7.9 Diagnosis of Congenital Heart Disease in Adults and Children

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7.9.1 Introduction

An accurate, 3D evaluation of the cardiac and related arterial anatomy is critical for the clinical management of adult and pediatric patients with complex congenital heart disease. 3D imaging has to be able to demonstrate the shapes of, and spatial relationships between, the great arteries, proximal branch pulmonary arteries, and anomalous pulmonary venous or systemic connections. Three-dimensional information about extra-cardiac morphological characteristics may determine subsequent surgical intervention.

Magnetic resonance imaging has been recommended by a task force report as the first-choice technique for many congenital heart diseases (Association of European Pediatric Cardiologists 1998). MRI appears, on initial consideration, to be an ideal technique because there is no radiation burden, which is a substantial advantage, especially in neonates and young children. However, it also suffers various limitations: the major one being the need for prolonged sedation and close monitoring, especially of infants with cyanotic heart disease, whose condition is often unstable. For such patients, intensive-care pediatricians must be present during the MRI examination. Also, the spatial resolution of MR images is lower than that of CT images, which can be a significant drawback for visualization of small anatomical structures.

More recently, helical CT has been proposed for 3D anatomical visualization in patients with congenital heart disease (Kawano 2000). Helical technology allows volume acquisition in a short period of time and provides good-quality 3D vascular images, even for neonates and infants. The multi-slice CT technology now available has much faster acquisition times , which substantially reduces respiratory artifacts. Furthermore, image synchronization with the cardiac rhythm is now possible, and this should reduce problems associated with heart motion. In our surgical center, which specializes in congenital heart disease, multi-slice CT with the evolution from 4- to 16- and, very recently, 64-slice technology has rapidly become an important complementary imaging technique for both pre- and post-operative management of patients (Lee 2004).

7.9.2 Technical Aspects and Imaging Protocols

The first issue is whether ECG-gated acquisition should be used for congenital heart disease patients, and the second concerns optimization of radiation exposure.

7.9.2.1 Neonates and Young Infants

It is not possible to make neonates and young infants hold their breath. Consequently, we do not use ECG-gated techniques for acquisition, because respira-

tory artifacts greatly degrade images, and such artifacts are substantial with ECG-gated acquisition (because acquisition is slow) (Paul 2002). Similarly, the cardiac rhythm of babies with cyanotic congenital heart disease is very high, generally between 140 and 180 bpm, making cardiac motion-free images impossible. The third reason for not using retrospectively ECG-gated cardiac acquisition is that it requires a higher radiation dose than non-ECG-gated thoracic CT, because only a part of the dose (i.e., the dose delivered during diastole) is used for creating images. Organ sensitivity to radiation is much higher in babies than in adults. Indeed, recent reports suggest that the risk of developing cancer in the future cannot be totally ruled out (Brenner 2001) after medical radiation exposure. This risk increases with radiation dose.

In our experience, the principle of "going as fast as possible" still allows good image quality in neonates with congenital heart disease; furthermore, the short acquisition times minimize respiratory artifacts. With 4-slice CT, the thorax of babies can be scanned in 3–4 s using 2.5-mm collimation, 0.5-s rotation time, and a table-feed of 20 mm per s (pitch 2). The images are of higher quality than those obtained using a slice thickness of 1 mm, which requires longer acquisition times associated with thinner collimation, and thus more respiratory artifacts. Very short acquisition times (≤ 5 s) allow apnea in intubated babies, and the images obtained are free of respiratory artifacts. With 16-slice CT, the thorax of a baby can be scanned in about 4 s using 0.75-mm slices or in 2 s using 1.5-mm collimation.

7.9.2.2 Infants Over 7 Years of Age and Adults

There are two options for older infants and young adults: either conventional breath-hold angio-CT acquisition or ECG-gated acquisition. The protocol should be chosen according to clinical considerations. If, for example, coronary visualization is required to detect a possible anatomical variant, ECG-gated protocols are recommended. ECG gating is often not required in other cases and should only be applied when appropriate due to the related increase in radiation dose.

