Kawasaki Disease

Kawasaki disease is a generalized vasculitis of unknown etiology that occurs in small children and may eventually give rise to coronary vascular aneurysms. Subsequent thrombosis of these aneurysms and a reduction in blood flow may lead to myocardial infarction. The aneurysms often reach considerable size, and most are easily detected by MRCA.

Magnetic Resonance Angiography of the Great Vessels

K.-F. Kreitner

Although MRI has been used since its introduction in clinical practice for the investigation of flow effects, the development of clinically useful MR angiography techniques (MRA) did not begin until the mid-1980s. The first techniques to be developed were time-of-flight angiography and phase-contrast angiography. Used without intravenous contrast agents, they were the first techniques that permitted the selective visualization of flowing blood, making it possible to image the interior of vessels. However, they did not reach any clinical significance in the diagnosis of intrathoracic vascular diseases, because the intraluminal signal in both techniques is dependent on complex flow effects. Furthermore, respiratory movements and cardiac pulsations in the chest lead to motion artifacts, and the air-filled lung tissue induces susceptibility artifacts.

With advances in gradient technology, T1-weighted 3D sequences were developed that made it possible to acquire a complete dataset in less than 30s. These sequences could be acquired during a single breath hold. The heavy saturation of flowing spins requires the use of an effective T1-shortening contrast agent to produce an intraluminal signal that is very bright in relation to the background. These enhanced intraluminal signals are virtually independent of flow phenomena, making contrast-enhanced MRA somewhat similar to digital subtraction angiography (Fig. 6.41).

Fig. 6.39 a Unenhanced free-breathing MRCA demonstrates a stenosis in the proximal portion of the left coronary artery (arrow).

Fig. 6.40 a, b MRCA of the left coronary system in a patient with inflammatory signs and suspected Mediterranean fever shows no apparent abnormalities (a). Conventional angiography (b) confirms this finding (with kind permission of T. Ibrahim and R. M. Botner, Technical University of Munich).

Fig. 6.41 a, b Finding at the same location on conventional angiography (arrow) (with kind permission of T. Sommer, Bonn University Hospital).
Magnetic Resonance Imaging

Basic Technical Principles

Scanner Hardware and Software

Although MRA of the great thoracic vessels can be performed successfully at 1.0 T, 1.5 T is considered the standard field strength. Increasingly, 3.0-T magnets are becoming available for routine clinical imaging, and their use is currently being optimized for thoracic MRA.

The speed of an imaging sequence mainly depends on the performance of the gradient system. To achieve short acquisition times, it must be possible to produce magnetic field gradients of very high amplitude that can be switched rapidly on and off. Manufacturers’ specifications for maximum amplitudes range from 30 to 45 mT/m with rise times ranging between 100 and 200 mT/m/ms and a maximum field of view (FOV) between 40 and 53 cm. Another important parameter is the number of receiver channels that are available to receive the signal from a single surface coil element. While four receiver channels are sufficient for conventional thoracic imaging, additional channels must be available for parallel imaging techniques that are used in modern protocols.

Pulse Sequences

The standard pulse sequence for contrast-enhanced MRA is a spoiled three-dimensional (3D) T1-weighted gradient-echo sequence. It provides rapid imaging with acceptable resolution and coverage, enabling the images to be acquired during breath holds. The 3D acquisition provides high spatial resolution across the imaged plane with no breaks between the individual partitions. A “spoiled” sequence is one in which all residual transverse magnetization is destroyed before the next excitation. This increases the T1 weighting of the sequence and increases the contrast between the blood vessels and surrounding tissue.

The repetition time of the MRA sequence should be as short as possible, as this setting will directly affect the acquisition time (TA) of the sequence:

\[ TA = N_y \times N_z \times TR \]  

where \( N_y \) is the number of phase-encoding steps in the y direction, \( N_z \) is the number of phase-encoding steps in the z direction (number of partitions), and TR is the repetition time. The TR should be shorter than 5 ms to maintain a reasonable duration of breath holding. High-performance scanners can now achieve TR times of less than 2 ms. This makes it possible to expand anatomical coverage without lengthening the acquisition time or losing spatial resolution, or to improve spatial resolution while maintaining the same degree of coverage. When a very short TR is used, a 3D dataset can be acquired multiple times in succession, resulting in a time-resolved MRA study.

