A. F. Corno
P. Festa

Congenital Heart Defects

Decision Making for Cardiac Surgery

Volume 3 CT-Scan and MRI

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Dedicated to our loved children

Federica, Laura, Jonathan, Bean

Laura, Paula, Pablo, Kiko

A. F. Corno P. Festa

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Volume 3 CT-Scan and MRI

Foreword by Richard Jonas

With 198 Figures in 400 Separate Illustrations

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Foreword

The diagnosis and management of congenital heart disease has evolved rapidly over the last 30 years. In the 1970s, invasive cardiac catheterization was a high-risk procedure for the infant and particularly the duct dependent neonate. The introduction of noninvasive 2-dimensional echocardiography revolutionized the diagnosis and management of individuals with congenital heart disease. No longer was the cardiologist limited to a handful of images and patient positions but could spend hours if necessary defining every detail of an individual child's cardiac anomaly. The addition of Doppler and subsequently color Doppler further increased the importance of echocardiography in the decision making for congenital cardiac surgical procedures. And now we are fortunate to have further additions to the imaging and diagnostic armamentarium, namely cardiac CT and cardiac MRI.

In this most recent volume of the series entitled "Congenital Heart Defects: Decision Making for Surgery" Dr. Antonio F. Corno provides an up-to-date and comprehensive presentation of the new role that cardiac CT and cardiac MRI will play in the management of congenital heart disease. He has been ably assisted by a cardiologist, Dr. Gigi P. Festa.

The book provides a dazzling array of images derived by both techniques and covering the full range of congenital heart anomalies. Both the pre-operative and postoperative usefulness of these techniques is presented.

There is no doubt that these techniques will be particularly helpful in the older child and adult with congenital heart disease in assessing the late impact of a congenital heart anomaly and the surgical repair or palliation which may have been undertaken years previously.

The time is definitely right for a comprehensive presentation of these new diagnostic modalities and the manner in which they will influence the field of congenital heart management.

Dr. Corno is to be congratulated on seizing this opportunity and producing a beautifully illustrated textbook that will be the standard against which others will be measured.

Washington, DC
August 2008

Richard Jonas, M.D.

Preface

The first two volumes of this series on "Congenital Heart Defects – Decision making for surgery" have been dedicated to the "Most common defects (Volume 1)" and to "Less common defects (Volume 2)". The schema utilized for these two books was the same, with each chapter devoted to a single malformation, with incidence, morphology, associated anomalies, pathophysiology, diagnosis (including clinical pattern, electrocardiogram, chest X-ray, echocardiogram, and cardiac catheterization with angiography), indications for surgical treatment, details of surgical techniques, potential complications and literature references.

During the last few years computed tomography (CT) scan and magnetic resonance imaging (MRI) have emerged as valuable noninvasive cardiovascular diagnostic tools. Both CT scan and MRI are capable of producing stunning 3-dimensional pictures independent of body size. In particular cardiac MRI can provide unique anatomic and functional information not available by any other diagnostic modality previously available. Because of the progressively increasing role of CT scan and MRI in the pre- and postoperative evaluation of patients with congenital heart defects, it seemed natural to continue this series of volumes preparing a third book entirely dedicated to these two diagnostic techniques.

The sole author of the first two volumes (AFC) decided to involve Dr. Gigi P. Festa, cardiologist with extensive expertise particularly in cardiac MRI, in the current task. The choice was due not only to his competence in the matter, but to the fact that we worked together years ago, first in Milan, Italy, then in Paris, France, with Dr. Yves Lecompte. The mutual respect that develops among colleagues is not enough to write a book, particularly when working as nowadays in different departments and living in different countries, with different professional schedules and family commitments, despite the advantages of the communication feasible through the internet. A deep friendship is indispensable to reach agreement on every single detail, including the entire text and the choice of illustrations.

We agreed to maintain the extremely schematic format of the book, following the style of the first two volumes, in order to provide the essential information to the readers. Since our list of congenital heart defects is derived from the sum of malformations treated in the first two volumes in this series, most of the clinical data on conventional diagnostic techniques (electrocardiography, X-ray, echocardiography, cardiac catheterization with angiography), indication for surgery and surgical techniques have been omitted to avoid duplication with the other volumes of the series. Only a selected num-

ber of illustrations, as well as number of references, could be used because of space limitations. Finally, since the pre-operative and post-operative illustrations are coming from different departments and different periods, it is quite clear that the patients have been treated with either interventional cardiology or surgical techniques by different operators, not necessarily by the authors.

Liverpool and Massa, Summer 2008

Antonio F. Corno
Gigi P. Festa

Acknowledgements

Since this book is the result of our combined experience, we would like to thank all the individuals who contributed to developing our current knowledge in the field of congenital heart defects.

Acknowledgement begins with all the sick children encountered during our professional lives, and also their families, who gave us the permission to publish the images obtained from the investigations on their children. Then to all the colleagues, nurses and technicians who have participated in the pre-operative, intraoperative and postoperative care of all the patients, and of course to all the individuals (colleagues, technicians, nurses, secretaries) involved in the CT scan and MRI investigations, mostly performed at the Radiology Department of Alder Hey Children's Hospital, Liverpool, and the MRI Laboratory of Institute of Clinical Physiology, CNR, Pisa.

This book has been stimulated by the pioneering work of Dr. Tal Geva, Boston, who not only opened the pathway towards the investigation of congenital heart defects with these new diagnostic techniques, but was able to transmit the passion for the know-how thanks to his very extensive knowledge, didactical capability and passionate approach.

Special mention is addressed to Dr. Mohamed Tawil, Consultant Pediatric Radiologist and to Dr. Robert A. Johnson, Consultant Pediatric Cardiologist, Alder Hey Hospital, Liverpool, both with a special interest in pediatric cardiothoracic imaging, who were available to help not only in the selection of the illustrations, but also in reviewing part of the text. In particular Dr. Mohamed Tawil is responsible for all the pre- and postoperative investigations in the pediatric cardiac patients in Alder Hey Hospital.