7.9.2.3 Dose Considerations

Radiation exposure is a major public-health issue. CT contributes greatly to the population dose due to medical exposure, as it makes up 35% of the total dose delivered during diagnostic examinations although it represents only 4% of such examinations (Nagel 2002). The ALARA principle (as low as reasonably achievable) is a good rule of thumb: dose reduction is necessary but examination quality must be maintained without losing diagnostic information. While the thorax is a region of low-attenuation, substantial dose reduction during chest CT is feasible because of the high inherent contrast. In August 2001, the ALARA conference of the Society for Pediatric Radiology considered the issue of dose reduction by decreasing the kilovoltage (Slovis 2002). In our center, we decided to apply the ALARA principle as far as possible to neonates and babies with congenital heart disease, and then implement some systematic rules:

- No topogram (responsible for unnecessary additional radiation dose)
- Consistent use of 80 kV settings whenever feasible
- Adaptation of the mAs to the child's weight (starting from 17 mAs)
- Only one phase acquisition when possible
- Systematic protection of non-scanned organs

A setting of 80 kV for pediatric patients is the rule in our center. Reducing the kV from 120 to 80 kV decreases the radiation dose by 65% at constant tube-current setting, as radiation dose varies with the square of the kV. This setting is sufficient for good-quality images, as long as the mAs are adjusted according to the child's weight. The other advantage of using only 80 kV is that the amount of contrast medium injected can also be reduced – because low kV is more sensitive to contrast (iodine has a high atomic number) than higher settings.

The tube-current is adapted to body weight for neonates and infants. For example for thoracic imaging, we scan neonates using 17–30 mAs and 80 kV, babies using 30–45 mAs. The minimum exposure setting allowed with the 16-slice CT used in our institution is 80 kV and 17 mAs using

0.75-mm collimation and a table-feed of 18 mm/s. Previous studies suggested that CT delivers much lower radiation doses than conventional angiography (Westra 2002). Appropriate exploitation of anatomical data acquired from CT may therefore be used to limit the number of views acquired with angiography, and sometimes replace conventional angiography altogether. Thus, CT may allow total radiation exposure of congenital heart disease patients to be reduced.

7.9.2.4 Contrast-Injection Protocol

Neonates and babies. The injection dose must be adapted to the baby's weight. We currently use 2 cc per kg. At 80 kV, the rate of injection can be as low as 0.5 cc per s in neonates through a catheter placed in the vein of the hand. Higher rates may be used with a central catheter (femoral or jugular). We use a power injector to ensure a continuous and regular flow rate, and the rate of injection is 0.5 cc/kg to 1 cc/s depending on the site of injection. The start delay for neonates and infants is 15 s for peripheral injection, and 10 s for central venous injection. To ensure vascular contrast during the acquisition, we increase the amount of contrast medium in some cases according to the rule: Time of injection = start delay + time of acquisition. Accordingly, acquisition is never "too late" for good vascular enhancement, because acquisition ends with the end of injection, so the peripheral veins still contain contrast medium when acquisition ends.

Precautions for venous access. Peripheral venous access is always done in the pediatric unit. Injection in the right arm is preferable (but not obligatory) to avoid artifacts in the innominate left brachiocephalic vein. In some cases, venous connections are congenitally abnormal or surgically modified. Any available relevant information can be important before the scan procedure, as the scan injection protocol may have to be adapted accordingly. Venous visualization may be obtained at first pass, with a high concentration of contrast medium, or sometimes later, at the time of venous return. The optimal injection protocol depends on the particular venous anatomy. The catheter is tested for permeability before the injection. It is essential to avoid any air injection during the scan procedure. All bubbles should be removed when connecting the catheter to the power injector. Because many patients with congenital heart disease have right-to left-shunt, air injection through venous access could cause systemic air embolism, with possible fatal consequences.

7.9.2.5 Sedation of Infants

General anesthesia is never necessary in our experience. In addition, we do not administer any sedative drugs to neonates . For infants, we recommend oral or intra-rectal sedation (or both) before the CT procedure in order to prevent agitation during the acquisition, which may result in poorer image quality and, as a consequence, occasionally, re-examination. Sedation is not always necessary if the baby is quiet. Experienced technicians are required in the CT room for good management of the babies: experience and knowledge of baby management and a calm attitude are important. Our sedation protocol for infants includes intra-rectal administration of 0.3 mg/kg Midazomal 15 min before examination. Additional sedative drugs may be useful (1 mg/kg hydroxyzine, per os, 1 h before examination). With experienced technicians, the mean total examination time in the CT room is 20 min. Qualified medical monitoring may be sometimes necessary during the examination, depending on the clinical condition of the baby, and oxygen saturation should be closely monitored.

