussig</td>
<td>Uses graft material to connect the subclavian artery to a branch PA.</td>
</tr>
<tr>
<td>Potts</td>
<td>Anastomosis between descending aorta and left PA.</td>
</tr>
<tr>
<td>Waterston</td>
<td>Ascending aorta to right PA with side-to-side anastomosis</td>
</tr>
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Abbreviations are defined in the text.

Definitive surgical repair
Repair of TOF involves relief of RVOT obstruction (infundibular, valvar and supravalvar) and closure of the VSD. The diversity of anatomy and associated features of TOF, however, requires a wide range of surgical techniques with corresponding long-term complications.

- Transannular patches (TAP)
  - Many patients with TOF have a hypoplastic pulmonary valve annulus, requiring an incision with patching for adequate augmentation/enlargement of the annulus and RVOT. The patch extends for a variable distance into the RVOT and along the PA and can be definitively identified by delayed enhancement CMR techniques [3]. While this approach provides excellent relief from obstruction, it also undermines the structural support of the pulmonary valve leading to a significant degree of pulmonary regurgitation and depending on the RV extent of the patch, can also directly contribute to RV dysfunction.
  - For several decades after TOF repair was introduced, surgeons focused on ensuring complete relief of RVOT obstruction with little consideration paid to RV geometry or residual pulmonary regurgitation which was felt to be a benign lesion. Consequently, pulmonary regurgitation and its sequelae are the most common indications for reoperation in repaired TOF. Currently, there is more tolerance of
residual RV obstruction, but TAPs are frequently required in the presence of a severely hypoplastic pulmonary annulus.

- **Conduits**
  - In a subset of patients, the native RVOT cannot be surgically repaired, often due to pulmonary atresia or aberrant coronary anatomy. Conduits constructed of natural or man-made materials (homograft or prosthetic tube graft) – with or without a valve to prevent regurgitation – are commonly used between the RVOT and pulmonary artery. Conduits placed early in life usually require replacement as the older patient requires a higher cardiac output than its small diameter can sustain, or due to development of hemodynamically significant conduit dysfunction (stenosis or regurgitation). Note that conduits are often difficult to visualize by echocardiography due to their retrosternal position which makes CMR evaluation advantageous. However, conduits can become heavily calcified which may need special CMR techniques to visualize these structures optimally.

- **Unifocalization of aortopulmonary collaterals**
  - Patients with the pulmonary atresia form of TOF frequently have inadequately sized proximal pulmonary arteries. When feasible, the aortopulmonary collateral (APC) vessels supplying the pulmonary blood flow can be “unifocalized” allowing connection to the native RVOT directly or with a conduit. CMR assessment of these APCs frequently aids surgical planning.

- **Transatrial ventriculotomy approaches**
  - Early approaches to relief of RVOT obstruction and VSD repair involved right ventriculotomy. This approach results in residual RV scar of variable size, increasing the risk of arrhythmia and sudden death, as well as contributing to RV dysfunction. When possible, repair via an atrial approach is now favored, which decreases the likelihood of these long-term complications [4].

**Role of CMR in post-operative assessment**

Adolescent and adult patients have limited echocardiographic windows and CMR assumes an important clinical role, particularly for quantitative imaging [5]. Its outstanding soft tissue depiction capabilities permit detailed examination of the RVOT, which frequently exhibits aneurysmal dilatation following patch repair, and complicates measurement of RV function (Figure 21.1). CMR can evaluate the location and severity of residual RVOT obstruction, from dynamic obstruction due to hypertrophied RV muscle bundles to stenotic valved conduits as well as the severity of pulmonary regurgitation (PR) following repair. CMR holds a major advantage over echocardiography (echo) in measurement of regurgitant flows. The use of VEC techniques permits accurate measurement of the forward as well as regurgitant flow and thus the regurgitant fraction which aids in assessment of RV function and candidacy for pulmonary valve replacement in patients with chronic severe PR [6,7]. Quantification of flow to each lung can provide indirect evidence of the presence of branch PA stenosis. A further refinement of the VEC technique permits estimation of the severity of relative right and left pulmonary vascular resistances [8]. Left to right shunting can be quantified in order to measure the impact of residual VSDs or VSD patch leaks when present. CMR is also superior to echo for accurate measurement of RV volumes and function since contiguous multislice tomographic images can be acquired and quantified without the need for geometric models or other error-prone assumptions [9]. This feature permits serial non-invasive evaluation of ventricular volumes and function to detect trends in RV (and LV) sizes and performance, especially under chronic volume overload conditions such as PR, which promotes RV dilatation and dysfunction.

**CMR predictors of outcome in repaired TOF**

**Pulmonic regurgitation and regurgitant fraction**

Progressive RV dilatation and functional deterioration is unlikely if the regurgitant fraction (RF) is less than moderate (RF < 20%), however, other factors such as RV diastolic compliance are important factors in long-term outcomes related to PR.

**RV function and size**

Right ventricular dysfunction in TOF is commonly seen and multifactorial in origin. RV dysfunction
Figure 21.1 Adult after tetralogy of Fallot (TOF) repair. (a) Gadolinium enhanced magnetic resonance angiography in a patient with a right ventricular outflow tract (RVOT) aneurysm after distant TOF repair (oblique coronal plane). Gradient echo (b, bright-blood) and spin echo (c, black-blood) parasagittal short axis images are shown for comparison. A similar slice is shown after aneurysm resection and pulmonary valve replacement, with a notable lack of RVOT dilatation (d). In a different patient with repaired TOF, there is quantitative and qualitative evidence for moderately severe pulmonic regurgitation and a regurgitant fraction of 34% (e-g). Note the forward/antegrade systolic flow (positive y axis on graph (e)) and negative/regurgitant diastolic flow (negative y axis on graph (e)) across a slice oriented perpendicular (cross-sectional plane) to the proximal pulmonary artery (PA); (f, g). In the oblique axial (PA cross-sectional) magnitude image at the level of the main PA just proximal to the bifurcation (f), the chest wall (CW), left atrium (LA), and pulmonary artery (arrow) can be seen. In the velocity encoded cine image at the same level (g), diastolic pulmonary regurgitant flow is identified by the area of signal void (dark area, arrow). This is the area of regurgitant flow quantitated to produce the diastolic phases of (e) above. H: head; LV: left ventricle; RA: right atrium; RV: right ventricle.
can take several forms including dilation, poor contractility, regional wall motion abnormalities, decreased diastolic compliance causing restrictive physiology, and others. The most consistently reported parameters associated with poor outcome are right ventricular size (both RVESV and RVEDV) and RVEF [10,11].

LV function
While TOF most commonly affects the RV, there is an increased prevalence of LV dysfunction in these patients. The precise etiologies are undefined, but several potential factors include the influence of right ventricular dysfunction and adverse effects due to ventricular interdependence, dyssynchrony due to the very wide right bundle branch block (RBBB) seen in many patients, and myocardial fibrosis. Left ventricular dysfunction has been clearly demonstrated to be an independent risk factor for poor outcomes after repair of TOF, and evaluation of global LV function as well as analysis of regional wall motion is an integral
component of the comprehensive CMR examination in these patients [11,12].

Myocardial fibrosis
In adults with TOF, etiologies of myocardial fibrosis include infarction due to transection of an aberrant coronary, inadequate myocardial preservation intra-operatively, scar due to prior surgical incision or vent placement, and possibly fibrosis due to abnormal ventricular strain patterns. In patients with TOF, presence of greater degrees of late gadolinium enhancement in the RV and LV is associated with worse ventricular function, lower exercise capacity, and arrhythmia [13,14].

Transposition of the great arteries
Transposition of the great arteries (TGA) is defined as a condition comprising discordant connections between the ventricles and the great arteries where the pulmonary artery is connected to the LV, and the aorta is connected to the RV. This condition results in a well-known form of cyanotic congenital heart disease (second in incidence to TOF). The two most common types of TGA are D-loop TGA and L-loop TGA (or “congenitally corrected TGA”). D-loop TGA or TGA [S,D,D], is the most common form of the disease and accounts for nearly 5–7% of all congenital heart disease births. The anatomical designation of the two types of TGA depends on the viscero-atrial situs (situs normal or inversus), the ventricular looping (D or L), and the position of the aortic valve relative to the pulmonic valve (D or L). In D-loop TGA, there is situs solitus (S), a ventricular D-loop (D), and dextro-transposition of the aortic valve (D) or [S,D,D]. There is severe hypoxemia in this condition as the systemic venous blood is directed to the aorta and thus the major organs are fed with oxygen-poor blood, whereas the pulmonary venous return is directed back to the lungs (Figure 21.2). Obviously, survival is dependent on some shunting between the circulations such as an atrial septal defect, VSD, or patent ductus arteriosus. Surgical therapy was devised to redirect systemic venous blood to the lungs and oxygenated blood to the systemic arterial circulation. Until the mid-1980s, this correction was performed at the atrial level and known as an “atrial switch” procedure. The two most common procedures were the Senning and Mustard repairs, with the major difference being that the Senning repair used native atrial tissue whereas the Mustard procedure used pericardial tissue to augment the baffles. The technique consists of forming intra-atrial baffles between the two atria, thus returning deoxygenated blood
across the mitral valve to the morphological LV, then across the pulmonic valve to the lungs. Similarly, oxygenated blood from the pulmonary veins is directed across the tricuspid valve to the morphologic RV (now acting as the systemic ventricle) and across the aortic valve to the systemic circulation. Unfortunately, hemodynamic problems are common in the late post-repair state in this form of ACHD. The most common abnormality encountered is systemic right ventricular failure, the mechanisms of which are not entirely clear at present. This frequently occurs when the patient reaches their mid-40s, and relates in part to abnormal ventricular wall stress/mechanics in a chamber that was not structurally designed to handle chronic systemic pressures. Systemic atrioventricular (tricuspid) valve regurgitation frequently occurs in the setting of RV dilatation and chronic RV dysfunction. Other abnormalities include baffle leaks (systemic and pulmonary, with resultant intracardiac shunting) or outflow tract obstruction, atrial arrhythmias and sinus node dysfunction [15]. From an imaging standpoint, other common associated congenital defects that should be evaluated include VSD, coarctation/interrupted arch, and pulmonic stenosis.

In the mid-1980s, a new surgical procedure known as the arterial switch operation (ASO) became popular, and has demonstrated favorable long-term results [16,17]. This repair switched the great arteries immediately distal to the aortic and pulmonic valves and required reimplantation of the coronary arteries. The major advantages include the return of the LV as the systemic ventricle and the reduction of suture lines in the atria that promote arrhythmias. A common associated repair of the pulmonary arteries with an ASO is known as the LeCompte maneuver where the branch pulmonary arteries are positioned anterior to the ascending aorta. Echo views are very limited in this type of repair due to the retrosternal and near-field location of the branches whereas CMR provides a full assessment of anatomy, flow, and presence of stenoses [18]. Late complications relevant to imaging include suprapulmonary and supra-aortic stenoses, and compromise of the coronary ostia.

The next most common form of TGA is L-loop or [S,L,L] TGA consisting of situs solitus, a ventricular L-loop, and levo-transposition of the aortic valve relative to the pulmonic valve. This disorder is also known as “congenitally corrected TGA” since there is discordance at the atrio-ventricular as well as ventriculo-arterial levels. This double-discordance results in oxygenated blood being delivered to the systemic circulation, but the RV still functions as the systemic ventricle. Concomitant abnormalities can include VSD, subvalvular and valvular outflow tract stenoses, complete heart block, and tricuspid valve disorders. Long-term outcome is dependent on the severity of the associated abnormalities and systemic RV function.

A dedicated CMR exam should include the following objectives: SSFP cine imaging for RV and LV volumes and function, VEC sequences for baffle leaks (with low VENC, i.e., to detect low velocity flow), valvular regurgitation, and sub- or supra-valvular stenoses (with higher VENC, depending on expected jet velocity), and delayed enhancement imaging to evaluate for ventricular fibrosis. In patients after ASO, dedicated imaging of the coronary ostia or a dobutamine or adenosine stress CMR study may be useful to evaluate for coronary artery stenoses.

**Double-outlet right ventricle**

Double-outlet right ventricle (DORV) is a unique malalignment of the ventriculoarterial segment whereby both great arteries arise from the morphologic RV (or infundibulum) (Figure 21.3). To understand the post-repair state and potential complications, it is important to appreciate the various forms of DORV and their similarities to other congenital cardiac disorders. The size and location of the VSD, its relation to the semilunar valves, the presence of semilunar valve atresia, position of the infundibular septum, and the ventricular sizes and function are all integral parts of the database for surgical decision-making. The LV may be normal, hypoplastic, or not present at all. The RV is usually of normal size but may be abnormal also. Some of the more common forms of DORV that resemble other congenital heart diseases from a physiologic standpoint include the TGA type (Taussig–Bing–DORV; with bilateral conus, a subpulmonic VSD +/- aortic outflow obstruction), TOF type (subaortic VSD with sub-
reimplantation. The peculiarities of these complex repairs often require expert attention to performance and interpretation of CMR scans [19].