Another special mention goes to Dr. Lamia Ait-Ali, Cardiologist at Pasquinucci CNR Hospital, Massa. Thanks to her extensive experience with imaging post-processing and to her tireless dedication, she made a major contribution to preparing this book.

Special thanks are extended to Prof. Luigi Donato, Director of the C.N.R./Regione Toscana Foundation, whose whole life has been dedicated to the development of a particular environment with combined clinical activity and research in cardiopulmonary diseases, leading to the setting of the MRI Laboratory in Pisa, one of the few in Europe fully dedicated to cardiac investigations including congenital heart defects.

Strong encouragement to complete the book came from eminent pediatric cardiologists, including Roberta G. Williams and the late William F. Friedman from Los Angeles, California, and from Bruno Marino, Rome.

Also pediatric cardiac surgeons were very supportive of the idea of a book dedicated to pre- and postoperative CT scan and MRI of congenital heart defects, in particular Aldo R. Castañeda, Guatemala City, Yves Lecompte, Paris, and Richard A. Jonas, Washington, D.C., who very kindly contributed the Foreword.

Everyone from the Publisher's side has to be acknowledged, in particular Susanne Denskus and Annette Gasser, because of their patience and compliance with our increasing requests for more space for both text and illustrations.

Two companies were instrumental in supporting part of the costs of publication: Philips UK (in particular Alistair Howseman) and General Electric Italy (in particular Roberto Molinari).

We apologize in advance for any omissions or mistakes that might be called to our attention.

As a profession constitutes only a part of our life, we deeply acknowledge our families' unconditional support, particularly the patience from our wives Josie and Arantxa for the long hours spent evenings and on weekends working to complete this book.

Liverpool and Massa, Summer 2008

Antonio F. Corno
Gigi P. Festa

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Chapter 3.0 Introduction to CT scan and MRI

Introduction

Transthoracic echocardiography is the first-line imaging modality for diagnosis and follow-up of patients with congenital heart defects due to its low cost, widespread availability, portability, ease of use, and excellent temporal-spatial resolution, reducing the need for diagnostic cardiac catheterization and angiography in most patients. However, the diagnostic utility of echocardiography markedly diminishes with the growth of the patients and after surgical procedures through median sternotomy because acoustic windows become progressively more limited. Moreover extracardiac structures, such as the great arteries and great veins, complex intracardiac connections or surgically implanted conduits or baffles deeply located in the chest or behind the sternum, may also be difficult to resolve by ultrasound. Diagnostic cardiac catheterization carries risk of complications because it is invasive and is associated with radiation exposure; furthermore as a projection technique, it is limited in providing accurate anatomical information. In the last two decades, computed tomography (CT) scan and magnetic resonance imaging (MRI) have emerged as valuable noninvasive cardiovascular diagnostic tools capable of producing informative pictures, providing unique anatomic and functional information not available by any other diagnostic modality currently available. For the assessment of the anatomy of the extracardiac structures, the clinical indication for CT scan and MRI are somewhat similar, and very often the question which is better to use is raised. The main differences as well as advantages and disadvantages are highlighted in Table 3.0.1.

The easy of use and the short time required for the complete investigation make the CT very attractive, particularly in children. However, according to the recent literature, its radiation exposure represents a major concern, mainly in children and its use should be limited whenever possible as recommended by the FDA in 2001. While the CT scan still remains a very valuable tool to provide detailed information about extracardiac structures, the available clinical experience with MRI is becoming richer than that with CT due its better capacity to assess intracardiac morphology and cardiac function, and its better versatility.

The choice between CT and MRI has to be based on the available institutional equipment and scheduling, the capability of the

Table 3.0.1. CT scan and MRI

	MRI	CT
Examination time	+ (long)	++++ (short)
Great vessels image resolution	+++	++++
Tissue characterization	+++	+
Calcification analysis	No	Yes
Flow analysis	Yes	No
Coronary artery imaging	++	+++
Ventricular function analysis	++++	+
Airway analysis	++	++++
Ionizing radiation	No	Yes
User friendly	+	+++
Costs	+++	+

patient to cooperate, and the need to tailor the investigation to answer the specific question being asked. Cardiologists and radiologists should be provided accurate estimates of CT radiation doses to allow for a balanced and accurate assessment of the risk/benefits ratio when considering indication for a CT scan.

Computed tomography

General principles

Computed tomography (CT) is a medical imaging method using tomography where digital geometry processing generates a 3-dimensional image from a large series of 2-dimensional X-ray images. Although historically the generated images were in the axial or transverse plane, orthogonal to the long axis of the body, modern computational techniques allow this volume data to be reformatted into various planes or even as 3-dimensional representations. CT has undergone staged evolution leading to increased spatial resolution and temporal resolution. Initial single slice CT has evolved into helically scanning CT in which large volumes could be included by the translation of the subject along the axis of scanner. By introducing multiple detectors (multislice CT) including arrays detectors, commonly 64 in the current clinical practice, increased resolution and/or volumes can be imaged on shorter time scales. This has led to the ability to perform high-resolution scans gated to the cardiac cycle.

Patient preparation

The need of sedation in infants and children less than 6 years of age has decreased as the speed of CT scanning has increased. While the sedation rate for single-detector row CT was at least 30%, the sedation rate for young children undergoing multidetector row CT is less than 5%. Despite this improvement, sedation has not yet been eliminated, and knowledge of safe and effective use of sedation remains of paramount importance.

Oral administration of chloral hydrate is used as a sedative agent in children, while deep sedation (propofol) or general anesthesia (isoflurane) is used in infants. In older children adequate preparation with verbal reassurance and explanation of the procedure can generally obtain good cooperation.