7.9.2.6 Post-processing

Even if all information is available on axial CT images, 3D imaging with VRT is our first-line approach for interpretation due to the complexity of spatial variations of anatomical structures. VRT allows initial comprehensive imaging of anomalies whereas MIP images are used subsequently for vessel-by-vessel interpretation.

7.9.3 Clinical Indications

7.9.3.1 Pulmonary Arteries

Pulmonary artery evaluation is frequently required for patients with pulmonary atresia with ventricular septal defect, tetralogy of Fallot, truncus arteriosus, or suspicion of pulmonary sling. With a 16-slice CT, we usually use 0.75-mm collimation and obtain 1-mm slice width with an increment of 0.5 mm. High resolution is beneficial for evaluating pulmonary artery stenosis.

7.9.3.2 Coronary Arteries

Anomalous coronary arteries are frequently associated with congenital heart disease. The most frequent anomalous finding is a left coronary artery originating from the right coronary sinus, but many variants are possible, even the coronary artery originating from pulmonary arteries. The "normal" position of coronary origins may be different from usual; for example, in cases of tetralogy of Fallot, because of the rotation of the aorta: the origin of the LMA is typically at 6 o'clock and that of the RCA at 1 o'clock. Detection of an anomalous origin of the coronaries is especially important before surgery when a ventriculotomy is planned, as accidental lesion during intervention of the coronary artery crossing the right ventricle can be fatal.

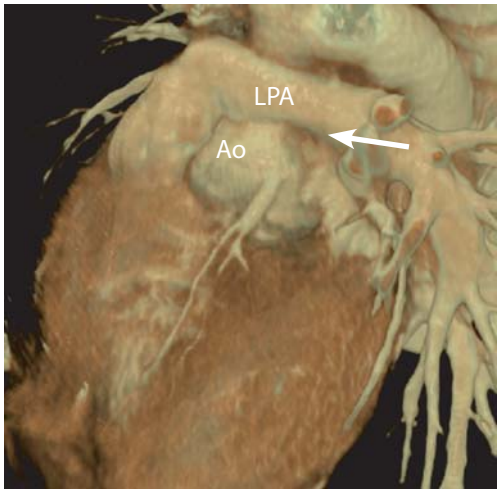


Fig. 7.64. 4-slice CT evaluation of re-implanted coronary arteries in a 9-year old girl, who underwent surgery just after birth for transposition of the great vessels (arterial switch). The 4-slice CT shows the normal anatomical position of the coronary arteries 9 years after surgery. Note the left pulmonary artery crossing in front of the aorta. *Ao* Aorta, *LPA* left pulmonary artery

In older patients with congenital heart disease, if the patient can hold his or her breath for a sufficiently long time, ECG-gated acquisition may be the technique of choice. Free-motion artifact visualization is then possible, allowing accurate evaluation of the coronary artery tree (Fig. 7.64). To avoid heart-motion artifacts, the heart rate must be regular and, if possible, < 70 bpm; excellent results are generally obtained at 55–60 bpm. We use an ECG pulsing technique except in patients with arrhythmia, so as to reduce the radiation dose by about 40%, all others factors being equal.

7.9.3.3 Aorta and Collaterals

Evaluation of the aortic anatomy is essential in cases of aortic coarctation (Fig. 7.65) or suspicion of aortic arch anomalies. In patients with pulmonary atresia with ventricular septal defect, major aorto-pulmonary collateral arteries (MAPCA) often originate from the origin of the descending aorta. It is essential to determine the size and spatial relationship of these arteries when planning surgical intervention. Thin collimation (1 mm with a 4-slice CT, 0.75 mm with a 16-slice CT), is of value as it provides high-resolution images. Such images are, in turn, useful for evaluating aortic stenosis, especially in patients with aortic coarctation, for better assessment of vessel narrowing.