**Single ventricle/Fontan repair**

While a detailed description of single ventricle physiology is beyond the scope of this chapter (see Chapter 16), more patients each year survive into adulthood with a single RV (e.g., hypoplastic left heart syndrome) or single LV circulation. Since the circulations are initially mixed and associated with systemic cyanosis, the purpose of surgical management is to reduce the dual volume load on this single ventricle by isolating the systemic and pulmonic circulations. This is performed in a staged manner using one of the Fontan procedures (classic, atroventricular [Figure 21.4], fenestrated, extracardiac), which routes systemic venous blood to the pulmonary arteries directly without a pumping chamber. Potential complications relevant to CMR imaging include ventricular dysfunction, atrial/Fontan circuit thrombus formation, Fontan circuit obstruction or baffle leaks, pulmonary vein compression, PA stenosis, and valvular regurgitation. Serial evaluation of ventricular size and function is important for chronic management. Dynamic first-pass imaging of pulmonary perfusion can aid in identifying PA stenosis (Figure 21.4) or other problems altering right vs. left lung perfusion. Evaluation for outflow tract obstruction or valvular dysfunction is recommended, as these disorders can add to chronic ventricular load. Novel techniques to evaluate dynamic flow patterns in the Fontan circuit have furthered the understanding of the complex underlying physiology. Myocardial tagging has also been used to investigate ventricular function in the single ventricle state [20,21].

**Incidental findings**

One must always be diligent for unexpected findings when interpreting images and be cognizant of the fact that congenital and hereditary disorders can present themselves in unique ways. In a patient referred for another indication, it was noted that the LV was symmetrically hypertrophied, but that the degree of hypertrophy was out of proportion to any underlying etiology (no history of hyperten-
origins to their insertions in 2 and 3 dimensions. This is particularly useful in defining abnormalities of the systemic and pulmonary veins. No other imaging technique provides the accuracy, anatomic coverage, ease of display/visualization, and safety as CMR.

Systemic and pulmonary venous abnormalities

The wide fields of view possible with CMR permit depiction of the full course of vessels from their origins to their insertions in 2 and 3 dimensions. This is particularly useful in defining abnormalities of the systemic and pulmonary veins. No other imaging technique provides the accuracy, anatomic coverage, ease of display/visualization, and safety as CMR.

**Systemic venous abnormalities**

Persistent left superior vena cavae (PLSVC) are common isolated findings; however, they attain greater significance when associated with other congenital defects. The typical PLSVC originates at the left brachiocephalic vein and drains to the coronary sinus and then to the right atrium. There is usually a right superior vena cava present, and sometimes a bridging vein connecting the two vessels in the retrosternal space. Alternatively,
**Pulmonary venous abnormalities**

While total anomalous pulmonary venous return is nearly always diagnosed in infancy, partial pulmonary venous return syndromes may not become clinically evident until middle age. The insertions of partial anomalous pulmonary venous return when the right superior vena cava (RSVC) is atretic, the left superior vena cava (LSVC) and coronary sinus are profoundly dilated. An unroofed coronary sinus may also be present, which acts as an interatrial shunt. The clinical significance of these abnormalities relates to intra-operative cannulation planning for cardiac surgery and the potential for shunting of flow away from the pulmonary system after certain types of shunt procedures.

An interrupted inferior vena cava (IIVC) has additional implications for imaging.

Typically, the IVC drains to the right atrium, however, in IIVC, the vessel frequently drains via an azygous continuation to the superior vena cava (SVC) and may even connect to the left atrium. The latter pattern is often associated with heterotaxy syndromes or isomerism and CMR permits accurate assessment of even the most complex anatomy.

**Figure 21.5** Three equivalent short axis images utilizing different CMR techniques in a patient with Fabry’s disease, demonstrating tissue characterization capabilities. (a) Uses HASTE imaging and demonstrates the “dark-blood” feature of the technique and the abnormal but symmetric hypertrophy characteristic of the infiltrative disease. The image shown in (b) was acquired using a balanced steady state free precession which results in a “bright-blood” image. (c) shows mid-myocardial delayed enhancement 10–12 minutes after administration of gadolinium. The intramyocardial hyperenhanced territories are best seen along the anterolateral (upper) and inferior (lower) walls of the LV.
on the heart and determine the optimal time for intervention [24]. Newer, faster techniques promise to simplify these measurements and accelerate image acquisition [25].

**Coarctation/patent ductus arteriosus/truncus arteriosus**

These collective abnormalities of the aorta and/or pulmonary artery are ideally evaluated by CMR techniques. Accurate anatomic depiction of these disorders in adults is frequently challenging for echocardiography due to a number of limitations including narrow retrosternal windows and difficulty identifying tortuous vessels. CMR provides a more comprehensive assessment of the vessel sizes, degree of stenosis and regurgitation of the truncal valve, and shunt fraction compared to the purely anatomic description afforded by CT. For example, CMR permits discrimination of coarctation from pseudocoarctation using the presence of turbulence and VEC imaging to measure peak flow velocity. A number of approaches to quantitate the severity of coarctation have been proposed over the years [26,27] including one study that used the Bernoulli equation adjusted for various hemodynamic parameters that previous studies proved correlated with stenosis severity [28]. Serial evaluation of patch or anastomotic aneurysms following repair is also straightforward with CMR techniques (Figure 21.7). In fact, one study suggested that CMR combined with clinical evaluation was more cost-effective than a combined clinical evaluations that used either echo or chest radiography to monitor for late complications after repair [29]. Treatment planning (surgical or interventional device) is greatly facilitated by CMR’s delineation of the location(s) of stenosis and the suitability for one therapy over another. This is particularly true in the case of a patent ductus arteriosus (PDA), where decisions regarding open surgical vs. percutaneous catheter-based closures are greatly aided. The unique anatomy associated with the various types of truncus abnormalities lends itself well to CMR evaluation. Again, not only are the anatomic features clearly evaluated but the degree(s) of truncal stenosis or regurgitation (or both), magnitude of left-to-right shunt via the frequently-present sub-truncal VSD, and degree of

(PAPVR) vary considerably, and include the SVC, innominate vein, right atrium, and inferior vena cava (Figure 21.6). CMR produces highly detailed 2D and 3D images of the origins, courses, and insertions of the normal vascular anatomy and the anomalous venous structures. The 3D datasets permit the interpreter to “fly” through the image space and reconstruct the data in any image plane. This added flexibility (with less acute nephrotoxicity than CT contrast and no ionizing radiation) allows for more accurate diagnoses and greater utility than conventional imaging display, particularly for surgical planning. CMR is more accurate than transesophageal echo and invasive cardiac catheterization in diagnosing PAPVR and its associated abnormalities (including sinus venosus atrial septal defects (ASD)) and permits optimal display of the various anatomic features [23]. The core elements of the CMR exam include axial cine CMR sequences to identify major anatomic features, black-blood imaging for greater spatial resolution to identify points of communication and short axis cine imaging to assess for right and left heart volumes, mass, and function. VEC sequences are then acquired to quantify flow in the ascending aorta as compared to flow in the pulmonary artery to compute shunt fractions (Qp/Qs: normally left to right but can be right to left in Eisenmenger’s syndrome). Serial evaluation of RV size, function and shunt fraction can follow the impact of shunts (PAPVR) on the heart.
Figure 21.7 Maximum image projection (a) of an adult with an incidentally discovered severe aortic coarctation. Note the aortic segments proximal (*) and distal (**) to the coarctation, demonstrating post-stenotic dilatation in the distal segment. Also note tortuous, “corkscrew-like” collateral vessels throughout the image that have developed due to chronic recruitment of these vessels in the setting of severe aortic obstruction. Maximum image projection (b) of a recurrent aortic coarctation (*) in a middle-aged woman with prior resection of aortic coarctation and end-to-end anastomosis at 1 month of age. There is an aneurysmal outpouching (**) at the takeoff of the left subclavian artery. Three-dimensional reconstruction (c) can provide additional information in some cases.
collateralization are definitively categorized, both before and after repair (Figure 21.8). There are frequently additional congenital cardiac and other defects associated with truncus arteriosus. Recent work has elucidated some of the potential molecular and genetic abnormalities that are shared with conotruncal abnormalities in the DiGeorge and velocardiofacial syndromes where the 22q11 deletion is frequently present [30]. CMR may help evaluate the presence and extent of these abnormalities in order to non-invasively phenotype individuals who may one day be candidates for future novel therapies.

**Septal defects**

While a full description of the individual atrial and septal defects is beyond the scope of this text, the common CMR advantages over CT, transesophageal echocardiography (TEE), or cardiac catheterization include an improved depiction of the atrial or ventricular septal anatomy [23], accurate quantitation of flows and the degree of shunting (left to right, or right to left), and evaluation of the suitability for percutaneous repair of the defects (Figure 21.9). Assessment of potential post-repair complications is also facilitated by examining for turbulent flow, or measuring residual shunt flows. Holmvang et al. used an en face orientation and VEC methods to prove superiority of this technique over spin echo sequences, which consistently overestimated ASD size [31]. This approach also accounts for multiple septal fenestrations, since flow is measured across the entire atrial septum, and not just the visualized defect(s). In a similar manner, CMR can measure the Qp/Qs in patients with multiple ventricular septal defects, and provide a more accurate assessment of the shunt magnitude. This is especially true in patients with complex congenital disease such as TGA with double-inlet morphology, where systemic blood flow must traverse...
the defect to exit the heart [32]. The high-velocity jet of small VSDs increases their visibility, since the jet will exhibit a dephasing of spins due to turbulent flow, and appear as a dark jet on bright-blood cine sequences. These techniques are very reproducible and are useful in serial evaluation of patients to evaluate for ventricular dilatation or dysfunction.

Conclusions
CMR is a vitally important tool for the assessment of adult congenital heart diseases and the quantitative capabilities of CMR for measurement of cardiac output, shunt fraction, regurgitant fraction, and other hemodynamic variables make it an invaluable resource. CMR is ideal for depicting the often complex anatomy of adults with repaired congenital heart disease who often have an incomplete knowledge of their previous surgery (or surgeries). The nuances of many of these repairs requires a detailed knowledge of the surgical techniques, the era in which the patient was repaired, and the potential complications of those procedures for an optimal study interpretation. For educational, training, and patient care purposes, it is often beneficial for imagers to partner with the treating physician on these cases to ensure that all questions are answered, and that all surgical possibilities are considered. New techniques, such as high-field scanners and more rapid parallel imaging methods will shorten imaging time and promise to provide greater accuracy in quantitative methods such as calculating flow and myocardial perfusion. Percutaneous placement of pulmonic valves may one day be routinely performed in CMR scanners, as recent work has foreshadowed [33]. As these patients continue to survive beyond their fifth decade, new complications may emerge that require the advanced non-invasive imaging capabilities that CMR provides. Based on current functionality, it appears certain that the evolution of CMR imaging techniques will continue to meet the needs of ACHD patients and their physicians.

Acknowledgments
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Cardiovascular computed tomographic angiography: complementary role to magnetic resonance imaging

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Introduction
Multidetector-row computed tomographic (MDCT) angiography has revolutionized diagnostic imaging of cardiovascular disease. The major limitation of this technology is that it utilizes radiation and requires iodinated, potentially nephrotoxic contrast medium. Initially applied for peripheral vascular disease [1] since the introduction of advanced generation of 64-channel MDCT scanners, MDCT angiography (CTA) has been accepted as a valuable tool for evaluating acquired adult cardiac disease [2]. Current recommended applications in adult cardiology include imaging coronary artery atherosclerosis, mapping pulmonary veins, and quantifying ventricular function.

Because of the radiation dose and the use of iodinated contrast, CTA plays a limited role in pediatrics and in congenital heart disease; if used selectively and applied with protocols that emphasize low radiation and contrast dose techniques, CTA can also play a role in evaluating pediatric cardiovascular disease (PCVD) and adult congenital heart disease (ACHD). While echocardiography and magnetic resonance imaging (MRI) and angiography (MRA) are the primary non-invasive diagnostic considerations for PCVD and ACHD, CTA should be considered as a complementary tool and a reliable alternative non-invasive modality when MRI or MRA cannot be used.

The purpose of this chapter is to review the role of CTA in clinical diagnostic algorithms for PCVD and ACHD. An overview clinical practice utilizing CTA is first presented, followed by discussions on CTA technique and clinical applications.