Methodology and technical aspects

Intravenous contrast material: The issues in the administration of intravenous contrast material for CT angiography in pediatric patients include type and volume of contrast material, method of administration (manual versus power injection), and delay between the start of injection and the initiation of scanning. Intravenous contrast material is not required for routine investigation of the airways, but is essential in children with suspected para-tracheal malformation such as vascular rings or pulmonary artery slings. Through an intravenous cannula placed in the CT suite, the nonionic contrast material is administered at a dose of 2.0 mL/kg. The use of a nonionic agent minimizes the gastrointestinal side effects such as nausea and vomiting, the discomfort at the site of injection, the patient reaction during the administration of intravenous contrast material, and potential complications from contrast material extravasations. With multidetector row CT volumes as small as 2 mL/kg allow successful contrast enhancement for CT angiography. Intravenous contrast material can be administered by manual (hand) or power injection. The advantages of power over manual injection are the ability to precisely determine the timing of delivery of the contrast material and the uniformity of enhancement. The flow rates vary with the size of the intravenous catheter, from 1.5 to 2.0 mL/s for a 22-gauge catheter to 2 to 3 mL/s for a 20-gauge catheter. During the injection

of contrast material, the site of injection needs to be closely monitored to minimize the risk of extravasations. As a general rule the contrast material is administered with a flow rate calculated to deliver the entire volume of contrast material in a period equivalent to, or slightly less than, the duration of the CT acquisition. There is a low but non-negligible level of risk associated with the intravenous administration of the contrast agent itself: severe and potentially life-threatening allergic reaction and renal insufficiency. Because of these risks, ionic contrast agents are generally avoided.

Scanning delay time: The time between the start of the injection of contrast material and the start of the scan data acquisition is the delay time. Empirically the duration of the scanning time should be set to obtain the maximum of contrast concentration in the region of interest, in order to provide excellent vascular enhancement in children. An automated tracking system, taking into account variable factors such as cardiac output and circulation time, allows customization of contrast enhancement for each individual patient.

Technical parameters: Before scanning is started, several technical parameters have to be selected, including collimator thickness, table speed, tube current or milliamperage, kilovoltage, and anatomic coverage required. Sections of 1 mm thickness are generally used in children, to maintain accurate definition without increasing the radiation dose. The table speed is set as fast as possible to increase the temporal resolution and to decrease the radiation combined with detector array dimensions. Electrocardiographic gating is able to reduce artifacts related to cardiac motion and arterial pulsation, but it does not affect the respiratory motion.

Radiation exposure: In children the issue of radiation exposure is extremely important. Since children's organs are more radiosensitive than adults' and they have a longer life span than adults, the development of radiation-induced malignancies is a real potential risk, particularly in younger children. On average, the radiation exposure during a CT scan reaches 2.0 to 2.5 rems, in comparison with 1.5 to 2.0 rems during a diagnostic cardiac catheterization with angiography. The radiation dose for a particular CT study depends on multiple factors: volume scanned, patient size, number and type of scan studies, and desired resolution and image quality. In general, multidetector row CT should be performed with techniques providing adequate quality of images with the lowest possible radiation exposure. These techniques include the use of the lowest milliamperage and kilovoltage settings, appropriate section thickness, and the fastest table speed. Adjustments of specific technical factors proved to minimize the radiation dose in children undergoing CT, particularly in state-of-the-art multisection spiral CT: such adjustments include setting the lowest diagnostic tube current according to the patient's weight and doubling the pitch, which reduces the radiation dose by half. The use of ECG prospective gating in multislice CT scan allows a further reduction of the radiation exposure, thanks to the acquisition of images limited to 70–80% of the R-R interval. In addition, multiphase studies should be used for selected indications rather than on a routine basis.

Anatomic coverage: A minimal coverage for CT angiography should extend from the level of the diaphragm inferiorly to just below the thoracic inlet, in order to include the proximal aspects of the common carotid arteries and of the subclavian arteries in the CT scan. This inclusion allows visualization of the position of the aortic arch, presence of aortic coarctation, presence and degree of hypoplasia of the aortic arch, and anomalous origin of the head and neck vessels. The selected field of view has to approximate the cross-sectional size of the region being studied. A smaller field of view can reduce a waste of matrix space, a loss of resolution, and incidence of poor quality images.

Breath holding: CT scan investigations are performed with breath holding at suspended inspiration in cooperative grown-up children and during quiet respiration in children under sedation.

Reconstruction algorithms: A standard reconstruction algorithm is generally enough in routine CT angiography studies. A high resolution algorithm is used for 3-dimensional reconstructions involving the airways.

Post-processing applications

Various options are available for post-processing the volumetric data set.

Multiplanar reconstructions: Sections of multiplanar reformations can be displayed in any plane: coronal, sagittal, parasagittal or in curved isotropic datasets, such as along the axis of the mediastinal vessels or airways. This technique is fast and can be easily performed at the CT scanner, but it provides only a 2-dimensional display of data (Figs. 3.0.1

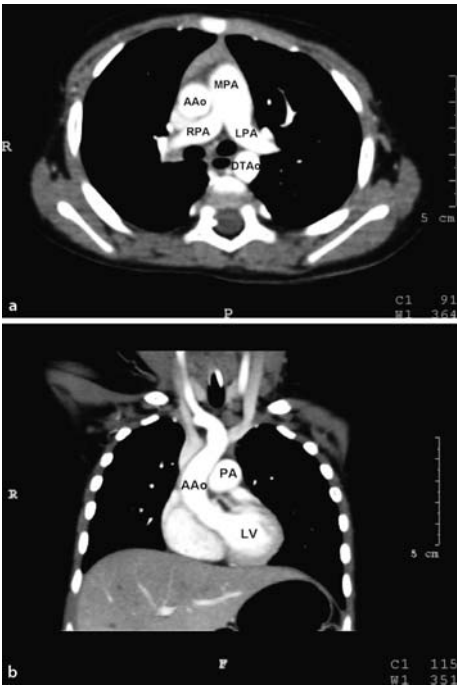

Fig. 3.0.1. CT scan of normal anatomy. **a** Axial section of a 2-dimensional study showing the relationship between the great arteries. **b** Para-coronal plan of a 2-dimensional study showing the relationship between the great arteries, in the presence of situs solitus, with the liver on the right side and the stomach on the left side (*AAo* ascending aorta, *DTAo* descending thoracic aorta, *LPA* left pulmonary artery, *MPA* main pulmonary artery, *RPA* right pulmonary artery, *LV* left ventricle, *PA* pulmonary artery)

