Anomalies of the Systemic and Pulmonary Arteries

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L. Grosse-Wortmann, M.D., F.R.C.P.S.C. Section Head, Cardiovascular Magnetic Resonance, The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada Congenital abnormalities of the thoracic systemic and pulmonary arteries can occur in isolation or in the setting of a complex cardiovascular malformation. Severe or complex vascular defects are usually diagnosed and corrected in childhood. Infrequently, anomalies of the systemic and pulmonary arteries present during adulthood. After correction of a cardiovascular malformation most children survive into adulthood. In adults, the most common indication for imaging is to evaluate post-treatment sequelae. Understanding the underlying pathology and possible complications that may occur during follow-up is essential for planning imaging studies. Furthermore, knowledge on the different imaging modalities, including their strengths and limitations for assessing anomalies of the systemic and pulmonary arteries, is important.

This chapter discusses the role of the different imaging modalities for the assessment of congenital abnormalities of the thoracic systemic and pulmonary arteries both at diagnosis and during follow-up after treatment. As follow-up already starts directly after surgical correction this chapter includes images of both children and adults.

Overview of Imaging Techniques for the Systemic and Pulmonary Arteries

Chest radiography is usually the first imaging modality used in adults with suspected large vessel abnormalities. Chest radiographs may provide clues to abnormalities of the thoracic vascular system. Signs that should raise suspicion of a vascular abnormality include abnormal location of the aortic knob, tracheal narrowing, rib notching, or an abnormal mediastinal silhouette (Ferguson et al. 2007).

Echocardiography is usually the next noninvasive imaging tool. In patients with favourable acoustic windows, echocardiography allows evaluation of the thoracic arterial vasculature in great detail and can detect abnormalities of the pulmonary and systemic arteries. Greyscale imaging in the parasternal short-axis view allows detailed depiction of the main pulmonary trunk and left and right pulmonary arteries. The aorta and its branching arch vessels are best visualised from the high parasternal and suprasternal views, allowing anatomical delineation to detect dilatation, narrowing or interruption of the aorta as well as an abnormal course of the aorta or its major branches. The descending and abdominal aorta can be appreciated from the subcostal views. Colour-Doppler flow shows the direction of blood flow and detects areas of turbulent flow, while pulsed-wave and continuous-wave Doppler assessment provides information on the location and severity of stenosis within the thoracic arteries. However, with increasing age and body mass index, echocardiographic evaluation of the thoracic vasculature is hampered by suboptimal acoustic windows. Furthermore, echocardiography cannot provide information on the surrounding structures, such as the trachea and oesophagus.

Oesophagography, using barium as a contrast medium, can be used to evaluate complaints of dysphagia, which is not infrequently the presenting symptom in adults with a vascular ring. Vascular rings cause a typical indentation on the oesophagus which give rise to the suspicion of the diagnosis (Berdon 2000). In clinical practice, however, this modality has been largely replaced by cross-sectional imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI).

CT and MRI are the preferred non-invasive imaging modalities in adults for assessment of abnormalities of the systemic and pulmonary arterial system as well as for follow-up and assessing complications due to prior interventions.

The typical MRI examination starts with scout images in the three orthogonal planes, on which one can appreciate the large systemic and pulmonary arterial anatomy, and these scout views are used for planning dedicated MRI sequences. Targeted spin echo ('black blood') as well as gradient echo ('white blood') MRI sequences are used for anatomical evaluation. For evaluation of the aorta and branching vessels, the imaging planes are planned in at least two perpendicular directions when a two-dimensional (2D) technique is used, including the oblique sagittal plane along the long axis of the aorta. For imaging the right ventricular outflow tract (RVOT), the pulmonary valve and the proximal main pulmonary arteries planning along the long axis of the RVOT are recommended, whereas the right and left pulmonary arteries can be evaluated in an axial or oblique axial stack (Fratz et al. 2013). Isotropic three-dimensional (3D) imaging using contrastenhanced or non-contrast-enhanced sequences can also be used to assess the thoracic vasculature.