Clinical practice
Diagnostic evaluation of congenital heart disease (CHD) patients generally begins with transthoracic echocardiography. Despite its wide application and advantages, echocardiography has limitations for imaging patients with known or suspected CHD. Its success is dependent on both the operator and an adequate sonographic window. Peripheral intra-thoracic segments may not be well depicted. Coils, surgical clips, stents, and other metallic devices etc may degrade the acoustic windows along with scar or chest wall deformities. On the other end of the spectrum for CHD evaluation is catheter angiography with its invasive nature, inherent risks of iatrogenic cardiovascular injury high healthcare costs, radiation and can cause patient and family inconvenience.
When additional imaging is required beyond echocardiography, cardiac MRI (CMR) is considered first followed by CT as alternatives to catheter angiography for CHD. They are both non-invasive modalities with lower cost and greater patient satisfaction. MRI should be considered prior to CT, while CT should be utilized selectively.

As with echocardiography and catheter angiography, MRI can evaluate morphology, function, flow, and hemodynamics. CTA, while inherently dependent upon the use of radiation and iodinated contrast medium, can define function as well as morphology. With the current state of technology, CTA cannot be utilized to directly evaluate flow and hemodynamics. Despite these disadvantages, CT has several advantages. These include flexible availability, short exam times, high patient tolerance, high spatial resolution, and high contrast detail. In addition, the complete airway and lungs can be evaluated at the time of CTA. The rapid scan times afford the opportunity to scan multiple cardiovascular territories with a single bolus of contrast in one simultaneous acquisition, while maintaining high spatial and contrast detail.

These advantages can be exploited to utilize CTA selectively in the PCVD and ACHD populations and enhance the diagnostic workflow by offering an alternative to both catheter angiography and MRI. CTA should be performed in preference to MRI when MRI is contraindicated (e.g. non-MRI compatible coils in place – Figure 22.1), is unavailable (Figure 22.2) or if MRI has failed to delineate the structures needed to be visualized (e.g. the lumen of a vessel with a stent in place – Figure 22.1). CTA can be performed in a very short period of time (Figure 22.3). In addition, if high spatial resolution requirements are needed (Figure 22.4) that exceed those available by MRI (e.g. coronary stenosis) or if detailed pulmonary imaging is needed, CTA should be performed (Figure 22.5). MRI and CT can be utilized to evaluate non-cardiovascular anatomy during a cardiovascular MRI or CT. On one hand, CT may provide superior detail – such as in a higher resolution.

Figure 22.1 Cardiac MRI was attempted in a patient who had undergone right ventricular outflow tract and main pulmonary artery graft augmentation followed by left pulmonary artery (LPA) stent placement. (a) shows extensive ferromagnetic artifact, precluding a diagnostic cardiac MRI. A retrospective ECG-gated cardiac CT angiogram was subsequently performed. (b) is an oblique short axis reconstruction from the CTA. Note the calcified augmented graft segment (long arrow) and the LPA stent (short arrow). Most stents do not cause this type of MRI artifact and can be imaged safely.
PART III Special topics in cardiac MR of pediatric and congenital heart disease

The integration of sound preparation and patient specific acquisition and contrast medium protocols. To minimize radiation exposure and contrast medium dose and to avoid repeated acquisitions, fundamental knowledge of strategies to optimize technique is paramount. Equally important is the ability to adeptly use advanced visualization techniques for image display and interpretation.

**Preparation**

Patient preparation begins with a thorough understanding of the patient’s cardiovascular history. A detailed imaging request form and routine discussions between the CHD imaging and clinical teams is recommended as the patient’s history will help decide on the intravenous catheter placement, the acquisition protocol, and the contrast injection protocol. Included in this decision process is whether ECG-gating is required for either (1) assessment of cardiac function; (2) evaluation of the coronary arteries; or (3) optimization of struc-

**CTA technique**

Clinical success of CHD CTA is dependent on technical and interpretative success. Synchronizing the cardiovascular acquisition volume and enhancement with a single bolus of contrast requires the integration of sound preparation and patient specific acquisition and contrast medium protocols. To minimize radiation exposure and contrast medium dose and to avoid repeated acquisitions, fundamental knowledge of strategies to optimize technique is paramount. Equally important is the ability to adeptly use advanced visualization techniques for image display and interpretation.

**Preparation**

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cavo pulmonary shunts, to achieve homogeneous opacification using a split-bolus technique, it is necessary to access a foot vein in addition to an upper extremity vein [3].

Following these initial measures, patients are positioned supine and feet first on the scanner. Blankets and pillows should be used, as needed to position and support the patient. To optimize image quality, arms should be raised above the patient’s head. If ECG-gating is required, leads are placed in appropriate position and secured with tape. Excess wires should be removed out of the field of view.

**Acquisition**

**Protocol series**

A CHD CTA protocol includes up to five acquisition series. The first series is a required low dose anterior–posterior scout topogram through the entire region of interest. For precise coverage and field of view, a lateral view may be required. The second series is an optional non-enhanced acquisition (1.5–3.0 mm thick images). Coverage may be identical to the planned angiographic acquisition or may target a selected region. The objective of the non-contrast acquisition is to identify high density material, which may degrade image quality and interpretation or may itself be obscured by the contrast medium. Calcifications, endovascular stents and stent-grafts, surgical clips, surgical grafts, catheters, active bleeding, and hematomas should be addressed. The third series is a low dose timing acquisition, which is essential for precise synchronization of the image acquisition with either arterial or venous enhancement. Scan timing is determined either with a timing bolus acquisition or bolus tracking software. The fourth series consists of the contrast enhanced CT angiogram, discussed in further detail in the next section. The fifth series is an optional delayed post-contrast acquisition. The delayed acquisition is useful if the CT angiogram did not have adequate cardiac and vascular opacification. The delayed phase can be prescribed routinely to assess systemic and pulmonary veins, vasculitis, vascular masses, and hemorrhage. When cardiac function, the coronary arteries, or both require assessment in addition to the intra-thoracic vasculature, the ECG-gated CT angiogram can be obtained through the heart, and

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**Figure 22.3** A four-month-old male with tetralogy of Fallot underwent right ventricular outflow tract graft augmentation. Surveillance echocardiography on post-operative day two identified a possible dissection flap in the distal thoracic aorta. High resolution CT angiography was performed emergently. A volume rendered oblique projection through the aorta demonstrates a spiraling dissection flap extending from the distal ascending aorta to the celiac axis (large arrows). Note the partially thrombosed false lumen (small arrows).
tion, the required temporal resolution, the scan distance, and the scan duration.

A key technical option for CHD CTA is ECG synchronization. ECG-gating is applied to suppress cardiac motion and evaluate cardiac function. Two methods are available. The first method, prospective ECG-gating, is a non-helical “step-and-shoot” technique. Images are acquired at a pre-determined phase of the cardiac cycle. Traditionally this technique has been utilized for evaluation of coronary calcium. However, it now a recommended option for low dose angiographic cardiac imaging [4]. The second method, retrospective ECG-gating, is a low pitch, highly overlapping, volumetric acquisition which generates multiple cardiac phases per each Z-axis position. Raw data is reconstructed as a function of both the table position and user-defined cardiac phases. To optimize temporal resolution, the fastest gantry rotation speed is used. Temporal resolution is further enhanced by using multisector reconstructions.

As retrospective ECG-gating is dependent upon a low pitch and overlapping anatomical coverage, it inherently yields more patient radiation exposure than prospective ECG-gating. Despite the increased exposure, advantages include: (1) time resolved four-dimensional (4D) CINE visualization of the heart, valves, and thoracic aorta; (2) qualitative and quantitative evaluation of cardiac chamber, valve, and aortic function; and (3) multiphase morphology assessment. Temporal resolution is much less than CMR.

With routine single source 64-channel MDCT scanners, in addition to limiting exam coverage, strategies for reducing radiation exposure include employing automated tube current modulation

Table 22.1 MDCT technical parameters.

<table>
<thead>
<tr>
<th>Data acquisition</th>
<th>Reconstruction</th>
</tr>
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<tbody>
<tr>
<td>Detector row width</td>
<td>Slice thickness</td>
</tr>
<tr>
<td>Pitch</td>
<td>Reconstruction interval</td>
</tr>
<tr>
<td>Gantry rotation speed</td>
<td>Field of view</td>
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<tr>
<td>Tube current</td>
<td>Reconstruction algorithm</td>
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<tr>
<td>Tube voltage</td>
<td>Half Scan reconstruction</td>
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<tr>
<td>Prospective ECG-gating</td>
<td>Multisector ECG-retrospective gating</td>
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<td>Retrospective ECG-gating</td>
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Figure 22.4 A 14-year-old with exertional chest pain underwent retrospective ECG-gated coronary angiography (CTA). A left anterior oblique multiplanar reconstruction from the CTA demonstrates an anomalous right coronary artery (RCA, long arrow) arising from the left coronary cusp (short arrows). Note the acute ostial angulation with an approximate 1–2 mm intramural course and high grade stenosis involving the retro-pulmonic RCA segment (short arrows).

CT angiogram

For complete imaging of the cardiac chambers, systemic thoracic veins, pulmonary vasculature, thoracic aorta, and proximal supra-aortic branch segments, coverage should include the entire chest. To minimize radiation exposure, however, the CTA scan length should always be applied with the minimum extent necessary for anatomical coverage. Appropriate exam coverage may be achieved on one acquisition (non-ECG-gated) or two acquisitions (cardiac ECG-gated angiogram with an immediate delayed upper thoracic non-ECG-gated angiogram). If assessment of upper abdominal viscera is required, coverage should be extended caudally.

Table 22.1 lists technical parameters for CT angiograms performed on currently available MDCT scanners. These scan options are applied to generate high and isotropic resolution datasets and reflect a balance between the desired spatial resolu-
Cardiovascular computed tomographic angiography

rather than retrospective ECG-gating [4]. Dual source MDCT scanners and volumetric 256- and 320-channel MDCT scanners offer unique means to reduce radiation by using flexible pitch values [7] and potentially eliminating dependence on pitch, respectively.

Contrast medium administration
Diagnostic quality CT angiograms for CHD require contrast medium administration. Software [5], using the lowest possible voltage and reference amperage, and using ECG-pulsing – if the study is acquired with retrospective ECG-gating [6]. Depending on the patient’s weight and whether ECG-gating is required, tube voltage may range between 80 and 120 kV. Reference amperage may range between 20 and 500 mAs (automated tube current modulation). If multiphase 4D imaging is not required, when ECG-gating is necessary, prospective ECG-gating is recommended rather than retrospective ECG-gating [4].

Findings are consistent with tracheobronchomalacia. Note the associated advanced cystic changes in the right lung and atelectasis in the left lung. As shown in (b) and (c), main pulmonary artery (PA), right ventricle (RV), and right atrium (RA) are enlarged and there is flattening of the interventricular septum and posterior displacement of the left ventricle (LV), consistent with pulmonary hypertension.

Figure 22.5 High resolution CT angiography was performed in a 7-month-old ex-25-week premature boy who now has bronchopulmonary dysplasia and pulmonary hypertension. The exam was obtained to evaluate the airway, lung parenchyma, and cardiopulmonary morphology. (a) Coronal minimum intensity projection image demonstrating moderate narrowing of the central left main bronchus (arrow) with diffuse irregularity of the mid and distal central airways.

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Achieving diagnostic intracardiac and intravascular enhancement is dependent on synchronizing the delivery of contrast with the scan acquisition and delivering an appropriate amount (iodine dose) of iodine at an appropriate rate (iodine flux). CHD CTA injection protocols can achieve iodine requirements by adjusting the contrast medium concentration (300–370 mg iodine per milliliter), injection rate (0.7–6 ml/second), injection volume, injection duration, or a combination of these parameters based upon a patient’s body weight, the scan distance, and the speed of the scanner [8]. When imaging Fontan shunts, the injection volume can be split into two doses. One portion is delivered prior to the CTA via a foot vein for antegrade IVC-Fontan shunt opacification and the other is delivered via an upper extremity vein for the CTA acquisition and opacification of the hemi-Fontan or Glenn shunt [3].

To account for the variable time for contrast medium to travel from the site of intravenous injection to the heart, it is necessary to determine the transit time using either a test-bolus injection or automatic bolus triggering. The reference level will vary depending on the known or suspected congenital heart lesion(s) and any surgical palliation shunts.

Immediately following contrast medium infusion, saline flush is administered via a dual-chamber injector. The saline injection improves contrast utilization and reduces perivenous streak artifacts.

Depending on the patient’s body weight, a small volume (10–50 milliliters) is injected at a rate equal to the contrast medium injection rate.