7.9.3.4 Upper-Airways Compression of Vascular Origin

Central-airway compression of vascular origin can result from various situations. The most frequently observed are: aortic arch anomalies (Fig. 7.66), pulmonary artery sling, dilated pulmonary arteries, and posteriorly displaced aorta (switch intervention). Non-enhanced CT is sufficient to detect steno-

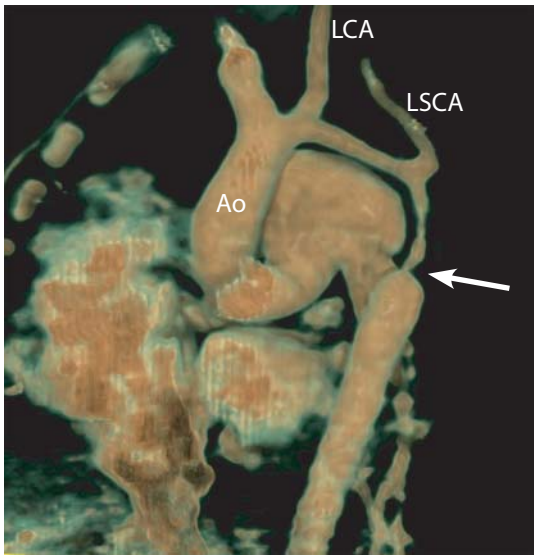
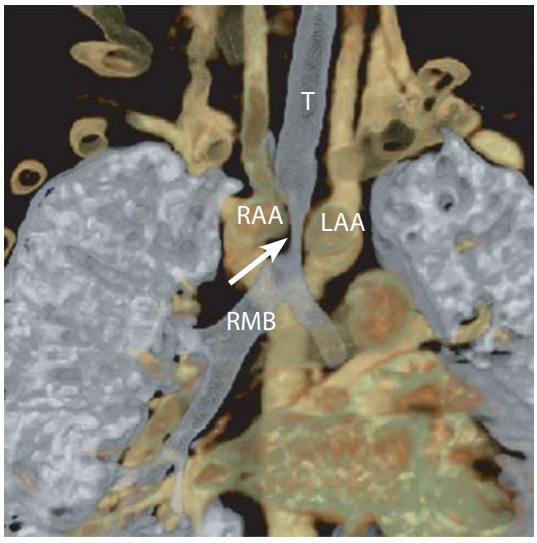


Fig. 7.65. Aortic coarctation in a 1-month-old baby examined with 16-slice CT. Total interruption of aortic arch was suspected from echocardiography. The 16-slice CT showed long stenosis of the aorta with post-ductal severe narrowing (*arrow*). *LCA* Left carotid artery, *LSCA* left subclavian artery

sis of the central airways, but contrast enhancement is required to identify the vascular origin. In babies, we currently use the same protocol as for visualization of the pulmonary arteries or aorta. In addition to MIP reconstructions, VRT is very effective for showing central-airway narrowing (Fig. 7.66b). However, airway compression can be more distal, which makes diagnosis of vascular compression difficult. Evidence of air trapping on parenchymal windowing is indicative of this type of compression.

7.9.3.5 Anomalous Venous Return

Multi-slice CT is very effective in the detection of pulmonary or systemic anomalous venous return. ECG-gated acquisitions are usually not necessary because venous structures are not very sensitive to cardiac motion (Fig. 7.67). The injection site and timing of acquisition must be chosen carefully, since the timing at which opacification is best depends on the venous drainage, and any anomalous venous drainage may affect optimal timing. Additional delayed acquisition may be necessary to opacify the entire venous system. To avoid artifacts associated