Besides detailed anatomical evaluation, MRI also provides functional information on the hemodynamic consequences of vascular abnormalities. For example, phase-contrast flow velocity mapping is used to assess the shunt volume in extracardiac shunts, to quantify differential lung perfusion in the presence of branch pulmonary artery stenosis, and help gauge the hemodynamic severity of (re)coarctation of the aorta.

CT is especially useful to determine the relationship between the vessels and the airways for planning an intervention. Furthermore, CT is the preferred imaging modality in patients with contraindications to MRI or for stent evaluation. Correct contrast-bolus timing is essential and should be adjusted to the vasculature under investigation: CT pulmonary angiography, CT aortic angiography or both. Although radiation dose has been reduced considerably with new scan and reconstruction techniques (Xu et al. 2014; Kroft et al. 2010), the radiation burden of repeated CT scans is of concern and should be taken into account, especially in children and young adults with congenital heart disease, making CT less attractive for serial follow-up (Baumgartner et al. 2010). Nuclear imaging techniques may be used to assess heterogeneity of pulmonary perfusion in patients with branch pulmonary artery stenosis and contraindications for MRI.

Cardiac catheterisation for diagnostic evaluation of systemic and pulmonary arteries is currently largely replaced by MRI and CT. However, fluoroscopic angiograms are still obtained prior to and during percutaneous interventions. Occlusion devices, balloons and stents are placed under fluoroscopy guidance. Furthermore, rightheart catheterisation is used in cases of suspected pulmonary hypertension in the rare adult patient with a long-standing unrepaired left-to-right shunt, such as septal defects, an aortopulmonary window or a patent arterial duct.

2 Development of the Systemic and Pulmonary Arterial System

The development from the embryological initial symmetrical cardiovascular system to a unilateral left-sided aortic arch and arterial duct is an intriguing and complex process that is not completely understood. The aortic and pulmonary vascular system develops from the aortic sac and the pharyngeal arch arteries (PAA) through extensive remodelling driven by flow-mediated mechanisms, with angiogenesis and apoptosis in a spatio-temporal pattern (Gittenberger-de Groot et al. 2006). In early embryonic stages, the aortic sac is divided by the aortopulmonary septum into ascending aorta and main pulmonary artery (Bartelings and Gittenberger-de Groot 1989). During cardiovascular morphogenesis five paired PAAs develop, numbered I-VI, with the fifth PAA never reaching full maturation. Eventually, the first and second PAAs contribute to the vasculature of the head, and the third forms the carotid artery system. During normal cardiovascular development the left fourth PAA forms the main part of the left-sided aortic arch and the left sixth PAA eventually connects the pulmonary artery system to the aorta through the arterial duct (Gittenberger-de Groot et al. 2006). Disruption of these complex events leads to a variety of abnormalities of the systemic and pulmonary arteries, occurring de novo or in the setting of known syndromes.

3 Individual Lesions

3.1 Coarctation of the Aorta

Coarctation of the aorta (CoA) accounts for 4.2% of all congenital heart diseases (Hoffman and Kaplan 2002) and most commonly occurs in a juxtaductal position distal to the origin of the left subclavian artery (Fig. 1) (Backer and Mavroudis 2000a). A clear association with bicuspid aortic valve (Fig. 2) exists with a reported prevalence as high as 85% in patients with coarctation (Roos-Hesselink et al. 2003). CoA is associated with Turner, Williams(-Beuren) and congenital rubella syndromes. CoA can be associated with intracardiac malformations including ventricular septal defects and left ventricular outflow tract obstruction. CoA may also develop secondary in Takayasu aortitis (Baumgartner et al. 2010).

Interrupted aortic arch (IAA) is a rare congenital cardiac defect. Given the near-universal mortality with closure of the arterial duct, presentations during adulthood are exceedingly rare (Knapper et al. 2014). After correction complications are similar to those after CoA correction and include aortic restenosis and aneurysm formation (Yang et al. 2008). Diagnosis and follow-up imaging are similar to CoA (Dillman et al. 2008; Yang et al. 2008).