Analogously to the TR, the echo time TE should be as short as possible in MRA to minimize artifacts due to flow dephasing (e.g., in high-grade stenoses) and susceptibility differences (air–tissue interface in pulmonary angiography). Usually the TE is less than 2 ms, and high-performance scanners can achieve values less than 1 ms.

Various other techniques can be used to expedite data acquisition. The use of partial Fourier techniques is based on the fact that information contained in the upper half of the raw data matrix duplicates information in the lower half. This makes it possible to carry out a complete image reconstruction with equivalent contrast and resolution using only slightly more than 50% of the raw data, enabling us to modify equation (6.1) as follows:

\[ TA = N_y \times N_z \times TR \times 0.6 \]  

The cost of this shortcut is reflected in the signal-to-noise ratio, which is decreased by the square root of the time saved.

As an alternative, slightly more than half of the raw data matrix can be scanned in the frequency-encoding direction. This registers only a portion of the complete signal echo. Also known as “fractional echo” or “asymmetric echo,” this technique allows a primary reduction of TE, leading secondarily to a shortening of TR.

Zero filling is an interpolation algorithm for improving the apparent resolution of individual images. With a zero filling factor of 2, the raw data matrix is doubled, the new points at the periphery of the matrix are filled with zeroes, and the images are then subjected to Fourier transformation. The use of zero filling does not primarily affect image acquisition, but it prolongs the reconstruction time. When zero filling is done in all three spatial planes with a filling factor of 2, eight times as...
many voxels must be calculated as when interpolation is omitted. But if we assume that an imaging protocol provides sufficient diagnostic information without zero filling, we can use the zero filling strategy to reduce the number of necessary phase-encoding steps \((N_x \text{ or } N_z)\), which in turn will shorten the acquisition time.

A recent approach to accelerated data acquisition is parallel imaging. In this technique the sensitivity profile of the coil elements used for signal detection is utilized for spatial encoding, so that ultimately the number of necessary phase-encoding steps can be reduced at either the raw-data or image-data level. Prior to application, the sensitivity profile of the coil elements that can be integrated into the imaging sequence must be determined by calibration. An acceleration factor of 2 leads to an -48% reduction of acquisition time. The cost of this accelerated acquisition is reflected in the signal-to-noise ratio, which is decreased by a factor of \(1/\sqrt{\text{acceleration factor}}\) (Fig. 6.42b, c).

### Essential Point

The current standard magnetic field strength for contrast-enhanced MRA of the thoracic vessels is 1.5 T. The speed of data acquisition depends critically on the performance of the gradient system. The goal is to shorten the TR and TE times as much as possible. Partial Fourier techniques, zero filling, and parallel imaging have become established techniques for accelerating data acquisition. Parallel imaging requires multiple coil elements for signal detection and a corresponding number of interconnectable receiver channels.

### Techniques of Examination and Interpretation

#### Contrast Agents

The extracellular contrast agents that have been approved for clinical use are low-molecular hydrophilic gadolinium chelates. The agents that have been approved in Germany for MR angiography of the thoracic vessels are the open-chain complex Gd-DTPA (Magnevist, Bayer Schering Pharma), gadodiamide (Omniscan, GE Amersham Buchler), and the neutral macrocyclic agent gadobutrol (Gadovist, Bayer Schering Pharma). Gadobutrol is characterized by its double concentration of gadolinium compared with other extracellular contrast agents (1.0 mmol/L), so that only half as much needs to be injected to achieve an equal dose.

The use of extracellular contrast agents temporarily shortens the T1 relaxation time of the blood to values between 30 and 80 ms.

Original dose recommendations for MRA of the thoracic vessels were in the range 0.2–0.3 mmol/kg bw or a total dose of 40–60 mL. But with current acquisition times of 15–25 s it is sufficient to administer smaller doses in the range 0.1–0.15 mmol/kg bw, or a total dose of 20 mL.