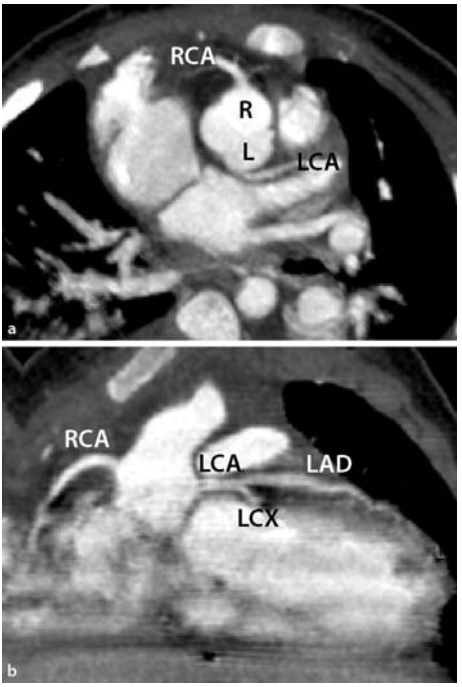

Fig. 3.0.2. CT scan of normal coronary arteries. CT angiography with axial (**a**) and coronal (**b**) projection showing the normal origin and course of right and left coronary arteries, respectively, from the right and left aortic sinus (*LAD* left anterior descending coronary artery, *LCA* left coronary artery, *LCX* left circumflex coronary artery, *RCA* right coronary artery) (reproduced with permission from Goo HW, Park IS, Ko JK, Kim YH, Seo DM, Yun TJ, Park JJ, Yoon CH (2003) CT of congenital heart disease: normal anatomy and typical pathologic conditions. Radiographics 23:S147–165)

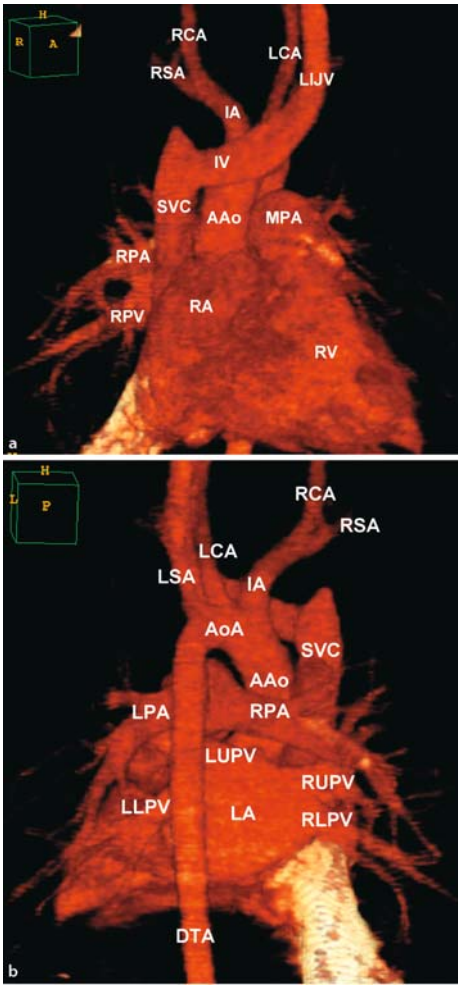

Fig. 3.0.3. CT scan of normal anatomy. **a** CT angiography with 3-dimensional reconstruction, oblique right anterior view of the normal cardiac anatomy. **b** CT angiography with 3-dimensional reconstruction, posterior view of the normal cardiac anatomy (*AAo* ascending aorta, *AoA* aortic arch, *DTA* descending thoracic aorta, *IA* innominate artery, *IV* innominate vein, *LA* left atrium, *LCA* left carotid artery, *LIJV* left internal jugular vein, *LLPV* left lower pulmonary vein, *LPA* left pulmonary artery, *LSA* left subclavian artery, *LUPV* left upper pulmonary vein, *MPA* main pulmonary artery, *RA* right atrium, *RCA* right carotid artery, *RLPV* right lower pulmonary vein, *RPA* right pulmonary artery, *RPV* right pulmonary veins, *RSA* right subclavian artery, *RUPV* right upper pulmonary vein, *RV* right ventricle, *SVC* superior vena cava)

and 3.0.2), and information regarding the 3-dimensional spatial relationship of anatomic structures is absent. Curved planar reconstructions are a variant of the routine multiplanar reconstructions, allowing curved or tortuous vessels such as the aorta and the pulmonary artery to be visualized in a single tomography volume.

Variable-thickness displays: With this technique, CT images are acquired at their routine thickness section, and then combined in multiples ("slabs") to create a thicker image. This allows better visualization of smaller pulmonary vessels and airways.

3-Dimensional volume rendering: Volume rendering has largely replaced other 3-dimensional reformatting techniques, because the use of a transfer function allows mapping of a CT number to brightness and color. This provides valuable information regarding the spatial relationship of anatomic structures, and it also allows the data to be displayed from an external or internal perspective. The volume-rendering technique is particularly useful for structures which are 3-dimensional and cannot be easily captured in a planar image (Fig. 3.0.3). The creation of volume-rendered images requires secondary processing, and the images can generally be produced in a few minutes.