**Image display**

Interpretation and communication of CHD CTA requires efficient and effective image display. Two (2D) and three (3D) dimensional visualization techniques are fundamental and necessitate the use of an advanced workstation. Four principle visualization techniques may be utilized (Table 22.2): maximum intensity projection (MIP), volume rendering (VR), multiplanar reformations (MPR), and curved planar reformations (CPR). Each technique has advantages and disadvantages, which impact how they are integrated for interactive real time interpretation and for generation of protocol driven, static post-processed images. It is important to keep in mind that the quality of all post-

| Table 22.2 Visualization techniques. |
|-----------------|-----------------|-----------------|-----------------|
| **Display** | **Principle use** | **Advantages** | **Disadvantages** |
| MIP | 2D | • Angiographic overview | • “Slice” through dataset in transverse, coronal, sagittal, and oblique projections | • Vessel, bone, visceral overlap |
| | | • Depict small caliber vessels | • Limited stent lumen evaluation |
| | | • Depict poorly enhancing vessels | • Limited by heavy calcium |
| | | • Communicate findings | • Opacity-transfer function dependent |
| VR | 3D | • Angiographic overview | • “Slice” through dataset in transverse, coronal, sagittal, and oblique projections |
| | | • Structural overview | |
| | | • Spatial perception | |
| | | • Communicate findings | |
| MPR | 2D | • Vessel analysis | • “Slice” through dataset in transverse, coronal, sagittal, and oblique projections |
| | | • Structure | • Accurate display of stenoses, occlusions, calcification, stents |
| | | • Flow lumen | • Limited spatial perception |
| | | • Vessel wall | |
| CPR | 2D | • Vessel analysis | • Complete longitudinal vessel cross sectional display |
| | | • Flow lumen | • Accurate display of stenoses, occlusions, calcification, stents |
| | | • Vessel wall | | 2D = two dimensional; 3D = three dimensional. See text for other abbreviations. |
processed images is inherently dependent on the acquisition technique and the degree of contrast enhancement. In addition, with all techniques, flexible angiographic window and level settings are used. This includes a wide window setting for vascular calcification, high contrast attenuation, or both.

Source images should always be viewed to confirm cardiovascular findings shown on real-time and static post-processed images. They are also reviewed to verify image quality and assess non-cardiovascular anatomy (Figure 22.6). The ability to evaluate non-cardiovascular anatomy allows a CHD CTA to be used as a multi-organ system exam. Relevant non-cardiovascular structures within the scanned volume are assessed with the same bolus of contrast. Review of the axial source images is facilitated by generating 2.0–5 mm thick reconstructions. Alternatively coronal and sagittal reformations (1.5–2.0 mm thickness) can be used.

CTA applications

Cardiac CT applications and interpretation for CHD focus on up to three areas. These include cardiovascular morphology, cardiac function, and coronary arteries.

Structural morphology

Assessment of structural morphology is the essence of CHD MRI and CT. The key objectives include establishing the situs, recognizing abnormal and anomalous structures and connections, identifying patent and obstructive arteries and veins, classifying the type(s) of lesion(s), assessing post-operative morphology, and confirming post-endovascular stent patency. With CT, ECG-gating may not be necessary. If anatomy on CT can be assessed without ECG-gating, patient radiation exposure can be minimized.

With MRI and CT, both upper abdominal and intrathoracic structures are evaluated. The congenital septal defects, to complex, such as heterotaxia. To optimize interpretation accuracy and efficiency, a segmental anatomical approach is recommended. While the primary emphasis of this process is to establish abdominal and thoracic situs and cardiac chamber and conotruncal looping, it is also provides a practical checklist for characterizing intra-abdominal and intra-thoracic cardiovascular structures.

The process begins in the abdomen, evaluating the presence and location of the stomach, liver, spleen, and inferior vena cava. Next, attention focuses on the venous-atrial, atrioventricular, and ventriculoarterial connections as well as the position of the atrial appendages, the cardiac apex, aortic arch and the branching pattern of the tracheo-bronchial tree. The final step is a detailed evaluation of the intra-thoracic systemic veins, pericardium, right cardiac chambers, left cardiac chambers, pulmonary arteries, pulmonary veins, thoracic aorta, and supra-aortic branch arteries, assessing structural size, course, contour, luminal caliber, patency, and enhancement.

MRI/MRA and CTA high resolution datasets can equally be viewed, manipulated, and interrogated using segmental analysis on Picture Archive and Communication System (PACS), an advanced 3D workstation, or both. To define concordant and discordant cardiovascular relationships and identify hypoplastic, atretic, stenotic, dilated, and anomalous anatomical segments, it is recommended to use an interactive multiplanar approach that applies multiple visualization techniques. For reviewing on PACS, post-processed static and rotating batch coronal, sagittal and oblique images can be generated on an independent workstation, These images are subsequently transferred to PACS. For reviewing on an advanced 3D workstation, datasets are manipulated dynamically using a combination of the volume rendered, maximum intensity projection, multiplanar reformations, and rotating curved planar reformations techniques with flexible opacity transfer functions, window and level settings, and slab thickness.

Whether on PACS or a 3D workstation, by nature of the CTA examination, as there is one volumetric series, the analysis is simplified and the time per each case is reduced. Optional pre-contrast and delayed acquisitions provide targeted information and add only minimal additional time.

CTA can offer a number of advantages in interpreting structural morphology, some of which have already been briefly discussed. The first regards characterization of airway and lung parenchyma.
At present, CT is considered the standard. CTA offers complete, high resolution airway and lung evaluation in addition to cardiovascular evaluation. Tracheal morphology; eparterial and hyparterial bronchial morphology; lobar anatomy; central and peripheral airway thickening, narrowing, and debris; and acute and chronic parenchymal changes can all be assessed. The second regards ferromagnetic materials, which on MRI, can lead to signal and flow void artifacts. Although artifacts from
CHAPTER 22 Cardiovascular computed tomographic angiography

Cardiovascular computed tomographic angiography requires precise synchronization of contrast delivery and image acquisition. If structures are not adequately opacified, such as cavopulmonary shunts, an additional acquisition may be necessary to distinguish technical artifact from occlusive disease. The penalty for this is greater patient radiation exposure. MR angiograms are more flexible as multiple phases can be obtained at no expense to the patient. The third important pitfall is that with current technology, CT angiograms do not directly reveal dynamic flow. However, flow dynamics can indirectly be inferred by observing the enhancement of lung parenchyma and cardiovascular structures, analyzing the size of native vessels, and recognizing collateral vessels.

Cardiac function

Echocardiography is accurate in the assessment of left ventricular size and shortening in patients with CHD. However, in assessing right or single ventricular function, the geometric assumptions used to calculate ventricular function can potentially lead to diminished diagnostic performance [9]. Advances in cardiac MRI have yielded accurate and reproducible methods to assess ventricular volumes.
and function in both left, right, and single ventricular chambers [10,11] and is considered the gold standard. Contiguous short-axis cine images provide three-dimensional measurements without geometric assumptions. This is important as late deterioration of right ventricular function is an important determinant of clinical outcome in various forms of congenital heart disease [12,13]. With its high temporal resolution, cardiac MRI currently remains the diagnostic modality of choice to quantify ventricular volumes and function. Every attempt should be made to determine accurate ventricular volumes and function at the time of the cardiac MRI.

MRI performance for evaluating cardiac function is degraded in the setting of ferromagnetic material. Coils, stents, and surgical clips may result in varying degrees of susceptibility artifact, limiting the ability of MRI to accurately assess ventricular volumes, mass, and ventricular function. The ability of cardiac MRI to accurately evaluate ventricular volumes and function in the Fontan population has been evaluated in the presence of ferromagnetic implants [14]. When compared to patients without implants, cardiac MRI was less accurate in its overall ability to assess ventricular volumes and function. This highlights the importance of selecting devices that produce negligible artifacts on cardiac MRI (nitinol or platinum rather than stainless steel) [15].

Cardiac CT offers an alternative method to assess ventricular volumes and function in CHD patients with limited temporal resolution. This requires that the CT is acquired volumetrically with retrospective ECG-gating. Accurate qualitative and quantitative datasets are dependent on a low heart rate to optimize temporal resolution but it should be understood that the lower temporal resolution of cardiac brings with it inherent inaccuracies of timing end-systole and therefore, stroke volume and ejection fraction. The dependence on a low heart rate is a greater issue with single source 64-channel MDCT scanners rather than with dual source 64-channel MDCT scanners or scanners with 256 or 320 detectors.

As with cardiac MRI, the end-diastolic and end-systolic phases in a cardiac CT dataset are processed in short axis using manual, semi-automated, or automated workstation algorithms. A study has shown agreement between cardiac CT measurements for both left and right ventricular size and function compared to echocardiography as well as cardiac MRI [16] (Figure 22.8).

In addition to cardiac motion, CT quantification of ventricular volumes and function may be degraded by suboptimal enhancement, noise, respiratory motion, and streak artifact. Contrast medium injection, saline flush, and scan acquisition parameters should be optimized to ensure the highest visualization of endoluminal and epicardial borders in both the left and right ventricles.

**Coronary arteries**

Assessment of the coronary arteries is an important component of CHD evaluations, whether as part of a general CHD assessment or a targeted investigation for a suspected or known coronary anomaly. This applies equally to initial, diagnostic investigations, as well as surveillance investigations, prior to and following endovascular and surgical interventions.

Compared to the general population, the incidence of anomalous coronary arteries is greater in patients with CHD (3–36%) [17,18]. Knowledge of coronary anatomy and the existence of an anomaly is paramount at the time of surgical palliation to avoid iatrogenic injury and also plan and perform any required corrective surgical intervention. Diagnostic imaging should assess the coronary origins and course, define coronary dominance, confirm patency, and exclude fistulas and aneurysms. Verifying patency is particularly important following coronary re-implantation (Figure 22.9). In adult CHD patients as well as pediatric CHD patients with metabolic cholesterol disorders, coronary artery atherosclerotic disease should also be characterized.

The standard methodologies to assess the coronary arteries in the CHD population include selective coronary catheter angiography and aortic root angiography. Although catheter angiography is highly sensitive for detection of coronary artery anomalies, angiography is a two-dimensional projection invasive technique. To completely characterize the three-dimensional spatial anatomy of the coronary tree, multiple injections from multiple projections are required. Another limitation of catheter angiography is that while it assesses the
CHAPTER 22 Cardiovascular computed tomographic angiography

Coronary MRA (CMRA) and CTA techniques provide methods to accurately assess the coronary arteries in the CHD population. Both techniques overcome the limitations of catheter angiography. They are three-dimensional techniques which afford evaluation of coronary and non-coronary structures and offer the ability to assess the coronary lumen and wall.

In neonates, infants, and young adolescents, echocardiography is the first-line alternative modality to non-invasively screen the coronary arteries. The sonographic window is often suitable to reliably image the origins and most proximal segments although false negatives have been known to occur when imaging coronary artery origins by echocardiography. When advanced coronary imaging is required, particularly for older adolescents and adults, MRA or CTA should be considered.

Coronary MRA (CMRA) and CTA techniques provide methods to accurately assess the coronary arteries in the CHD population. Both techniques overcome the limitations of catheter angiography. They are three-dimensional techniques which afford evaluation of coronary and non-coronary structures and offer the ability to assess the coronary lumen and wall.

CMR has been shown to be accurate in defining the origin and proximal course of the coronary arteries in a small cohort of adults with CHD when compared to conventional X-ray angiography [19]. Using a respiratory gated or navigator echo technique, the origin and the proximal course of the coronary arteries are well visualized. CMR is

Figure 22.8 Short axis reconstructions of axial multiphase reformatted cardiac computed tomography images obtained at end-diastole (a) and end-systole (b) with superimposed contours do not differ significantly from cine images in end-diastole (c) and end-systole (d) obtained from a cardiac magnetic resonance imaging examination in a 26-year-old with d-transposition of the great arteries who underwent a Mustard repair.
the preferred modality for imaging anomalous coronaries because of radiation considerations as stated in a 2008 American Heart Association document [20].

CTA coronary evaluation falls into two categories. The first is an evaluation during routine chest CT angiograms. If the heart rate is low enough (<70 bpm) and there is sufficient temporal and spatial resolution, proximal segments can be defined (Figure 22.10). The second is coronary assessment when using ECG-gating. Like CMR, ECG-gated CTA defines coronary origins and proximal segments well. ECG-gated CTA can display the entire coronary luminal tree with high spatial resolution, including the distal right, left anterior descending, and left circumflex coronary artery segments, as well as acute marginal, diagonal, and acute marginal arteries. This achieved in a short acquisition, which improves patient tolerance and minimizes potential artifacts. The great spatial resolution afforded by CTA provides superior routine assessment of the coronary arterial wall to characterize atherosclerotic plaque, inflammatory changes, aneurysm morphology, and coronary remodeling. In addition intramural and intramyocardial segments are defined with great definition.