Considerable interest has focused on the development of "blood-pool" contrast agents that remain within the blood vessels for an extended period and cause little or no enhancement of extravascular tissues. In MRA of the great vessels, these blood-pool agents could make it possible to examine various regions without the need for additional contrast injection. They
would be particularly useful in the quantification of cardiac perfusion and imaging of the coronary arteries (see p. 111 ff)

**Bolus Timing**

For successful contrast-enhanced MRA of the thoracic vessels, acquisition of the 3D dataset should coincide with the arrival of the contrast bolus in the vessels of interest.\(^{140,141,150}\)

Various methods have been described for the optimum timing of contrast-enhanced MRA. In the test bolus method, a small initial bolus is administered to determine the physiological transit time of the contrast agent from injection to its appearance in the ROI so that MR data acquisition can be initiated precisely when the contrast bolus arrives in the target region (Figs. 6.41, 6.42).\(^{152}\) There are also techniques in which continuous SI measurements are taken to time the arrival of the contrast bolus in a predefined test region. When the rise in SI per unit time exceeds a designated threshold, MRA is automatically initiated (e.g., Smart Prep, SmartScan). Another approach is the MR fluoroscopic detection of arrival of the contrast bolus in the target region with manual initiation of the MRA sequence (e.g., CareBolus).

Both methods involve centric rather than linear k-space acquisition, because ideally MRA is initiated to coincide with the peak concentration of gadolinium in the desired vessel. Large clinical studies have confirmed the reliability of these techniques in ensuring diagnostic image quality.\(^{146,150,153}\) A major advantage of the test bolus method is that it can be used in any MR system and does not require special hardware or software.

The imprecise timing of contrast-enhanced MRA can affect image quality in various ways.\(^{154}\) If the contrast bolus appears too early in the ROI relative to acquisition of the 3D dataset, the contrast agent will already have washed out by the time imaging is initiated, resulting in the undesired enhancement of veins and/or other vascular and tissue structures.

If the contrast bolus arrives too late, little or no contrast agent will be present in the vascular region of interest. “High-pass filter” artifacts may be observed; these occur when the TI time of the blood changes rapidly because of contrast wash-in during acquisition of the central k-space lines, which determine image contrast. This results in poor visualization and enhancement of the target vessels, with or without “ringing” artifacts along the vessel walls, making it difficult to detect abnormalities.

**Essential Point**

For successful contrast-enhanced MRA of the thoracic vessels, acquisition of the 3D dataset should coincide with the arrival of the contrast bolus in the vessels of interest. Strategies for bolus timing include the test bolus method as well as manufacturer-specific automated or semi-automated techniques. The main advantage of the test bolus method is that it does not require special hardware or software.

**Planning the Examination**

The pulmonary circulation can be imaged in one coronal or two sagittal slice packages\(^ {151,155}\). In the coronal acquisition, both halves of the lung are imaged simultaneously. Generally, a larger field of view is needed to prevent wrap-around artifacts from the shoulders and apposed arms, and this adversely affects the voxel size of the 3D dataset. Another option is to extend the arms above the head before imaging. The coronal technique is satisfactory for evaluating abnormalities of the pulmonary veins, but a 3D volume 12–14 cm thick cannot cover the entire pulmonary tree, and the voxel size remains anisotropic even when a high-end system is used. These restrictions may be overcome by use of multichannel phased-array coils, parallel imaging techniques, and imaging at 3T: here, whole coverage of the lung is realizable with acquisition of isotropic voxel sizes of 1 mm × 1 mm × 1 mm in a total of 20 s.\(^ {144}\)

Sagittal data acquisition permits the use of smaller FOVs, which has a favorable effect on voxel size and makes it easier to acquire isotropic voxels. Because of the smaller 3D volume, the acquisition time is shorter than with coronal data acquisition. The main disadvantage of sagittal acquisition is the fact that separate datasets must be acquired for each side, which doubles the required contrast dose\(^ {151,155–157}\) (Fig. 6.43).

An oblique sagittal plane is usually recommended for imaging the thoracic aorta. Generally the prescription of this plane is based on the orientation of the aortic arch in the transverse plane. Coronal data acquisition is recommended in cases where it is necessary to investigate the supra-aortic branch vessels or detect congenital anomalies of the aortic arch.\(^ {158–160}\) ECG triggering is not absolutely essential but does improve visualization of the ascending aorta and is therefore recommended in examinations for ascending aortic disease.\(^ {150}\)

In time-resolved contrast-enhanced MRA, the time needed to acquire a 3D dataset can be shortened to less than 4 s, which is a particular advantage in severely dyspneic patients. With the advent of multichannel coil technology, receiver channels and modified strategies for k-space sampling, the reduction of in-plane resolution, using thicker partitions, and decrease of the number of partitions may be minimized.\(^ {139,143,149,161}\) With time-resolved examination techniques, higher injection rates (up to 6 mL/s) should be used with a concomitant reduction in the contrast dose\(^ {140}\) (Fig. 6.44). Time-resolved imaging techniques can be useful in congenital vascular malformations (e.g., duc tus arteriosus) and aortic dissections (perfusion characteristics of the true and false lumina), and to document steal effects in patients with subclavian artery stenosis.\(^ {145,148,162}\)

The recommended protocol for contrast-enhanced 3D MRA of the thoracic vessels is outlined in Table 6.8.