3.1.1 Presentation and Diagnosis

Critical CoA presents at young age with signs of cardiac failure. In adults, CoA often presents inconspicuously with a murmur, hypertension, weak femoral pulses, headaches or, occasionally, claudication. Of note, in patients with extensive collateral circulation induced by a CoA, femoral pulses or lower extremity blood pressure may appear almost normal (Torok et al. 2015).



Fig. 1 Coarctation of the aorta. 9-year-old male with bicuspid aortic valve and coarctation of the aortic. Gadolinium-enhanced MR angiography. Parasagittal orientation showing the coarctation with collateral arteries. (a)

Maximum intensity projection image demonstrating prominent intercostal- and internal mammary arteries (*arrows*). (b) Volume-rendering reconstruction of the aorta and collateral arteries showing the site of coarctation (*arrow*)



Fig. 2 Bicuspid aortic valve. 28-year-old male patient with bicuspid aortic valve and mild dilatation of the ascending aorta. Axial views of through-plane velocity-encoded phase-contrast MRI acquisition of the flow velocity through the valve ((a) magnitude image, (b) phase image). In (c) a double-oblique lateral view of the aortic outflow tract with the bicuspid aortic valve is presented

with aortic regurgitation (*arrow*). (**d**) shows streamline visualisation with colour coding of systolic outflow, with abnormal circulating blood flow (*arrowhead*) and helical flow in the aortic arch. LV left ventricle, LA left atrium, Ao aorta. Figure from unravelling cardiovascular disease using four-dimensional flow cardiovascular magnetic resonance. (Adapted from Kamphuis et al. 2017)

Chest radiography may provide clues to the presence of CoA. In 50-66% of adults with CoA, a figure of three sign can be appreciated on the posterior-anterior chest radiograph, caused by dilatation of the aorta proximal to the stenosis, indentation at the site of stenosis and dilatation distal to the stenosis (Ferguson et al. 2007). Rib notching (deep grooves at the caudal part of the ribs, especially of the 4th-8th ribs) can sometimes be observed in adult patients with CoA, and is caused by collateral circulation via dilated intercostal vessels to bypass the site of stenosis. In patients with favourable windows, echocardiography has proved to be accurate in detecting aortic arch obstruction including CoA (Nihoyannopoulos et al. 1987; Tworetzky et al. 1999). In the suprasternal view, the aorta can be evaluated in its long-axis projection with special attention to the predilection site of CoA and diameters of all segments of the aortic arch. Doppler techniques are used to assess the velocities in the aorta, estimate the pressure gradient over the CoA site and evaluate the presence of the typical CoA flow pattern of antegrade diastolic forward flow. In adults with suspected CoA, CT and MRI are frequently used as the aortic arch, the site of coarctation and the collateral circulation can be appreciated in greater clarity and more detail (Karaosmanoglu et al. 2015). A stan-

dard MRI examination for the evaluation of a (re) CoA may include cine imaging in the long-axis plane of the aortic arch, contrast-enhanced MRA of the aorta (Fig. 1) and phase-contrast MRI at several sites within the aorta, including the aortic valve level, aorta just proximal to the CoA and distal to the CoA at the level of the diaphragm (Fratz et al. 2013). Based on the flow volume measurements just above the CoA site and at the level of the diaphragm the amount of collateral flow can be estimated (Fig. 3) (Steffens et al. 1994). Similar to echocardiography, phasecontrast MRI distal to the stenosis can reveal increased blood velocity and abnormal flow patterns. The combination of indexed minimal aortic cross-sectional area (derived from contrastenhanced MRA) and heart rate-corrected deceleration time in the descending aorta at the level of the right pulmonary artery (derived from phasecontrast MRI) has been used to predict a transcatheter peak-to-peak pressure gradient of \geq 20 mmHg with a sensitivity of 90% and a specificity of 76% in native as well as recurrent CoA (Muzzarelli et al. 2011).