Regarding clinical applications of coronary CTA, evaluation of coronary anomalies has been shown to be reliable and accurate [21] although CMR is preferred [20]. Multiplanar reformations, volume rendered three dimensional projections, and maximum intensity projections can distinguish a malignant (retro-pulmonic; Figure 22.4) from non-malignant (non-retro-pulmonic) coronary origin and course. Multiplanar and curved planar reformations are useful to define intramural segments, ostial narrowing and kinking, post-ostial stenosis, and myocardial bridging. Endoluminal

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**Figure 22.9** A volume rendered three-dimensional reconstruction depicts the right coronary artery (RCA), including the proximal portion that had been reimplanted in a patient who had undergone a Ross-Konno procedure for severe aortic valve insufficiency.

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**Figure 22.10** An oblique axial view (a) demonstrates a moderate size aneurysm of the proximal right coronary artery, measuring 11 mm at its largest diameter. The aneurysm has a moderate burden of thrombus (arrow). Of note, the aneurysm is also calcified (arrowhead) as demonstrated in the oblique sagittal view (b).
views can be applied to further assess ostial relationships.

CTA has also been shown to be a useful modality to assess coronary patency and negative remodeling following coronary re-implantation (Figure 22.9). Initial experience with early generation multislice computed tomographic angiography demonstrates the ability to diagnose intimal hyperplasia and coronary lesions [22].

Finally, CTA can be quite useful for the long-term follow-up of patients with Kawasaki disease and other acquired diseases. CTA provides an accurate assessment of aneurysm location, size, thrombus, and calcifications in proximal and distal segments, offering insight into coronary artery remodeling in Kawasaki disease (Figure 22.10).

**Conclusion**

Selective use of computed tomographic angiography can greatly enhance pediatric cardiovascular and adult CHD diagnostic evaluations. When MRI is contraindicated, CTA should be considered prior to catheter angiography. When applied in this capacity, CTA can serve as a useful adjunct to MRI, obviate the need for invasive angiography, and minimize patient risk. CTA protocols should always strive for the lowest possible radiation (ALARA concept) and contrast medium doses to obtain diagnostic image quality of both cardiovascular and non-cardiovascular structures. To achieve these goals, comprehensive, fundamental understanding of CT acquisition, contrast medium delivery, image display techniques is essential.

**References**


CHAPTER 23

Radiation in cardiac imaging in congenital heart disease

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Introduction

When the clinician attempts to answer a clinical question via imaging, there are a number of techniques in use in congenital heart disease (CHD) for him or her to choose from. Some have overlapping capabilities (e.g. visualizing valve morphology by echocardiography and cardiac magnetic resonance [CMR]) while others have unique applications (myocardial or blood tagging by CMR); each with their own advantages and disadvantages. Obviously, one consideration is picking the imaging modality that will best answer the specific question in the particular patient, but another is both the short and long-term risk of obtaining the information and conversely, of not obtaining the data.

One major concern is the use of ionizing radiation in cardiac imaging which is especially important to be aware of, since the major morbidity and mortality from this exposure, neoplastic disease, is usually manifested years after the test and long after the clinician ordering the test may have retired from medical practice. The extended time to manifest this side effect of the intervention, despite its dire consequences, is the major reason why it is so difficult for clinicians and patients to pay attention to it. The long-term cost to the patient and the healthcare system is enormous, yet the immediacy of medical concern at the time of ordering the test (e.g. visualizing anomalous coronary origins in a teenager with chest pain because of a suggestion of this on echocardiography) may blind clinicians to the risk and the alternatives to the test (e.g. ordering a CMR to visualize an anomalous coronary rather than cardiac computed tomography [CT]). It is analogous to the present situation in the United States economy, where long-term borrowing for the immediacy of the economic downturn will impact the ability of future generation that will need to repay these loans. In addition, because of “relative” ease in which a CT scan can be ordered, which has various reasons (e.g. relatively simple to learn and use, its explosive proliferation – see below), the “immediate gratification” for physician and patient can be too tempting.

A complete treatise on radiation in cardiac imaging, as with a number of chapters in this textbook, can in and of itself take up volumes. Any textbook on CMR, which competes as an imaging modality not only with echocardiography but with angiography, CT and nuclear scans, should delineate the risks of radiation exposure, especially in a textbook on imaging in CHD where a substantial portion of patients are children and will have multiple exams (see below). The purpose of this chapter is to give the reader at least a cursory glance at the field and to be somewhat knowledgeable regarding the issues involved. This chapter is not at all intended to deter the physician or patient from obtaining imaging utilizing ionizing radiation when it is absolutely necessary; it is, however, intended to raise the level of awareness that potential alternatives which do not expose the patient to ionizing radiation exist, that alternatives should be attempted first, and that the long-term
Table 23.1 Summary of types of measurement of radiation.

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Description</th>
<th>Unit – new (symbol)</th>
<th>Units – old (symbol)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absorbed dose</td>
<td>Energy deposited in a volume of tissue divided by the mass of the tissue</td>
<td>Gray (Gy) = 1 joule/kilogram</td>
<td>Rad 100 rads = 1 Gy</td>
</tr>
<tr>
<td>Equivalent dose</td>
<td>Takes type of radiation delivered into consideration. “Radiation weighting factor” is utilized. Radiation weighting factor = 1 for X-rays in CT</td>
<td>Sievers (Sv) = absorbed dose (in Gy) averaged over an entire organ and multiplied by the radiation weighting factor</td>
<td>Rem 100 rem = 1 Sv</td>
</tr>
<tr>
<td>Effective dose</td>
<td>Takes organ receiving radiation into consideration. “Tissue-specific weighting factor” is utilized</td>
<td>Sievers (Sv) = the products of equivalent doses and tissue weighting factors are then summed over all the irradiated organs to calculate the “effective dose”</td>
<td>—</td>
</tr>
<tr>
<td>Dose taking into account age of patient</td>
<td>No measurement unit. Need to assess in graphical format (Figure 23.1)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

consequences of ionizing radiation exposure need to be taken into consideration.

**How radiation is measured**

As easy as it might sound to measure radiation, it is actually not so simple and numerous terminologies are used. Here are the basics (summarized in Table 23.1):

**Absorbed dose** – The fundamental quantity for describing the radiation effect in humans is the absorbed dose which is the energy deposited in a volume of tissue divided by the mass of the tissue. Absorbed dose is measured in joules/kilogram and a special unit called gray (Gy) has the quantity of 1 joule/kilogram (International System). An older unit which the reader may hear or read about is called the rad of which 100 rads = 1 Gy (i.e. 1 mGy = 0.1 rad.)

**Equivalent dose** – The biological effects of radiation are not only a function of the absorbed dose but are also dependent on the type of radiation delivering the energy (i.e., whether it is X-rays, gamma rays, electrons [beta rays], etc.). The reason for this is because different types of radiation interact with tissue differently. Because of this, a “radiation weighting factor” is utilized for the specific radiation type which is a dimensionless constant. Therefore, an “equivalent dose” is the absorbed dose (in Gy) averaged over an entire organ multiplied by the radiation weighting factor and is measured in sieverts (Sv). As with absorbed dose, there is an older system of measuring effective dose and the unit used in that system is called the rem; similarly 100 rem = 1 Sv (1 mSv = 0.1 rem). Specifically, for X-rays in CT, the radiation weighting factor = 1, meaning that for CT, the absorbed dose in Gy is equal to the equivalent dose in Sv.

**Effective dose** – Cancer induction by radiation is not only a function of the amount of radiation absorbed (absorbed dose) and the type of radiation depositing the energy (equivalent dose), it is also a function of the organ receiving the dose. So, for example, reproductive organs which have millions of dividing cells are more susceptible to cancer induction than bone or brain. To permit comparison of the risks when different organs are irradiated, the quantity “effective dose” is used which is calculated by determining the “equivalent dose” to each organ irradiated and then multiplying by a tissue-specific weighting factor for each organ or tissue type (note how this is similar to the radiation weighting factor or the equivalent dose – don’t
were first recognized shortly after the discovery of the X-ray by Roentgen in 1895. Marie Curie, the discoverer of radium, and her daughter are believed to have died of radiation-induced leukemia. Skin reactions were observed in many people working with early X-ray generators, and by 1902 the first radiation-induced cancer was reported in skin. Within a few years, a large number of such skin cancers had been observed. Even Thomas Edison, who experimented with radiation and whose assistant died in 1904 with skin carcinoma noted that "the X-rays had affected poisonously my assistant" [1]. He stopped experimenting with radiation shortly afterwards. The first report of leukemia in radiation workers appeared in 1911 and since then, it has been well established that radiation is carcinogenic. The most definitive and updated publication on radiation effects on humans is the BEIR VII report published by the National Academy of Sciences [2].

The reader will no doubt hear or read about a number of "risk models" which attempt to describe the level of radiation risk as a function of dose (important in determining the effect of low levels of radiation in medical practice—Figure 23.2). These were models examined by the BEIR VII scientists from the National Academy of Sciences [2]. The "linear, no threshold model" states that as the dose of radiation increases, the level of cancer risk increases linearly from no exposure to increasing doses of radiation. The "linear quadratic model" states that as the dose of radiation increases, the level of cancer risk increases linearly at low levels of radiation whereas at higher exposure, the risks increase quadratically. The "linear model with a threshold" states that at low levels of radiation, there is no radiation risk until a threshold exposure is reached as radiation exposure is increased; afterwards, the risk increases linearly. The BEIR VII committee, after extensive review of data, accepted the "linear, no threshold model" for all cancer risks except leukemia where they accepted the "linear quadratic model" instead. This linear relationship at low-doses of radiation is borne out by epidemiologic studies [2,3]. They estimated that when exposed over a lifetime to 10 mSv above background, there was an increased

![Figure 23.1](image_url)
cancer risk of 1 per 1000 and 1 per 100 when exposed over the course of a lifetime to 0.1 Sv above background (100 mSv).

A good portion of what we know about exposure to radiation and the risk of cancer derives from survivors of the atomic bomb blasts in Japan in 1945 (The Life Span Study – LSS). There were 25,000 survivors who were followed over multiple decades for medical application purposes; the survivors who were exposed to low levels of radiation are of interest. In the group who received low-dose radiation of 5–150 mSv, a substantial increase in cancer risk was seen [4–7]. The data are too extensive to detail in this brief chapter and the reader is referred to the BEIR VII [2] report and the references (see below) for greater detail. The effect of radiation dose on the excess cancer risk of these patients are seen in Figure 23.3 with increasing risk noted for an increasing dose; note that by definition, an excess cancer risk must be zero if the radiation exposure is zero. Even more disturbing is the fact that cancer risks for patients today may even be higher than put forth by the “linear no-threshold” model as applied to this cohort; current radiation protection guidelines assign a radiation weighting factor of 1 to X-rays – the biological effectiveness per unit-absorbed dose of X-rays may be double the high-energy photons [2]. This would essentially underestimate the cancer risk in patients exposed to medical radiation. In addition to cancer, a dose–response relationship to mortality from non-neoplastic disease has been demonstrated in survivors of the bombings in Hiroshima and Nagasaki with statistically significant associations for the categories of heart disease; stroke; and diseases of the digestive, respiratory, and hematopoietic systems. However, these non-cancer risks at the low-doses were uncertain in the BEIR VII study and it was not modeled.

Besides the survivors of atomic bomb blasts, a number of epidemiological studies of cohorts support the notion that low-dose radiation exposure can induce cancer. A large scale study [8,9] of 400,000 radiation workers in the nuclear industry across multiple countries were exposed to an average radiation dose of ~20 mSv; a significant association was reported between radiation dose and mortality from cancer in this group of individuals. Interestingly, there was a significant increase in cancer risk in the group that received 5–150 mSv. Carr et al. studied 3719 patients with radiation to the abdomen for peptic ulcers between
1936 and 1965 who were followed over an average 25-year period and found a significant increase in stomach (relative risk $[RR] = 2.6$), pancreas ($RR = 2.73$) and lung cancer ($RR = 1.5$) in a dose-related manner compared to patients who were not irradiated [10]. There are a huge number of other studies from medical exposure to radiation, occupational exposure and environmental exposure, besides the atomic bomb survivors, and they are too long to list here or discuss. The reader is referred to the BEIR VII report which can also be accessed on-line [2].

The explosive use of CT in the past few years has made radiation a potential public health hazard. On the basis of CT use from 1991 to 1996, it is estimated that 0.4% of all cancers in the United States may be attributable to CT scanning [11,12]. At the rate of increasing CT use, this estimate may now be in the range of 1.5–2% [12]. See Figure 23.4 and text below.