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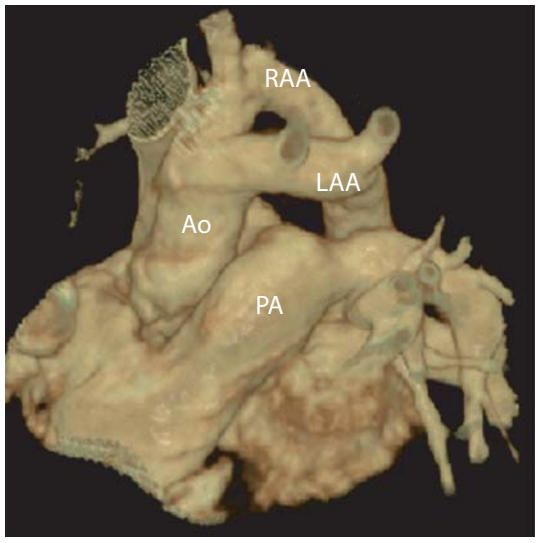

b

Fig. 7.66a,b. Severe respiratory distress in a newborn. **a** The 16-slice CT examination using VRT clearly shows a complete double aortic arch responsible for tracheal compression in a 5-month-old patient weighing 5 kg. **b** Using the same data set, VRT (displaying both airways and vascular structures) clearly reveals compression of the trachea (*T*) by the right aortic arch (*RAA*). *LAA* Left aortic arch, *Ao* ascending aorta, *PA* pulmonary artery

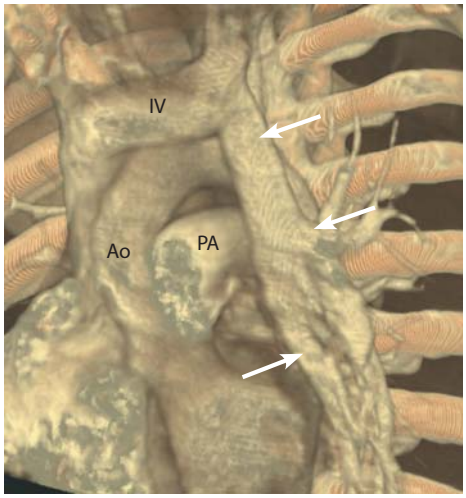

Fig. 7.67. 16-slice CT image of totally abnormal left venous return in a 25-year-old man with dyspnea. All left pulmonary veins (*arrows*) are connected to the innominate vein. *IV* Innominate vein

with concentration of the contrast medium in the veins, a low rate of injection is recommended.

7.9.3.6 Post-intervention Evaluation

Post-operative evaluations are required in various clinical situations, for example, for assessment of bypass patency (Fig. 7.68) or suspicion of mediastinitis. The CT protocol should be adapted to the clinical context. In many cases, radiation exposure can be less than that of the standard protocol because there is generally no need for detailed anatomic information. For example, to search for mediastinitis, a single, delayed (3–5 min) acquisition may be sufficient. In contrast, conduit patency can be tested by acquisition at the arterial phase alone. CT assessments of the altered vascular anatomy may also be useful for follow-up after complex surgical repair.

7.9.4 Improvements with 64-Slice CT

With the recently developed 64-slice CT scanners, the thorax of a baby can be scanned in 1–2 s with the thinnest collimation, usually 0.6 mm (Fig. 7.69). Therefore, no compromise in spatial resolution has to be made even at the minimum scan time. The faster rotation time of the new scanners, down to 0.33 s, improves image quality also at higher heart rates, and heart rate control is no longer mandatory in adult patients. The increased spatial resolution of 0.4 mm that is possible with 64-slice CT even allows for visualization of the coronary arteries in babies, thus enabling the detection of congenital coronary artery disease at a very early age (Fig. 7.70). In older children and adults, the increased volume-coverage speed provides very comfortable breath-hold times of 5–10 s, during which the thorax can be covered with ECG gating at the maximum possible resolution (Fig. 7.71).

7.9.5 Conclusion

Precise 3D visualization of anomalous extra-cardiac anatomy in congenital heart disease patients can be routinely obtained using multi-slice CT. In addition to providing a major, additional, non-invasive diagnostic tool for the evaluation of congenital heart disease, multi-slice CTA offers an alternative to angiography. While radiation-dose considerations, especially for neonates and infants, remain a draw-