**Essential Point**

With acquisition times from 10 to 20 s, a contrast dose of 20 mL or 0.1–0.15 mmol/kg bw is generally adequate. The shorter the acquisition time, the higher the recommended injection rate (up to 4 mL/s). A rate up to 6 mL/s is recommended for time-resolved MRA.

**Interpretation**

The interpretation of thoracic MRA is based on a detailed analysis of the source images. Combined with multiplanar reor-
Congenital Heart Disease in Adults

Congenital cardiovascular anomalies are present in 0.8–1% of all newborns. The number of different anomalies is so large that their complete description would be beyond the scope of this book. This chapter therefore focuses on the capabilities of modern cardiac imaging techniques in the most common types of congenital heart disease in adults—atrial septal defects, patent foramen ovale, and ventricular septal defects.

Atrial Septal Defect

Anatomy and Pathophysiology

Atrial septal defects (ASDs) account for ~10% of all congenital heart diseases and for 22–40% of congenital heart disease in adults. They are associated with varying degrees of left-to-right shunting of arterialized blood into the pulmonary circulation, depending on the size of the septal defect and the relative pressures. In extreme cases the shunt flow may be several times the volume flow of that in the systemic circulation. Atrial septal defects are primarily characterized by a volume overload on the right heart, which eventually lead to cardiac failure. With a left-to-right shunt above the level of the tricuspid valve, the great dilatory capacity of the pulmonary vessels can forestall a pressure rise in the pulmonary artery and right ventricle. Comparable to the ventricular septal defect however, a functional and/or organic rise in vascular resistance will develop over time in the pulmonary circulation, causing the pressure in the right ventricle to rise. When the pulmonary vascular resistance exceeds that of the systemic circulation in the presence of a ventricular or atrial septal defect, a "shunt reversal" occurs, leading to marked cyanosis. This phenomenon is termed the Eisenmenger reaction.

Three etiological types of atrial septal defect are distinguished:

- An ostium secundum atrial septal defect (ASD II) is located in the central portion of the atrial septum in the region of the fossa ovalis. It is the most common type, accounting for 60–70% of atrial septal defects.
- An ostium primum atrial septal defect (ASD I) is usually a large defect that comprises 15–25% of atrial septal defects. It results from a failure of fusion of the septum primum with the endocardial cushion between the atrioventricular valves. As a result, it is commonly associated with mitral valve defects and occasionally with tricuspid valve defects.
- A sinus venosus atrial septal defect is present in 5–15% of cases. It occurs in the posterosuperior portion of the atrial septum between the termination of the superior vena cava and the fossa ovalis. It is frequently associated with anastomotic termination of the right pulmonary veins in the right atrium.

Clinical Features

Symptoms

- Generally asymptomatic until the third decade; 70% of patients are symptomatic by the fifth decade
- Dyspnea, rapid fatigability (manifestations of heart failure).
- Palpitations (in patients with atrial arrhythmias)
- Proneness to respiratory infections
- Approximately 15% of patients show clinical symptoms of associated mitral insufficiency

Complications

- Atrial fibrillation or flutter
- Mitral insufficiency

Imaging

Echocardiography

2D Echocardiography

- Transthoracic scanning enables direct visualization of the septal defect (excepting sinus venosus defects) [Fig. 7.1].
- Transesophageal scanning permits direct visualization of a sinus venosus defect [Fig. 7.2].
- Enlargement of the right atrium and right ventricle
- Detection of associated congenital anomalies

3D Echocardiography

- Enables direct visualization of the ASD in the frontal view to assess the location, size, and shape of the defect.
- Allows direct visualization of a cleft in the anterior mitral valve leaflet in ASD I.
- Can detect an AV canal in patients with ASD I.
- Detection of an AV canal in patients with ASD I
- Detection of associated mitral insufficiency in ASD I

Color Doppler Echocardiography

- Detection of shunt flow by demonstrating a flow jet through the atrial septum into the right atrium
- Detection of shunt flow by demonstrating a flow jet through the atrial septum into the right atrium
- Detection of an AV canal in patients with ASD I
- Detection of associated mitral insufficiency in ASD I

Contrast Echocardiography

- Detection of a left-to-right shunt based on the washout phenomenon in the contrast-filled right atrium [Fig. 7.1b].
**CW Doppler Echocardiography**

- Pulmonary hypertension can be diagnosed by assessing the systolic pulmonary artery pressure based on the regurgitant signal from the tricuspid valve.