**Radiation and cancer – childhood exposure**

As this textbook concentrates on CHD, childhood exposure to radiation is an especially important topic and the evidence is even clearer in this area. As a general rule, the National Cancer Institute website put it succinctly and clearly: “Major national and international organizations responsible for evaluating radiation risks agree there probably is no low-dose radiation ‘threshold’ for inducing cancers, i.e., no amount of radiation should be considered absolutely safe” [13]. Multiple studies have demonstrated that exposure to radiation in childhood is a significant risk factor for cancer development and there are several reasons, unique to children, which explain this:

- Children are considerably more sensitive to radiation than adults, as demonstrated in epidemiologic studies of exposed populations (see below).
• Children have a longer life expectancy than adults, resulting in a larger window of opportunity for expressing radiation damage.
• Children receive a higher dose than necessary when adult CT settings are used for children [13].

This is especially important in patients with CHD who generally undergo numerous cardiac catheterizations, nuclear scans and occasionally CT scans during their lifetime. The cumulative increased risk from multiple exposures to radiation-based techniques put these children at an unusually high risk.

Studies from all over the world come to the same conclusion about children, radiation and cancer. A study from Israel published in 2000 looked at 674 children who underwent cardiac catheterization between 1950 and 1970 with a mean follow-up of 28.6 years [14]. Skin dosages (not to be confused with the organ doses received which are generally lower) ranged from 5–40 cGy; they found an excess of malignancies relative to the general population with a standardized incidence of 2.3. There was an overabundance of lymphomas (standardized incidence of 6.3) and melanomas (standardized incidence of 4.9). In a population based case-control study from China, 642 childhood cancer cases who had post-natal X-ray exposure were investigated along with 642 controls; there was an excess of cancer cases in the radiation exposed group. The odds ratio of all cancers was 1.3 with acute leukemia (odds ratio 1.6), brain cancer (odds ratio 1.5) and lymphoma (odds ratio 1.3) being the highest groups. In another population based case-control study of postnatal diagnostic X-rays and cancer, this time performed in Canada, 491 acute lymphocytic leukemia cases less than 10 years of age and an equal number of controls were examined between 1980 and 1993 [15]. The cases were matched to their controls on multiple levels including prenatal radiation exposure and family educational level. Since radiation doses were not known, the number of X-rays served as a surrogate measure of the amount of radiation the children received. There was a strong radiation-exposure response, with leukemia risk increasing with the number of X-rays (mostly bone X-rays, very few CT scans); with one exposure, the odds ratio was 1.04, however, with two or more exposures, the odds ratio went up to 1.61. In females, this was even more striking with the odds ratio for one exposure of 1.41 and two or more being 6.66. In an analysis which pooled seven clinical studies in the literature from around the world on radiation exposure to the thyroid gland in children and thyroid cancer [16], a clear linear dose response was demonstrated for children who were exposed to radiation before age 15. For this group, a very steep dose-response was observed with radiation and thyroid cancer. Among the children who were <15 years old and exposed to radiation, there was a strong trend for the risks of getting cancer to increase as the age at radiation exposure decreased.

The previously mentioned studies are just a sampling of the investigations that link radiation to cancer. For the reasons mentioned previously, the younger the age of exposure, the more likely an individual will develop cancer from the exposure. This has been quantified by a number of scientific organizations such as National Academy of Sciences Biological Effects of Ionizing Radiations Committee [2] and International Commission on Radiological Protection [17]; the graphs are delineated by Brenner et al. in their 2001 publication [18] (Figure 23.1). In the pediatric age range, the increased risk per unit gray exposure is 8–14%, depending upon which model is used and as shown, it is higher the younger the exposure. Even young adults between 20 and 40 years of age have a 3.5–12% increased risk per unit gray exposure, again depending upon which model is used and the age of the patient. Besides younger age, females are more susceptible than males to this increased risk. They state that “The larger doses and increased lifetime radiation risks in children produce a sharp increase, relative to adults, in estimated risk from CT.”

As mentioned, not all risks are equal. Dovetailing with the previous investigation is another one performed by Einstein et al. [19] published in JAMA in 2007 which used Monte Carlo simulations, phantoms (computational models) and the BEIR VII approach to estimate lifetime attributable risk (LAR) of cancer to patients undergoing 64 slice CT scans. Age, sex and scan protocols were taken into consideration. LAR estimates for standard cardiac scans ranged from 1 in 143 for a 20-year-old woman to 1 in 3261 for an 80-year-old man.
combined cardiac and aortic scans.

**Radiation from background and various imaging modalities**

It is true that we all are exposed to background radiation while performing our everyday tasks. This comes from “the ground, building materials, air, food, the universe, and even elements in their own bodies” [2]. It is estimated that worldwide, the background radiation per year is approximately 2.4 mSv and is slightly over 3 mSv in the United States because of higher levels of radon [2]. It is important to understand that all estimates of excess cancer risk due to radiation exposure is above and beyond this background exposure.

As official statements from various international and national organizations such as the American Heart Association, the American College of Cardiology and the European Society of Cardiology are produced, they are recognizing that radiation plays an important role in determining the choice of imaging modality. For example, the 2008 statement from the American Heart Association on non-invasive coronary imaging states that “anomalous coronary artery evaluation can be performed by either CTA or MRA; radiation-protection concerns indicate that MRA is preferred when it is available (Class IIa, level of evidence B)” [20]. In a statement published in the *European Heart Journal* in 2004 [21], The European Society of Cardiology prefers CMR because “the lack of ionizing radiation is an important consideration when performing sequential studies in children and young adults.”

One must know approximately how much radiation each imaging modality generally exposes the patient to so as to understand the risk involved. The following data are quoted in mSv; however, in pediatrics, one should be cautious about this. Although mSv is designed to estimate biological risk and takes into account the amount of radiation deposited, the type of radiation and the organ susceptibility to the radiation, it does not take into account one specific risk factor – age (see Table 23.1). The risk factor for age must be obtained from the graphs of age of exposure versus risk per Gy or mSv in various publications [12,18] and is represented in Figure 23.1.

Approximate radiation doses for various imaging modalities are given in Table 23.2, in the two left columns. It is instructive to put these radiation doses in perspective. Since one of the most common radiation-based imaging modalities is a postero-anterior chest X-ray (chest PA), other imaging modalities are assigned a chest PA equivalent or how many chest PAs would one of these imaging tests be; this is located in the 2nd from the right column. In addition, since worldwide background radiation is a mean of 2.4 mSv per year, other imaging modalities are assigned a background radiation equivalent or how many days or years would one of these imaging tests be; this is located in the right-hand column. All values are referenced. As you can see relative to cardiac imaging, cardiac catheterization and cardiac CT are hundreds of chest PAs and years of exposure to background radiation. It would be prudent to avoid this if other imaging modalities, such as CMR, are available as alternatives, especially in younger patients.

**Another viewpoint**

There are those who would say that the issue of radiation exposure in medical imaging is overstated. A manuscript published in 2009 is instructive [22]; this article makes “excuses” as to why medical radiation can be acceptable and encompasses a number of viewpoints from this side of the radiation issue. In this article, the authors state that “there is no consensus as to whether the effects observed in Japanese individuals who experienced whole-body acute exposures to primarily high levels of radiation can be extrapolated to the partial-body exposures at much lower levels of radiation that are delivered to patients of different ethnic origins who are undergoing medical imaging procedures.” Indeed, BEIR VII from the National Academy of Sciences [2], the International Commission on Radiological Protection [17], the National Cancer Institute of the National Institutes
of Health [13] as well as the numerous studies of risk mentioned above do come to this consensus. Dovetailing with this, the article also questions the validity of the “linear no-threshold” model; although not stated in the article, it is not seen as valid by such societies as the American Nuclear Society [23], the Health Physics Society [24] and the France Academy of Sciences/National Academy of Medicine [25]. The other side of this question has a whole host of reputable and prestigious national and international organizations backing its validity and its use in medicine such as the National Academy of Sciences [2], the International Commission on Radiological Protection [26], the US National Council on Radiation Protection and Measurements [27], the United Nations Scientific Committee on the Effects of Atomic Radiation [28] and the UK National Radiological Protection Board [29] to name but a few. BEIR VII went to great pains to discuss why low doses of radiation may be “less harmful” and why they may be “more harmful” than the linear no-threshold model. As mentioned earlier in this chapter, even more disturbing is that cancer risks to patients exposed to radiation-based medical imaging modalities may even be higher than put forth by the generally accepted “linear no-threshold” model as applied to survivors of the atomic bomb survivors. Current radiation protection guidelines assign a radiation weighting factor of 1 to X-rays – the biological effectiveness per unit absorbed dose of X-rays may be double high-energy photons [2]. This would essentially underestimate the cancer risk. The article [22], which questions the cancer risk estimates, needed to quote from the “public summary” of BEIR VII by stating that at doses <100 mSv, it “is difficult to evaluate cancer risk in humans.” Difficult but not impossible, as the actual “body” of BEIR VII did make a good estimate of this risk.

The authors of this article also question Monte Carlo simulations which in a number of studies are used in estimating radiation dose. This technique involves using random numbers and probability to solve problems and has been validated and in use for more than 60 years. It has contributed

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Radiation in mSv [ref]</th>
<th>Chest PA equivalent</th>
<th>Background Rad equivalent (2.4mSv)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest PA</td>
<td>0.01 [12]</td>
<td>1</td>
<td>1.5 days</td>
</tr>
<tr>
<td>Chest lateral</td>
<td>0.03–0.06 [13]</td>
<td>3–6</td>
<td>4.5–9 days</td>
</tr>
<tr>
<td>Screening mammography</td>
<td>3 [12]</td>
<td>300</td>
<td>456 days (1.3 years)</td>
</tr>
<tr>
<td>Head CT</td>
<td>2–4 [34]</td>
<td>200–400</td>
<td>304 days (0.8 years)–608 days (1.7 years)</td>
</tr>
<tr>
<td>Diagnostic coronary angiogram</td>
<td>3–6 [34]</td>
<td>300–600</td>
<td>456 days (1.3 years)–912 days (2.6 years)</td>
</tr>
<tr>
<td>Pediatric diagnostic catheterization</td>
<td>4.6 [35]</td>
<td>460</td>
<td>699 days (1.9 years)</td>
</tr>
<tr>
<td>Pediatric therapeutic catheterization</td>
<td>6 [35]</td>
<td>600</td>
<td>913 days (2.5 years)</td>
</tr>
<tr>
<td>Chest CT</td>
<td>5–7 [34]</td>
<td>500–700</td>
<td>761 days (2.1 years)–1065 days (2.9 years)</td>
</tr>
<tr>
<td>Abdominal or pelvic CT</td>
<td>8–11 [34]</td>
<td>800–1100</td>
<td>1217 days (3.3 years)–1674 days (4.6 years)</td>
</tr>
<tr>
<td>Sestamibi myocardial perfusion</td>
<td>9 [22]</td>
<td>900</td>
<td>1368 days (3.9 years)</td>
</tr>
<tr>
<td>Barium enema</td>
<td>15 [12]</td>
<td>1500</td>
<td>2282 days (6.3 years)</td>
</tr>
<tr>
<td>ECG-gated cardiac CT</td>
<td>4.2–18.1 [36,37]</td>
<td>420–1801</td>
<td>639 days (1.8 years)–2754 days (7.5 years)</td>
</tr>
<tr>
<td>Thalium stress/rest</td>
<td>41 [22]</td>
<td>4100</td>
<td>6150 days (16.8 years)</td>
</tr>
</tbody>
</table>

CT: computed tomography; ECG: electrocardiography; mSv: milliseverts; PA: postero-anterior.
immensely to the physical sciences; found utility in business and telecommunications; is used in the biological sciences [30]; and has made things such as nuclear physics, regulating traffic flow and spaceflight possible. That’s a pretty good track record.

There are three other issues the alternative viewpoint may highlight. The first is a lack of discussion regarding alternative imaging modalities such as CMR which can, in most cases, substitute in CHD for radiation-based examinations, especially CT; this discussion was relegated to just a few words in the article mentioned above [22] in their recommendations section. They point out the risk of not obtaining the required diagnostic information which is indeed a very important factor; however, alternatives to radiation-based techniques to obtain that information should always be considered first. Another issue is a lack of discussion regarding age which this article [22] did not mention and as much of this chapter discusses, is an extremely important aspect to consider. Finally, it is recognized that CT scanning and other radiation-based techniques may have higher spatial resolution than techniques such as CMR; this may be true for many cases; however, the question of “added diagnostic value” must be considered. How much extra does the added spatial resolution give to solving the clinical problem? Added spatial resolution is certainly important if the issue is coronary stenosis where submillimeter resolution is important. However, to determine anomalous coronary origins, pulmonary artery size, coarctation of the aorta and spatial relationships such as in transposition of the great arteries for example, the spatial resolution of ~1 mm which CMR can yield is certainly more than adequate. There would be no reason to expose patients to ionizing radiation to get a slightly “prettier picture” if the diagnosis can be made without radiation risk.