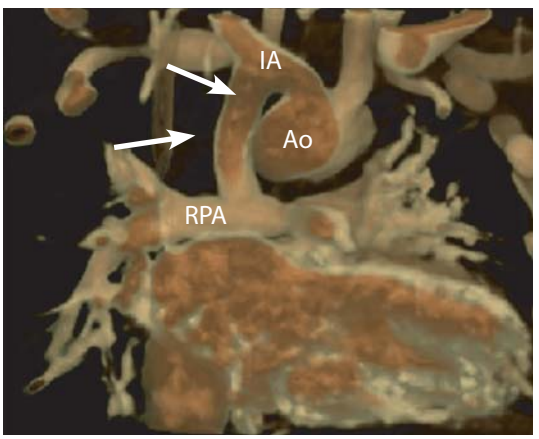

Fig. 7.68. Post-operative evaluation in a newborn with pulmonary atresia and ventricular defect, examined with 16-slice CT. A shunt between the innominate artery and the right pulmonary artery (Blalock anastomosis) is clearly apparent (*arrows*). *RPA* Right pulmonary artery, *IA* innominate artery

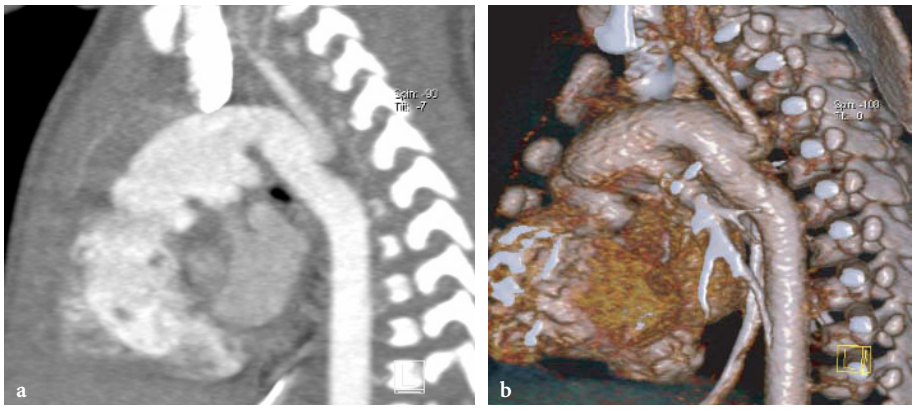

Fig. 7.69a,b. 64-slice CT examination of a neonate. The image is displayed in MIP (a) and VRT (b). With a 0.33-s rotation time, the thorax of the neonate is covered in less than 2 s