Magnetic resonance imaging

General principles

Cardiac MRI in the pediatric population is accompanied by a unique series of technical challenges superior to those encountered in imaging of adult patients: the anatomical structures are smaller, thus, demanding greater spatial resolution; the heart rates are generally higher, requiring a greater temporal resolution; and the pediatric patients can be either noncooperative or may require full sedation. Despite these technical difficulties,

cardiac MRI offers several characteristics favorable in comparison with the conventional imaging modalities, including soft tissue contrast, capacity for true 3-dimensional imaging, accurate flow measurements, freely selectable imaging planes, and lack of ionizing radiation. These advantages, in addition to continued advances in MRI hardware, software, and imaging techniques, are progressively increasing the widespread utilization of cardiac MRI in pediatric cardiology.

Through the use of several MRI techniques, examiners can obtain high-quality 3-dimensional images of the cardiovascular anatomy, accurately quantify volumes and mass of cardiac chambers, ejection fraction, stroke volumes, regurgitation volume and fraction, as well as regional left and right ventricular myocardial function. Therefore, MRI is ideal for noninvasive pre-operative evaluation of patients with complex intracardiac anatomy, anomalous connections of the systemic and pulmonary veins, anomalous pulmonary arteries, defects involving the aorta and its branches, single ventricles, systemic right ventricles and abnormally shaped left ventricles. MRI has also acquired a precise role in the postoperative evaluation of patients after complex surgery, such as, after repair of tetralogy of Fallot, univentricular type of circulation, and in the presence of abnormal blood flow dynamics.

The clinical indications for cardiac MRI are rapidly evolving due to recent considerable technical advances and potential benefits. In 2004, MRI clinical indications were published by a consensus panel composed of European and American cardiologists and radiologists with major input from members with additional established expertise in pediatric cardiology, nuclear cardiology, magnetic resonance physics and spectroscopy, as well as health economics (Table 3.0.2). Interestingly, cardiac MRI expertise is highly recommended in centers dedicated to the treatment of congenital heart defects. Moreover evaluation and follow-up of adults with congenital heart disease is considered as a Class I indication for a comprehensive cardiac MRI investigation, since it is now possible to often answer anatomic and functional questions unresolved with other noninvasive investigations.

Cardiac MRI techniques are generally more operator dependent than other MRI techniques; however, some points should be highlighted: for a reliable study, a thorough understanding of the anatomic and functional principles, as well as the knowledge of the available interventional and surgical therapeutic options for congenital heart defects is nevertheless required. A comprehensive pathophysiological cardiac MRI evaluation, especially if dealing with complex congenital heart defects, requires a long time and may not be well tolerated by the patient. Therefore, accurate and detailed preparatory steps are required in planning the cardiac MRI investigation to precisely define the specific questions to be answered as some lesions may have already been well characterized by other diagnostic modalities. Hence the complete knowledge of all the details of the anatomic and functional findings derived from prior clinical, echocardiography or cardiac catheterization and angiographic studies, as well extensive communication with the referring physician are essential. Frequently, a direct review of previous echocardiography investigation is warranted to check some details of the myocardial function such as ventricular stroke volume in case of atrioventricular valve regurgitation. When dealing with patients with previous interventional and/or surgical procedures, direct access to the previous reports is extremely helpful, as well as the knowledge of the presence of implanted devices as potential sources of artifacts. Furthermore, excellent knowledge of the MRI techniques and physics is warranted to reach high accuracy both in terms of anatomical detail and, more importantly, functional information, often heavily determinant in view of the surgical indication. Thus, every cardiac MRI laboratory should strictly test their own interobserver reproducibility as well as a rigorous quality control of instruments.

Table 3.0.2. Indication for cardiac MRI in congenital heart defects (reproduced with permission from Pennell DJ, Sechtem UP, Higgins CB, Manning WJ, Pohost GM, Rademakers FE, van Rossum AC, Shaw LJ, Yucel EK (2004) Clinical indications for cardiovascular magnetic resonance (CMR): Consensus Panel Report. Eur Heart J 25(21):1940–1965. By permission of Oxford University Press)

Indication	Class
General indications	
1. Initial evaluation and follow-up of adult congenital heart disease	I
Specific indications	
1. Assessment of shunt size (Qp/Qs)	I
2. Anomalies of the viscero-atrial situs	
▪ Isolated situs anomalies	II
▪ Situs anomalies with complex congenital heart disease	I
3. Anomalies of the atria and venous return	
▪ Atrial septal defect (secundum and primum)	II
▪ Anomalous pulmonary venous return, especially in complex anomalies and cor triatriatum	I
▪ Anomalous systemic venous return	I
▪ Systemic or pulmonary venous obstruction following intra-atrial baffle repair or correction of anomalous pulmonary venous return	I
4. Anomalies of the atrioventricular valves	
▪ Anatomic anomalies of the mitral and tricuspid valves	II
▪ Functional valvular anomaly	II
▪ Ebstein's anomaly	II
▪ Atrioventricular septal defect	II
5. Anomalies of the ventricles	
▪ Isolated ventricular septal defect	III
▪ VSD associated with complex anomalies	I
▪ Ventricular aneurysms and diverticula	II
▪ Supracristal VSD	I
▪ Evaluation of right and left ventricular volumes, mass and function	I
6. Anomalies of the semilunar valves	
▪ Isolated valvular pulmonary stenosis and valvular dysplasia	III
▪ Supravalvular pulmonary stenosis	II
▪ Pulmonary regurgitation	I
▪ Isolated valvular aortic stenosis	III
▪ Subaortic stenosis	III
▪ Supravalvular aortic stenosis	I
7. Anomalies of the arteries	
▪ Malpositions of the great arteries	II
▪ Postoperative follow-up of shunts	I
▪ Aortic (sinus Valsalva) aneurysm	I
▪ Aortic coarctation	I
▪ Vascular rings	I
▪ Patent ductus arteriosus	III
▪ Aortopulmonary window	I
▪ Coronary artery anomalies in infants	Inv
▪ Anomalous origin of coronary arteries in adults and children	I
▪ Pulmonary atresia	I
▪ Central pulmonary stenosis	I
▪ Peripheral pulmonary stenosis	Inv
▪ Systemic to pulmonary collaterals	I

Methodology and technical aspects

To discuss the MRI technique is not the main aim of this chapter, however, we will give an overview on cardiac MRI methodology and technical aspects in order to make the readers more familiar with the technique.