Finally, the alternative viewpoint is of the opinion that the incremental increase in cancer risk is minimal compared to the background risk of fatal cancer, dying of a motor vehicle or pedestrian accident or of smoking [22]. The argument is that because of this minimal increase, radiation-based imaging procedures should be used with less caution than those sounding the alarm. It should be asked of those who think the cancer risk of radiation is minimal that if indeed they believe this to be true, they should remove the lead aprons placed over them when a dentist performs a dental X-ray or when a technician performs a chest X-ray. In addition, this side of the debate fails to recognize that the radiation risk of cancer is cumulative so that continued exposure with multiple exams over the course of years can substantially increase risks so that even “minimal” risks can become substantial over the course of a lifetime of exposure. This is especially important in children with CHD who will need multiple imaging examinations over the course of their lifetimes. A few CT scans and cardiac catheterizations can add up quickly to significantly increase the patient’s risk even more than just the one exam.

Two examples from the adult literature are instructive with regard to multiple radiation-based scans. A recent article by Griffey and Sodickson looked at 131 patients in an emergency room setting over the course of an average of 7.7 years [31]. The number of scans were a mean of 13 with a maximum of 70! Cumulative CT doses of 122 and 579 mSv respectively were calculated and lifetime attributable risk of cancer was one in 82 and one in 17 respectively. In another study, Sodickson et al. [32] reviewed the records of 31,462 patients who underwent diagnostic CT in 2007; they had also undergone 190,712 CT examinations over the prior 22 years. Thirty-three percent underwent ≥5 CT scans lifetime and 5% underwent 22–132 scans. Fifteen percent received an estimated cumulative doses of >100 mSv and 4% received 250–1375 mSv. The lifetime attributable risk was a mean of 0.3% and a maximum of 12% for cancer incidence and a mean of 0.3% and a maximum of 6.8% for cancer mortality. CT exposures were estimated to produce 0.7% of total expected baseline cancer incidence and 1% of total cancer mortality.

The rising epidemic

The use of CT in adults and children has increased tremendously in recent years (Figure 23.4). Since 1980, the number of scans has increased ~8-fold and annual growth rate is estimated to be ~10%/year [13]. In another report, it has increased by >20 times, with an estimated 62 million CT scans currently obtained each year in the United States as
compared with \( \sim 3 \text{ million in 1980} \) [12]. Although CT scans comprise \( \sim 12\% \) of diagnostic radiological procedures, they contribute \( \sim 45\% \) of the collective radiation dose from all medical radiological procedures in the US [13]. CT is the largest contributor to medical exposure to the US population and this has not escaped the pediatric population. Estimates of the proportion of CT studies that are currently performed in children range between 6% and 11% [12]. This is a looming epidemiologic disaster and will contribute to the overall cancer rate in upcoming years.

Conclusion and a rational approach

Radiation is a dangerous tool that has been shown by many well-known and respected researchers and organizations to cause cancer as well as other non-neoplastic side effects. Because cancer occurrence usually happens years after the exposure in medical practice, it is not at the forefront of the physician or the patient’s mind. Nevertheless, radiation-based imaging procedures can yield valuable diagnostic information, but the risk of cancer and other side effects should limit its use. As Benz and Benz [33] as well as the National Cancer Institute [13] have noted, “avoid unnecessary examinations.” When the information is truly needed, the healthcare provider is ethically obligated to “consider other imaging modalities” which do not use ionizing radiation such as CMR and echocardiography. For nearly all CHD and pediatric applications, CMR can substitute for CT scans unless there is a contraindication to CMR. Just making a “prettier picture” is not acceptable – the radiation-based imaging modality must demonstrate added diagnostic value to the patient’s care. Finally, if cardiac catheterization, CT or nuclear scans are thought to be the only way to obtain the necessary information, the ALARA concept (as low as reasonably achievable) in the radiation dosing should be used. The smallest amount of radiation exposure to obtain the necessary information is prudent. Always remember the words from the National Cancer Institute: “Major national and international organizations responsible for evaluating radiation risks agree there probably is no low-dose radiation ‘threshold’ for inducing cancers, i.e., no

amount of radiation should be considered absolutely safe” [13].

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CHAPTER 23 Radiation in cardiac imaging in congenital heart disease

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Pediatric cardiovascular MRI in the outpatient private practice setting

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Introduction

Patient procedures and testing that once were only done during an inpatient setting are now more commonly being done on an outpatient basis. The outpatient setting provides unique advantages over the inpatient setting. Studies can usually be scheduled and performed more quickly, convenience is enhanced usually because of easier access and parking; and patients are usually more comfortable having procedures done outside the hospital. Children in particular may benefit from testing in the outpatient setting. In particular magnetic resonance imaging (MRI) testing can be frightening at times, even for adult patients. Therefore providing maximal comfort in a safe environment may help achieve increased diagnostic accuracy.

However since children are not just little adults, different steps are required to ensure their safety during outpatient testing and procedures. Procedural steps, equipment, and medications need to be tailored to meet the needs of the pediatric patient. This includes having appropriately sized equipment, accurate pediatric medication doses, and staff trained to monitor and respond to pediatric emergencies.

MRI pre-procedure routine

The pediatric patient can present some unique challenges that need to be addressed prior the MRI procedure. Before the testing day, the parents are called and initial questions are answered and directions to the MRI center are verified. Communicating with parents prior to the day of testing helps allay pre-test anxiety for the family. On the day of MRI, the child and family are required to arrive at least one hour prior to the study. This allows the child and family to become comfortable with the surroundings. It also allows time for hearing a description of the procedure and for filling out paperwork, verifying insurance and for giving informed consent. Since the pediatric patients are unable to give consent for procedures, it is important not only to involve the patient but the parents and/or guardian in the procedural description. Body weight, height and baseline vital signs are also measured and recorded. Accurate weights are important because, unlike adults, pediatric medications are commonly weight-based. A weight and height allows for body surface area to be calculated as structures need to be indexed in the growing child.

Preparing children prior to an MRI procedure can reduce anxiety and fears that are experienced on the day of the study. The first step is to explain the procedure in detail in terms that the child will understand. Parents should also be involved in this step. Making sure that both parents and children have an understanding of everything from proce-
Prior to the day of testing, detailed instructions need to be given to the family. The child needs to have had nothing to eat or drink (NPO) for several hours prior to arriving at the facility in order to reduce the risk of aspiration should an adverse reaction to medication or contrast occur. A good and conservative rule of thumb would be to follow the 1999 American Society of Anesthesiologists Fasting Guidelines [4]. These guidelines state that a patient should refrain from ingesting the following substances prior to general anesthesia: (1) clear liquids for the preceding 2–3 hours; (2) breast milk for the preceding 4 hours; (3) infant formula for the preceding 6 hours; (4) non-human milk for the preceding 6 hours; (5) a light meal for the preceding 8 hours, and (6) a heavy meal for the preceding 8 hours. These guidelines apply to general anesthesia but can be extrapolated to also apply to conscious sedation.

As mentioned previously, patients and parents/guardians are also instructed to arrive 1 hour prior to the scheduled procedure in order to prepare for testing. Intravenous therapy needs to be established prior to any administration of conscious sedation medications even if the route for the sedation medication is by mouth. Commonly used medications for minimal to moderate sedation in the outpatient setting are choral hydrate, midalozam, diphenhydramine, and fentanyl. (Table 24.1 lists sedation medications and dosages). It is useful in the outpatient setting to avoid longer-acting drugs such as pentobarbital, since long-term monitoring can be difficult.

Prior to sedation administration, a thorough patient history is obtained, baseline vital signs are taken, and an accurate weight in kilograms is established. At Oklahoma Heart Institute, a pediatric sedation score is used both before and after sedation. A similar scale is the Aldrete Recovery Scale [5] (Table 24.2). Emergency and monitoring equipment must be checked and in place prior to the administration of sedation. Since it is difficult to visualize the patient while they are undergoing MRI, it is important that equipment be functioning properly. Blood pressure, pulse oximetry, and EKG must be monitored continuously after sedation medications are given to assess for decline in the patient’s status. These should be recorded every 5 minutes until sedation

**Sedation**

Sometimes children must undergo MRI procedures that require them to be still for long periods of time. Since some children are unable to remain still, conscious sedation may be needed to complete the MRI study. Many steps must be taken prior to an outpatient MRI to prepare children, parents, and facility staff for sedation. Parents need to understand one of the main goals conscious sedation is to relieve anxiety, relieve pain, and control excessive movement during the MRI study [3].
score of 8 or greater must be obtained prior to patient discharge. When the child’s vital signs and mental status return to baseline the child may then be discharged to home with instructions. Parents/guardians are given instructions on what to expect with regards to their child’s activity and mental status. Oklahoma Heart Institute recommends with infants and toddlers that a parent ride in the back seat with them on the way home to make sure that airway issues do not arise while the child is in a car seat. Parents are also provided with contact information if they have any questions or concerns after the procedure.

**Emergency situations**

Outpatient MRI differs from the inpatient setting because there are fewer ancillary staff available for emergencies. However having fewer staff should not reduce the quality of a response should an emergency arise. Having well-trained physicians, nurses, and MRI technologists who are comfortable with pediatric emergencies is important. Routine practice of emergency situations should be implemented on a regular basis. All nursing and physician staff should have training and certification in Pediatric Advanced Life Support. This course is offered locally by the American Heart Association. The course teaches participants to handle many pediatric emergencies including how to deal with adverse events that may occur during conscious sedation.

Planning for emergencies is essential for the safety and well-being of the pediatric patient. Emergency equipment needs to be in place and in good working order prior to any pediatric MRI study. A pediatric code cart with appropriate medications and equipment needs to be readily available for all pediatric procedures. Common monitor equipment includes, but is not limited to, appropriate sized blood pressure cuffs, pulse oximetry probes, bag valve masks, endotracheal tubes, IV catheters, and EKG patches. Another helpful tool is a color coded resuscitation guide such as the Braslow’s measuring tape [6]. This tape provides accurate medication dosages quickly and with minimal calculations. Physicians and nurses also need to be familiar with the sedation reversal agents and they need to be readily availa-

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dosage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fentanyl</td>
<td>1 to 2 micrograms/kg IV</td>
</tr>
<tr>
<td>Midazolam</td>
<td>0.5 to 1 mg/kg IV</td>
</tr>
<tr>
<td>Chloral hydrate</td>
<td>50–60 mg/kg po</td>
</tr>
<tr>
<td>Diphenhydramine</td>
<td>0.5 mg/kg po</td>
</tr>
<tr>
<td>Pentobarbital</td>
<td>1 to 3 mg/kg IV</td>
</tr>
</tbody>
</table>

**Table 24.2 Aldrete recovery scale.**

<table>
<thead>
<tr>
<th>Activity</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Voluntary movement of all limbs to command</td>
<td>2 points</td>
</tr>
<tr>
<td>Voluntary movement of 2 extremities to command</td>
<td>1 point</td>
</tr>
<tr>
<td>Unable to move</td>
<td>0 points</td>
</tr>
<tr>
<td>Apneic</td>
<td>0 points</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Respirations</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breathe deeply and cough</td>
<td>2 points</td>
</tr>
<tr>
<td>Dyspnea, hypoventilation</td>
<td>1 point</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Circulation</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>BP +/- 20 mmHg of pre-anesthesia level</td>
<td>2 points</td>
</tr>
<tr>
<td>BP &gt; 20–50 mmHg of pre-anesthesia level</td>
<td>1 point</td>
</tr>
<tr>
<td>BP &gt; 50 mmHg of pre-anesthesia level</td>
<td>0 points</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Consciousness</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fully awake</td>
<td>2 points</td>
</tr>
<tr>
<td>Arousable</td>
<td>1 point</td>
</tr>
<tr>
<td>Unresponsive</td>
<td>0 points</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Color</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pink</td>
<td>2 points</td>
</tr>
<tr>
<td>Pale, blotchy</td>
<td>1 point</td>
</tr>
<tr>
<td>Cyanotic</td>
<td>0 points</td>
</tr>
</tbody>
</table>

Post sedation score must be >8 at conclusion of monitoring.