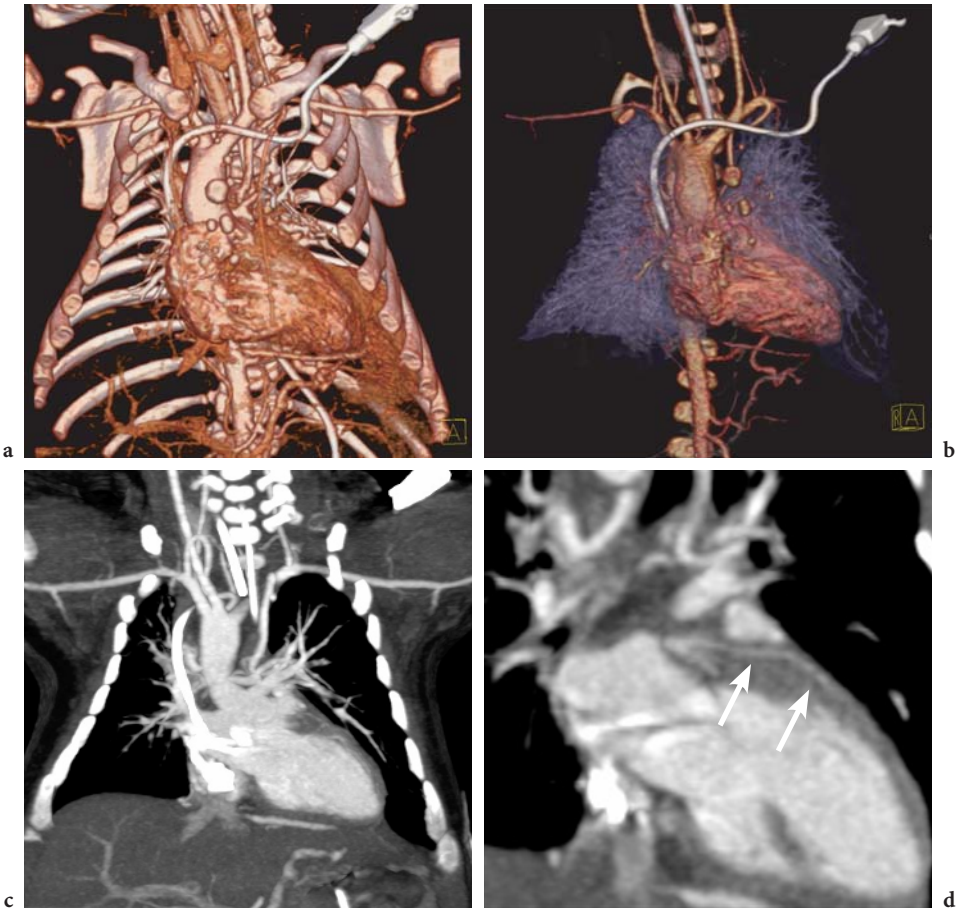

Fig. 7.70a–d. ECG-gated 64-slice CT examination of an 8-week-old baby. The cardiothoracic anatomy can be visualized with VRT (a, b) and MPR (c). Owing to the high spatial resolution, an abnormal course of the coronary artery could be ruled-out (*arrows* in d). (Case courtesy of Tübingen University, Germany)

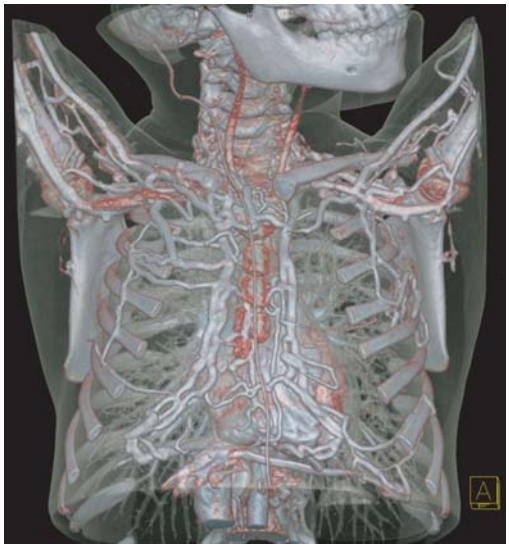

Fig. 7.71. 64-slice CT examination of an 8-year-old child with complex malformation of the thoracic vasculature. The scan was acquired in a 6-s breath-hold with 80 kV and without use of ECG-gating to minimize radiation exposure. (Case courtesy of MUSC, Charleston, USA)

back of this technique, excellent image quality is possible even at very low exposure.

References

Brenner D, Elliston C, Hall E, Berdon W (2001). Estimated risks of radiation-induced fatal cancer from pediatric CT. AJR Am J Roentgenol 176:289–96

Kawano T, Ishii M, Takagi J, Maeno Y, Eto G, Sugahara Y, Toshima T, Yasunaga H, Kawara T, Todo K, Kato H (2000). Three-dimensional helical computed tomographic angiography in neonates and infants with complex congenital heart disease. Am Heart J 139:654–660

Lee EY, Siegel MJ, Sierra LM, Foglia RP (2004). Evaluation of angioarchitecture of pulmonary sequestration in pediatric patients using 3D MDCT Angiography. AJR 183:183–188

Nagel HD et al (2002). In: Radiation exposure in computed tomography: Fundamentals, influencing parameters, dose assessment, optimization, scanner data, terminology. 2nd edn. Offizin Paul Hartung Druck, Hamburg, Germany pp. 1–3

Paul JF, Serraf A (2002). Truncus arteriosus and double aortic arch. Circulation 105:e170

Slovis TL (2002). ALARA conference proceedings. The ALARA concept in paediatric CT: intelligent dose reduction (Editorial). Pediatr Radiol 32:217–231

Task Force of the European Society of Cardiology, in collaboration with the Association of European Paediatric Cardiologists (1998). The clinical role of magnetic resonance in cardiovascular disease. Eur Heart J 19:19–39

Westra SJ, Hurteau J, Galindo A, McNitt-Gray MF, Boechat MI, Laks H (1999). Cardiac electron-beam CT in children undergoing surgical repair for pulmonary atresia. Radiology 213:502–512