The goals of a pre-operative cardiac MRI investigation in patients with congenital heart defects are the following:

- define the morphological characteristics and the location of the primary malformation and the potential presence of associated defects,
- evaluate and quantify the functional consequences of the primary malformation.

Morphological assessment: Although there are several cardiac pulse sequences for MRI morphologic imaging, many with manufacturer-specific features, they can be grouped into three classes:

- *Black-blood imaging:* black-blood images are images with high signal-to-noise ratio, resolution and contrast. Most techniques use radiofrequency refocusing, making them robust to metal artifacts. This is important in the evaluation of postoperative patients where surgical clips would other-

wise cause local image voids and anatomic disruption. Black-blood techniques become very useful when the relationships between the airway and vessels must be elucidated. They are also extremely useful in characterizing the abdominal situs, the presence of masses, thrombi, hematomas or other soft tissue details as fatty infiltration as well as edema.

- *White-blood cine imaging:* static pictures as black blood images can answer many questions in pediatric cardiology, but they incompletely characterize the details of the malformation. Any sonographer can relate to the difficulty of interpreting still-frames removed from their dynamic context. Although cine images in MRI can be prospective and retrospectively-gated (not real time), they offer the same physiologic context provided by echocardiography. In most instances the required spatial and temporal resolution makes it impossible to acquire a full series of images in a single heart beat. Instead, each image is assembled from data acquired during several heart beats (segmentation). Cine images represent the backbone of congenital cardiac MRI even though they have lower resolution and contrast than black-blood imaging. In recent years, steady-state free precession (sequences also known as balanced FFE, TrueFisp, and FIESTA) has practically replaced previously used spoiled gradient sequences. The steady-state free precession is extremely useful for imaging of the cardiac function because of its specific properties. Strong T2/T1-weighted and homogeneous contrast shows cavitary blood bright and myocardium with lower signal intensity. Contrast is virtually independent of inflow, imaging at consistent quality in all orientations, including horizontal and vertical long axes. The steady-state free precession sequences are also fast, allowing acquisition of a single slice heart phases in a breath-hold of a few seconds duration.

  However, some disadvantages of the steady-state free precession have to be mentioned: the ultra-short TE produces lower flow sensitivity for vascular or valvular obstructions. Even more important steady-state free precession is troubled by flow artifacts near regions of rapid blood acceleration. These artifacts can be even more crippling in infants and young children since they have significantly greater blood acceleration than adult patients in the aorta and the great arteries. Local shimming improves but does not eliminate these artifacts. Another limit of steady-state free precession is that reducing the field of view and slice thickness can greatly prolong the echo and repetition times (this is done to reduce the absorbed radiofrequency power and gradient switching rates).
- *3D gadolinium-enhanced angiography:* contrast-enhanced magnetic resonance angiography (CEMRA) is the most important 3-dimensional technique available for routine cardiac assessment. Since voxels are more isotropic than for 2-dimensional acquisitions, images collected by 3-dimensional sequences are ideally suited for multiplanar reformatting and volume rendering. CEMRA acquisitions are un-gated, so they are only useful for characterization of extracardiac structures. Despite short acquisition times, CEMRA has a high signal-to-noise ratio, allowing the highest resolution of any of the cardiac imaging sequences. Although large vessel disease may be well characterized by standard 2-dimensional imaging, CEMRA is absolutely essential to visualize small vessels such as in peripheral pulmonary stenosis or the presence of aortopulmonary collateral arteries. In fact, CEMRA offers comparable diagnostic accuracy as cardiac catheterization and angiography in evaluating the number and location of aortopulmonary collaterals in children and adults with pulmonary atresia.

Functional assessment: MRI provides unique functional information of major value related to ventricular function, flow as-

sessment, myocardial perfusion and viability. Information on function and flow is obtained from time-resolved series of gradient echo images (white blood), each time frame corresponding to a phase of the cardiac cycle.

- *Biventricular function:* one of the advantages of MRI over echocardiography in the assessment of ventricular volumes, ejection fraction and mass is its ability to reproducibly and accurately acquire parallel images in any orientation, eliminating geometrical assumptions. This advantage is especially important in congenital heart disease where the intracardiac morphology does not fit the usual shape assumptions. One example is the unique ability of cardiac MRI to determine ventricular function in patients with a single ventricle, either of right or left ventricular morphology. A single ventricle has an extremely variable and bizarre shape, difficult to mathematically model and which does not allow the geometric assumptions necessary to calculate mass, volume, or cardiac performance from a single image plane. Compared with other noninvasive imaging tools, cardiac MRI provides a precise assessment of the ventricular mass, volumes and function necessary for the early and late postoperative follow-up in these complex patients. The use of MRI has also been extensively validated for the quantification of the right ventricular function.