Recently, 4D flow MRI has been introduced to assess flow patterns in patients with CoA (Fig. 4) and allows estimations of regional pressure differences within the aorta (Rengier et al. 2015). Although the clinical significance of this technique



Fig. 3 Collateral flow quantification in coarctation of the aorta. Same patient as in Fig 1. MRI flow imaging with phase-contrast technique immediately distal to the coarctation (upper line in (a) and upper row (b and c)) and at diaphragm level (lower line in (a) and lower row (d and e)).

Flow in the descending aorta is 2.5 L/min immediately distal to the coarctation (\mathbf{c}), and 3.0 L/min at diaphragm level (\mathbf{e}), indicating haemodynamically significant coarctation for the aorta. *Ao* aorta

65 -10 cm/s Fig. 4 4D flow MRI of coarctation of the aorta. 29-year-

needs to be established, it has shown potential as a prognostic tool to assess the risk of aneurysms formation (Hope et al. 2010a, b, 2011).

CT evaluation of a (native) CoA can be performed with high diagnostic accuracy, but should

be reserved for patients in whom echocardiography is insufficient and who cannot undergo an MRI examination (Nance et al. 2016).

Treatment 3.1.2

Critical CoA typically is corrected early in life with low mortality and morbidity (Ungerleider et al. 2013). In adults with CoA, intervention is recommended in patients with a peak-to-peak coarctation gradient greater than or equal to 20 mmHg or peak-to-peak coarctation gradient less than 20 mmHg in the presence of anatomic imaging evidence of significant coarctation and evidence of significant collateral flow (Warnes et al. 2008). Balloon angioplasty for treatment of native CoA shows less favourable outcomes compared to surgery and stent treatment of CoA (Forbes et al. 2011), but can be used as a treatment of recoarctation after previous surgical repair of CoA (Torok et al. 2015). Stent treatment of CoA has significant less acute complications compared to surgery or balloon angioplasty. Intermediate outcomes show that stent treatment is safe in children and adults with CoA with persistent relief of the obstruction (Meadows et al. 2015).

3.1.3 Follow-Up Imaging

Patients after treatment of CoA require lifelong follow-up with imaging of the aorta by CT or MRI at intervals of 5 years or less, according to current guidelines (Baumgartner et al. 2010; Warnes et al. 2008). Common complications after CoA treatment are recoarctation, aneurysm formation, hypertension and stent fracture. Special notice should be given to pregnant women with (treated) CoA, because hypertension-related adverse outcomes are common. Especially, smaller aortic dimensions, as assessed with MRI, are related to an increased risk of stroke, newly diagnosed hypertension and arrhythmias (Jimenez-Juan et al. 2014).

Restenosis after previous CoA treatment is common, especially in patients who underwent CoA repair within the first year of age (Brown et al. 2010; Chen et al. 2013). Although echocar-

old male with a coarctation of the aorta. In (a) a bright blood image of the aortic arch and proximal descending aorta with coarctation of the aorta (arrow). (b) shows the streamline visualisation with colour coding. Distal to the coarctation site, abnormal circular flow is present (arrowhead). Ao aorta. Figure from unravelling cardiovascular disease using four-dimensional flow cardiovascular magnetic resonance. (Adapted from Kamphuis et al. 2017)



diography is the first-line modality for the followup, many adults require cross-sectional imaging for adequate visualisation (Karaosmanoglu et al. 2015). According to the guidelines, MRI is the preferred imaging method (Fig. 5) (Baumgartner et al. 2010), and combined with clinical assessment proved to be cost effective in adults to identify complications, including recoarctation, after CoA repair (Therrien et al. 2000).

Serial follow-up with CT and MRI is mandatory in CoA patients after treatment, given the considerable incidence of aneurysm formation (Fig. 6), particularly in those whom patch aortoplasty was performed (Cramer et al. 2013). Many of these patients are asymptomatic (Hoffman et al. 2014; Tsai et al. 2011) and echocardiography is hampered by a poor sensitivity for aneurysm detection of only 24%.