**Table 24.1 Oklahoma Heart Institute commonly used sedation medication for minimal to moderate sedation in the outpatient setting.**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dosage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fentanyl</td>
<td>1 to 2 micrograms/kg IV</td>
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<tr>
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</tr>
<tr>
<td>Diphenhydramine</td>
<td>0.5 mg/kg po</td>
</tr>
<tr>
<td>Pentobarbital</td>
<td>1 to 3 mg/kg IV</td>
</tr>
</tbody>
</table>
Other challenges

Other challenges may be evident prior to the procedure. Family dynamics play a significant role in a child’s life. When children feel overwhelmed from pain, anxiety, or distress they turn to their parents for support. Families of chronically ill children are faced with a multitude of challenges and problems on a daily basis. When incorporating the fact that they now have to take time from their schedules to come to an outpatient setting for a procedure, it can produce a wide variety of issues with family members. If relationships are strained due to family dynamics the child’s support system may not be available or sufficient. As children age their dependence on this support diminishes [7].

Another challenge that families face is the cost of healthcare. In 2005 about 45 million Americans were uninsured [8]. This leaves many children without access to essential healthcare. It also adds to the stress families feel on a daily basis. Even when families do have health insurance, their plans may only pay a small percentage of the total cost of the test. The plan may also require prior authorization for MRI testing. This may lead to a delay in testing thus adding to family stresses. Physicians ordering and/or performing the MR study may also need to call medical directors with additional justification for the testing also adding time and stress to the patient’s support system.

Conclusions

Caring for the pediatric patient can prove to be challenging if your outpatient center is not prepared. The pediatric patient is unique at each developmental stage. Children are not just little adults and care must be taken to ensure their safety in the outpatient setting by having appropriate sized equipment, appropriate doses of medications, and well trained staff. Frequent training in pediatric IV therapy, medication dosing, and pediatric emergencies will reduce stress with the patient, the family, and the staff. Encouraging family involvement by explaining the procedure to the child and parent will help reduce anxiety for all parties involved. Many challenges may arise but if you are well-prepared then having the procedures done in an outpatient setting can be beneficial to the family and healthcare system.

References

The role of the technologist in performing a cardiac MRI (CMR) exam

Christine Harris
Children's Hospital of Philadelphia, Philadelphia, PA, USA

Introduction

Magnetic resonance imaging has inherent advantages for evaluation of the heart and blood vessels. These include its non-invasive nature, excellent contrast between flowing blood and cardiovascular structures and the ability to produce images in any plane. Magnetic resonance imaging (MRI) has been a valuable tool in the evaluation of clinical diagnoses of brain, abdominal, and musculoskeletal disease. The application of cardiac MRI (CMR) in the last decade has become a clinical reality due to the advancements in image quality, and reduction in scan times, as well as the development of pulse sequences that generate high contrast, high resolution images. Cardiac MRI has become an essential non-invasive imaging modality for the assessment of cardiac function and vascular anatomy. Using complex, post-processing algorithms, the cardiologist, radiologist and or technologist can then construct a 3-dimensional model of the heart which can be viewed from all sides, and can separate out particular vessels for better visualization.

While CMR is becoming the standard of care for cardiac imaging at most major institutions, careful consideration should be given to understanding and performing these cardiac MRI exams on pediatric patients and the role of the technologist in ensuring that this exam is safe and diagnostic.

As an example of what a tertiary care center has, using the Children’s Hospital of Philadelphia (CHOP) as an example, we perform cardiac MR imaging (CMR) on Siemens 1.5T Avanto systems. Table 25.1 displays our three 1.5T systems, software levels, and number of channels for each system.

Example of a CMR program that works well

The team

It is important to have a team that is collaborative and cohesive for optimal outcome. We continually work to plan and implement policies and procedures that improve image quality, assure safety and help our program be efficient. With this mission in mind, we have formed a team that consists of cardiologists, radiologists, dedicated CMR technologists (with expertise in other areas of MRI as well), dedicated 3D lab technologists, cardiac anesthesiologists, cardiac schedulers and radiology nursing staff.

Cardiologist/radiologist

The cardiologist/radiologist is responsible for ensuring that the technical staff have performed the correct sequences in the proper views and post-
CHAPTER 25 The role of the technologist in performing a cardiac MRI (CMR) exam

The role of the technologist in performing a cardiac MRI (CMR) exam

Our radiology nursing staff understand not only patient care, but also MRI imaging. They assist technologists with setting up the MR environment and positioning the patient. They aid the cardiac anesthesia team with their sedation needs, assist with transporting the patient from the prep room, and help with monitoring of the patient. The nurses also help us keep the MR environment safe. You might say a radiology nurse is a “jack-of-all-trades.”

Our team of cardiologist/radiologists, cardiac anesthesiologists, cardiac schedulers, radiology nursing staff, and radiology management meet every three months to analyze the prior months of activity. It is our hope to refine the schedule so that it truly reflects appointment time and exam time while decreasing delay and allowing for the unexpected to occur.

The technology

As previously noted, as an example of a tertiary referral center, CHOP has three MRI scanners which perform CMR. Of these three MR units, one unit (MR6) is dedicated to cardiac and fetal imaging, while the other MR units (MR2 and MR4) are utilized for CMR when MR6 cannot be used, for certain research exams and as backup when all of the other scanners are full. The dedicated MRI scanner (MR6) is a hybrid system. The hybrid system combines MRI and cath labs with the use of a Myabia track system. This unique hybrid solution allows two different exams (CMR and cath) to be performed and combined together (cardiac physiology, function, 3D imaging, pressure measurements), while allowing the patient to be move from one modality to the other, and back if necessary, without physical movement of the patient (see Figure 25.1).

Table 25.1 Cardiac MR imaging (CMR) on three Siemens 1.5TAvanto systems.

<table>
<thead>
<tr>
<th>Manufacturer/CHOP designation</th>
<th>Software level</th>
<th>Number of channels</th>
<th>Number of cardiac slots</th>
</tr>
</thead>
<tbody>
<tr>
<td>Siemens Avanto/MR2</td>
<td>B15</td>
<td>32</td>
<td>20/monthly</td>
</tr>
<tr>
<td>Siemens Avanto/MR4</td>
<td>B15</td>
<td>32</td>
<td>As needed</td>
</tr>
<tr>
<td>Siemens Avanto/MR6</td>
<td>B15</td>
<td>32</td>
<td>78/monthly</td>
</tr>
</tbody>
</table>

processed the images correctly. They direct the technical staff when dealing with complex exams and refine image quality when needed. The cardiologist and radiologist provide the interpretation of the images in collaboration with the technologists and ensuring that the data has been processed to its maximum potential.

Cardiac technologist, 3D lab technologist

The technologists are responsible for ensuring that the patient is properly placed in the MRI scanner, that the correct exam is performed with the proper views and sequences, that the image quality is the best possible, and that the post-processing of data is done correctly and completely.

Cardiac schedulers

As with any radiology department, proper scheduling of the CMR, in conjunction with other imaging exams, is key. This is especially important in ensuring that sedated studies, studies requiring anesthesia and studies that do not require any medication to remain motionless are scheduled properly. Our cardiac center provides dedicated cardiac schedulers who work diligently to ensure the proper scheduling of all patients for their cardiac exams and procedures.

Cardiac sedation unit

It is important to provide the safest sedation for all our patients. To help us achieve the safest, yet most effective sedation, we employ a cardiac sedation unit run by a dedicated nursing staff, and cardiac anesthesiologists to perform our sedations/anesthesias. This team has the best understanding of our patients’ conditions and needs.

Radiology nursing staff

Our radiology nursing staff understand not only patient care, but also MRI imaging. They assist technologists with setting up the MR environment and positioning the patient. They aid the cardiac anesthesia team with their sedation needs, assist with transporting the patient from the prep room, and help with monitoring of the patient. The nurses also help us keep the MR environment safe. You might say a radiology nurse is a “jack-of-all-trades.”

Our team of cardiologist/radiologists, cardiac anesthesiologists, cardiac schedulers, radiology nursing staff, and radiology management meet every three months to analyze the prior months of activity. It is our hope to refine the schedule so that it truly reflects appointment time and exam time while decreasing delay and allowing for the unexpected to occur.
Education

An MRI technologist must practice reflective responsible scanning in order to be effective. The CMR technologist uses the same skill set as all MRI technologists; however their skills need to be especially advanced in order to perform a good pediatric exam. The age and size of the pediatric patient varies widely and for this reason, the MR technologist needs to employ a variety of techniques in order to obtain an optimal exam.

Educational opportunities, both in-house and outside, provide the technologist with the tools our hospital needs in performing a pediatric CMR exam.

Educational opportunities include the following:
- Dedicated weekly scheduled education and staff meeting time.
- Vendors’ system specific education.
- Dedicated cardiac imaging specific to the system.
- Quality insurance and education by the cardiologist.
- Education calendar includes lectures from invited guest speakers.
- Peer-to-peer feedback as education.
- Funding available for outside educational offerings.

Membership to organizations dedicated to cardiac imaging such as the Society for Cardiovascular Magnetic Resonance (SCMR) provide clinical education and the means to network with other facilities. Educational opportunities are invaluable for the development of the CMR technologist.

Our procedures

Screening

Since many of our pediatric patients are sedated, it is important that we design an MRI program that will keep them safe in our environment. A pediatric patient cannot communicate their needs once they are sedated. As with all MRI exams, we follow the guidelines of the American College of Radiology (ACR) and have our MR safety program.
reviewed yearly by an expert in the field of MRI safety, Frank Shellock Ph.D. Our MR safety program includes, but is not limited to, the following procedures:

- Check for all metallic devices.
- Pre-screening as far in advance as possible so that sedated patients are not delayed with metal, and devices discovered after sedation.
- For implanted devices, the manufacturer and serial number must be provided.
- Utilization of a two team screening process.
- Each patient should be visually checked before entering scan room.
- Infants 3 months and younger are provided with a cozy bunny suit to help regulate temperature.
- Obtain temperature reading on all infants 3 months of age and younger.
- All patients sedated or non-responsive must be monitored.
- If a child has a stent, a pre-scan of the heart must be done to evaluate metallic artifacts before sedation.
- Ears should be well-protected with age-appropriate ear plugs.

Due to the variations in the size of the child’s heart, we are imaging a structure that can be as small as a dime or as large as an adult fist. This variation in size requires careful selection of coils to provide the best performance and signal-to-noise to obtain the best image quality.

Currently we utilize one of the following coils when performing our CMR exam: 4-channel body array (Figure 25.2), head coil and neck array combination and a 32-channel cardiac coil (Figure 25.3).

Understanding how a child develops is effective in ensuring good communication between patient, parent and/or guardian. Age-specific competencies are essential and should cover all the stages of development including neonate, infant, toddler, preschool, school age, adolescent, young adult, and adult. Each patient is unique and it is important to keep in mind that while growth and development

**Room preparation**

**Coil selection**

It is crucial that we understand the development of the heart, from in utero to adulthood as well as basic cardiac anatomy. While the development of the heart can fill a chapter on its own, there are some key facts that will help the technologist in coil selection. The fetal heart occupies most of the fetus midsection. The heart size to body size ratio is nine times greater than an adult. The heart lies high in the chest of the fetus, then moves down to occupy the chest cavity.

After birth, the size of the child’s heart is about the size of his fist and it grows in this proportion until adulthood.

Pediatric patients are probably the most demanding patients for MR imaging due to the variation of size of the heart, growth, or clinical diagnosis. These patients utilize the smallest fields of view, thinnest slices, and highest matrix, with very little signal-to-noise. At the Children’s Hospital of Philadelphia patients range from 34 weeks in utero to 350 pounds.
Acknowledgments

It is a pleasure to express my sincere thanks to our technical team in taking the challenge of learning and understanding CMR. Thanks to Justine Wilson RT (R) MR, Eric Danz RT (R) MR and Deanna Tipton RT (R) MR. I would also like to thank our team of radiology nurses led by Terry Schultz, for their assistance in progressing this program to its current standard; our anesthesia team led by Dr. Susan Nicolson for providing us with their sedation expertise; our CMR physicians led by Dr. Mark Fogel, and last but not least our cardiac schedulers led by Marcie Wieder. Without this team, we could not be successful.

Clinical applications

Understanding the clinical diagnosis will help the technologist plan the basic CMR exam. While technologists may not understand complex clinical diagnoses of the cardiac patient, technologists have a basic understanding of routine CMR exams. Currently, our most frequent applications are tetralogy of Fallot (TOF), coarctation of the aorta, complex congenital heart disease after repair, such as transposition of the great arteries, truncus arteriosus and single ventricles.

In conclusion, an MRI technologist must practice reflective responsible scanning in order to be effective. It is essential to educate the technical staff in the capabilities of the MR system, patient’s anatomy, and development of the patient as well as MR safety. Educational opportunities, both in-house and outside, provide the technologist with the tools our hospital needs in performing the best pediatric CMR exam possible.

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