  For measurement of ventricular volumes, a number of adjacent slices are acquired in the short-axis orientation covering them from apex to base. For each short-axis slice, a region of interest is defined by outlining the endocardial contour of the left or right ventricle on the end-systolic and end-diastolic images. For each slice, the area of the region of interest is multiplied by the slice thickness. Summation over all slices results in end-systolic and end-diastolic volume, respectively. The difference between the two volumes corresponds to the stroke volume. From these numbers the ejection fraction is calculated as ejection fraction = stroke volume/end-diastolic volume. Cardiac index is calculated as cardiac output per minute, normalized by the body surface area.
- *Velocity mapping:* quantification of blood flow is made using velocity-encoded cine MRI sequence, also called phase contrast velocity mapping (PVC-MRI), through planes transecting the targeted vessel (Fig. 3.0.4), which can provide information on blood flow in the major vessels, such as

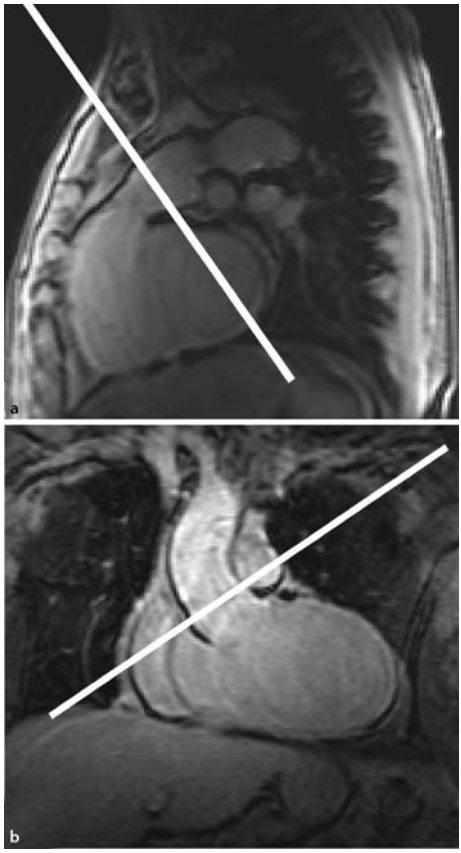

Fig. 3.0.4. MRI: measurement of blood flow. Coronal (**a**) and sagittal (**b**) projection to measure the aortic (*QS* systemic) and the pulmonary artery (*QP* pulmonary) blood flow (see Fig. 3.0.5)

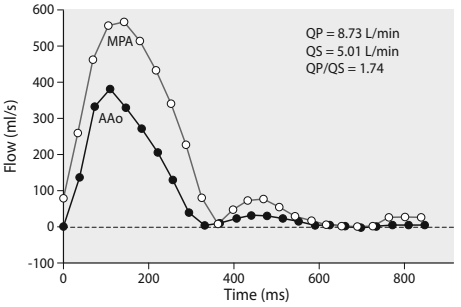

Fig. 3.0.5. MRI: measurement of blood flow. Graphic representation of the measurement of systemic (QS) and pulmonary (QP) blood flow, and calculation of the QP/QS

aorta, pulmonary arteries, central veins, and surgically implanted conduits. It represents a unique and unrivaled strength of MRI. The main clinical applications of PVC-MRI, very useful in the assessment of congenital heart defects before and after surgery, are the assessment of the systemic and pulmonary blood flow, the pulmonary to systemic flow ratio (QP/QS) (Fig. 3.0.5), valvular regurgitation, peak velocity measurement in stenotic vessels and valves, lung perfusion and flow dynamics. However, even when appropriate methods of acquisition have been used, there can be inaccuracies of flow measurement caused by background phase errors due to eddy current or uncorrected concomitant gradients. Furthermore for accurate measurements of aortic regurgitation or mitral valve inflow, motion tracking and velocity correction with respect to the cyclic displacements of the valves are needed, but few if any commercial systems provide this facility. Regarding the jet velocity, its measurement poses different challenges, mainly related to the voxel size and slice orientation relative to respectively a narrow jet and jet orientation. A recent application of MRI is to study the compliance of the wall of the great arteries (aorta and pulmonary artery) subjected to a chronic increase of the intravascular blood pressure.

- *Myocardial tagging:* myocardial tagging allows a more thorough investigation of the cardiac motion, providing detailed information on translation, rotation, and deformation, and allows calculation of measures, such as wall motion, regional wall thickening, and strain. This is obtained by destroying coherent magnetization periodically over space and subsequently performing a cine acquisition using a gradient echo sequence. Among other research applications, myocardial tagging has been used to investigate function in children and adult patients with single left ventricle and after heart transplant.
- *Myocardial perfusion and viability:* in adults with coronary artery disease contrast-enhanced MRI allows the identification of myocardial perfusion, extent and diffusion of myocardial ischemia, infarction, and viability. In patients with complex congenital heart disease, myocardial ischemia and fibrosis can also complicate the early and particularly the late follow-up after intracardiac surgery.
- *First pass perfusion:* the currently available MRI perfusion measurement techniques are all based on the dynamic imaging of the first-pass through the myocardium of gadolinium-based T1-shortening contrast agents. Adenosine is used as a coronary vasodilator to create a blood flow difference between the myocardial regions perfused by normal coronary arteries and those supplied by arteries with stenosis. As the contrast agent passes through the blood pool and myocardial tissue, the T1 is shortened, and the signal enhanced. Generally, 20 to 30 seconds are required for the first-pass of contrast agent through the myocardium from the time of injection. This time defines the length of the scan as images must be acquired throughout the passage of contrast agent through the myocardium. Ischemic regions with poor perfusion will not enhance at the same rate as normally perfused tissue and can be visualized as regions of transient dark signal.

Preliminary results demonstrate that MRI evaluation of myocardial perfusion and viability is feasible in pediatric patients with congenital and acquired heart disease, although so far there is a limited application of MRI perfusion imaging in the pediatric population. Cardiac MRI presents advantages in comparison with nuclear scintigraphy, including the lack of ionizing radiation, a very important factor when considering stress imaging in young patients who may require lifelong assessment of myocardial ischemia and function. In comparison with nuclear perfusion examination, which may require 2 to 6 hours, the cardiac MRI examination is generally completed in less than 1 hour. In addition the high spatial resolution of MRI is particularly helpful in pediatric patients because it allows even smaller perfusion defects to be detected. One all-inclusive study also provides complimentary data, such as regional wall motion abnormalities and delayed enhancement to evaluate viability. The diagnostic utility and clinical benefit in the pediatric population with congenital and acquired heart disease will be further addressed by future studies.