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**Magnetic Resonance Imaging and Computed Tomography**

The standard planes for detecting an ASD in MRI are horizontal long-axis slices and short-axis slices through the heart. Spoiled gradient-echo sequences are preferred. They are of advantage over SE and balanced SSFP sequences in that they can also detect smaller defects by the associated zones of turbulent flow, for example.

Determination of the maximum size of the ASD is essential in selecting patients for interventional repairs, an unsuitable procedure for large defects. Both MRI and multislice CT (MSCT) can document the enlargement of the right atrium and right ventricle (Fig. 7.3a). In addition to demonstrating morphologi-

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**Essential Point**

In the presence of clinical suspicion of a left-to-right shunt at the atrial level, it is possible to overlook a high-sited sinus venosus type of ASD if the atrial septum appears normal. Often a sinus venosus defect can be detected only by multiplanar transesophageal echocardiography in color Doppler mode.

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**Fig. 7.1 a, b** Transesophageal echocardiography of ASD II (arrow).

- a Prior to contrast wash-in.

- b Typical appearance of the washout phenomenon (arrow) in a left-to-right shunt at the atrial level. Various bubbles can be seen entering the left atrium after passing through the shunt.

**Fig. 7.2 a, b** Sinus venosus atrial septal defect documented by transesophageal echocardiography.

- a The size of the defect (arrow) is measured at the entry of the superior vena cava into the right atrium.

- b Abnormal flow signal detected by color Doppler echocardiography.
cal features, MRI also enables an accurate assessment of the $Q_p/Q_s$ ratio based on flow measurements in the ascending aorta and pulmonary trunk. This ratio is important in selecting patients for operative or interventional repair. The $Q_p/Q_s$ ratio can also be determined using cine MRI to identify right and left ventricular stroke volumes, provided the presence of an additional significant valvular defect can be excluded. MRI and CT additionally detect any anomalous pulmonary venous termination that may be present. Operative treatment is the only option available for correcting this anomaly² (Fig. 7.3b).

### Patent Foramen Ovale

#### Anatomy and Pathophysiology

Patent foramen ovale (PFO) is a valvelike opening that persists postnatally between the septum primum and secundum in the region of the fossa ovalis owing to failed fusion of these septal elements. A right-to-left shunt at the atrial level may occur spontaneously or in response to a Valsalva maneuver. Autopsy results indicate that PFO has a prevalence of ~25% in the general population. Its clinical significance is that it places patients at risk for paradoxical emboli. This risk appears to be particularly high in patients with a hypermobile atrial septal aneurysm, which frequently coexists with PFO. Because the opening and volume of the shunt are generally small, hemodynamic complications do not occur.

#### Clinical Features

**Symptoms**
- The majority of patients with PFO are asymptomatic.

**Complications**
- Central or peripheral emboli, stroke, transient ischemic attacks, dizzy spells, migraines

#### Imaging

**Echocardiography**

**M-Mode Echocardiography**
- M-mode can document hypermobile excursions associated with an atrial septal aneurysm (Fig. 7.4a).

**2D Echocardiography**
- Demonstrates the valvelike separation between the septum primum and secundum; but cannot reliably detect patent foramen ovale (2D visualization only by transesophageal scanning).
- Can detect an atrial septal aneurysm (frequently also detectable by transthoracic scanning).

**Contrast Echocardiography**
- Can reliably detect or exclude patent foramen ovale based on the right-to-left passage of contrast medium, either spontaneously or in response to a Valsalva maneuver (Fig. 7.4b).

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**Fig. 7.3a, b** Large ASD II in a 32-year-old woman.

- a Cine image in the four-chamber view demonstrates the large ASD (arrow).
- b Contrast-enhanced 3D MRA further demonstrates partial anomalous pulmonary venous termination on the right side. The upper and middle lobe veins (arrows) open into the superior vena cava.