- *Delayed myocardial enhancement:* gadolinium-based MRI contrast agents in clinical use are all T1-shortening, extravascular, extracellular agents, rapidly diffusing out of the capillaries into the interstitial space. In adult patients, the areas of acute or chronic myocardial infarction provide a larger distribution volume for these agents when compared to viable tissue. T1-weighted, inversion recovery imaging can differentiate necrotic regions of delayed-enhancement from viable myocardium. The inversion time is set to null the signal from viable myocardium, to produce images in which necrotic tissue is bright by virtue of its shorter T1. In practice, the optimal T1 is based on the dose of contrast administered and the elapsed time between injection and imaging. The assessment of the transmural extent of viability by cardiac MRI is not available from other noninvasive imaging techniques. In adult patients with coronary artery disease, the late gadolinium-enhanced cardiac MRI is able to predict whether regions of abnormal ventricular contraction will improve after myocardial revascularization. At the moment, there is only limited experience with this technique in children, even if in infants with an anomalous left coronary artery from the pulmonary artery syndrome and severe left ventricular dysfunction, the impact of myocardial viability, utilizing delayed enhancement to detect ventricular infarct and myocardial fibrosis, has already been reported to direct the surgical choice between the two options of coronary artery reimplantation versus cardiac transplantation.
  This technique of post-gadolinium myocardial-delayed enhancement is increasingly used in patients after repair of congenital heart defects to assess the presence and extent of area with myocardial fibrosis.

Sedation and anesthesia

A comprehensive cardiac MRI investigation requires approximately 30 to 60 minutes with the patient in a still position in an uncomfortable environment; therefore, children under the age of 12 will commonly require anesthesia. The use of conscious sedation (such as pentobarbital or chloral hydrate), deep sedation (propofol), or general anesthesia varies tremendously among different hospitals depending upon local preferences and resources. Although general anesthesia with intubation and paralysis is frequently seen as excessively invasive, it is probably safer and more predictable than other strategies in young children. However it becomes highly preferable in patients with vulnerable airways or in infants.

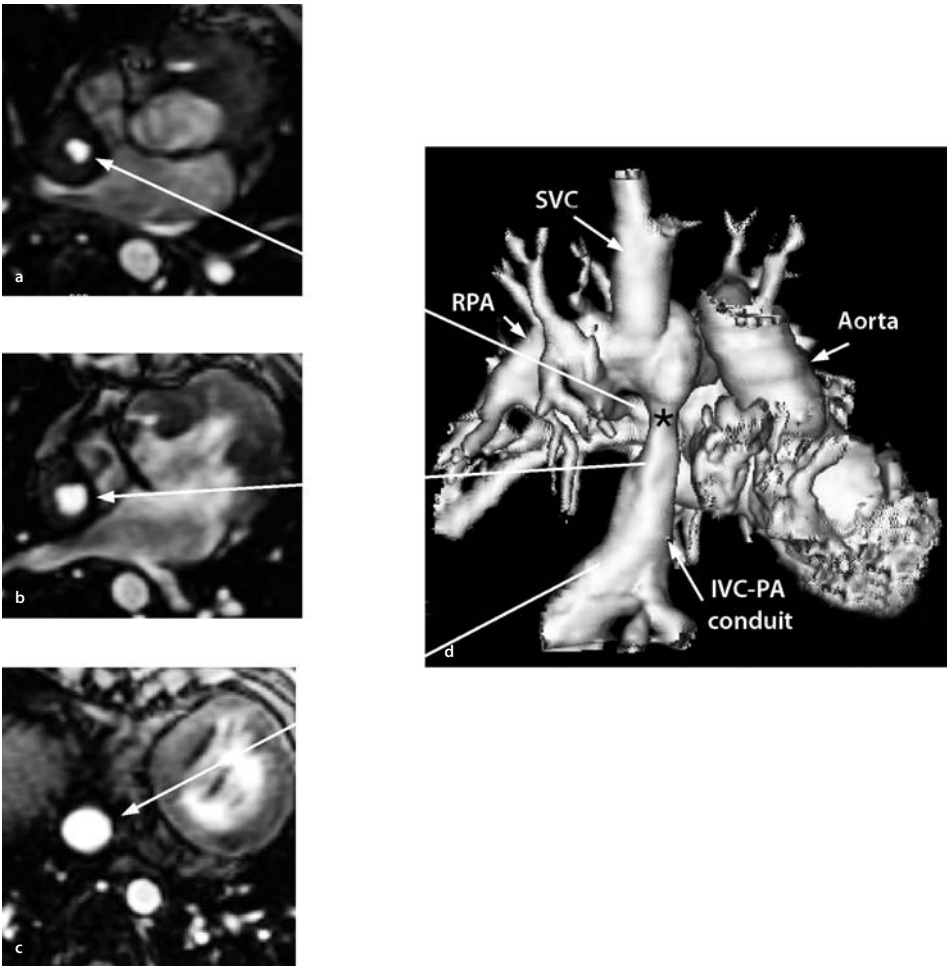

Fig. 3.0.6. Cardiac MRI. Axial time of flight images (**a**, **b**, and **c**) at different levels of the thorax in a patient after an extracardiac Fontan procedure. Arrows indicate the corresponding level to the *volume rendering reconstruction* (**d**) showing an unexpected narrowing of the extracardiac conduit between inferior vena cava and pulmonary artery (asterisk). Acquisition parameters were set as follows: flip angle 30°, trigger delay 400 ms; FOV 35×35 cm, fold over direction right-left in order to minimize respiratory artifacts, matrix: 256×160; slice thickness 3 mm; interpolated at 1.5 mm; NEX 2; TE 2.4 ms, TR 6.9 ms (*IVC* inferior vena cava, *PA* pulmonary artery, *RPA* right pulmonary artery, *SVC* superior vena cava)