Fig. 5 Follow-up imaging after coarctation repair with MRI. 11-year-old female patient after coarctectomy and end-to-end anastomosis. The non-contrast 3D steady-state free precession dataset and thick multiplanar reconstruction reveals no signs of re-coarctation (*). *Ao* aorta

Stent fracture or in-stent stenosis is a late complication after CoA treatment (Nance et al. 2016). Echocardiography can raise the suspicion of in-stent stenosis or fracture when increased velocity is encountered in the aorta distal to the stent. On MRI examination, metal stents lead to artefacts expanding beyond the immediate area around the stent. Spin-echo techniques and, to a lesser extent, gradient-echo techniques minimise the image degradation as compared to steadystate free precession pulse sequences, but in-stent stenosis remains impossible to be ruled out with MRI (Shepherd et al. 2015; Valsangiacomo Buechel et al. 2015). Therefore, CT is the modality of choice to assess stent patency and integrity (Fig. 7) (Nance et al. 2016).

3.2 Aortopulmonary Window

Aortopulmonary window, also known as aortopulmonary septal defect, is the rarest of the septal defects and accounts for 0.1% of all congenital cardiac anomalies (Jacobs et al. 2000). An AP window is usually diagnosed in infancy with signs of a large left-to-right shunt, such as respiratory distress, congestive heart failure or because of a murmur (Naimo et al. 2014). However, numerous case reports describe the primary diagnosis of an AP window in adulthood, often with concomitant pulmonary hypertension.

3.2.1 Diagnosis

Echocardiography is the principal method to evaluate an AP window (Mahle et al. 2010), although the diagnosis may be difficult even in experienced hands (Kiran et al. 2008). CT and MRI can aid in the diagnosis, particularly in adult patients with insufficient acoustic windows (Bobylev et al. 2014; Chattranukulchai et al. 2013; Rider et al. 2013; Wong et al. 2012). In addition, MRI allows quantification of the magnitude and net direction of the shunt. Cardiac catheterisation is recommended in adults with AP window to assess the pulmonary vasculature and pressure (McElhinney et al. 1998).





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Fig. 6 Aneurysm after coarctation repair. CT angiography of a 35-year-old male after end-to-end anastomosis for coarctation of the aorta (**a**). A small aneurysm (*arrow*

3.2.2 Treatment and Follow-Up Imaging

Some types of AP windows are suitable for catheter intervention (Barnes et al. 2011), but most warrant surgical correction, especially in the presence of associated anomalies. Early and late outcomes of correction of simple AP window are excellent, approaching 0% mortality (Naimo et al. 2014). Main issues during follow-up are in panel **b**) is present at the site of coarctation repair (asterisk in **a**). *Ao* aorta. Courtesy Dr. AM Spijkerboer, Amsterdam, The Netherlands

related to the associated anomalies. Especially in AP windows with interrupted aortic arch, recurrent aortic arch obstruction can develop. For follow-up of these patients echocardiography is the first-line imaging modality. When aortic arch obstruction is suspected or in patients with poor echocardiographic windows, CT and MRI are recommended with excellent visualisation of the aorta (Kimura-Hayama et al. 2010).



Fig. 7 Stented coarctation of the aorta. 26-year-old male after stent implantation for coarctation of aorta. The patency of the stent (*arrow*) is depicted in panel (**a**). Panel

3.3 Vascular Rings

Vascular rings refer to a group of thoracic vascular anomalies that encircle and may compress the oesophagus and trachea (Backer and Mavroudis 2000b). Vascular rings comprise 1.4% of all congenital heart diseases (Bjornard et al. 2013). The embryological development of vascular rings was proposed by Edwards (EDWARDS 1948) and is thought to be related to an abnormal sequence of regression of the individual components of the ventral and dorsal aorta and PAAs.

Double-aortic arch and right aortic arch with left ductal ligament are the classic complete vascular rings (Backer and Mavroudis 2000b). Other abnormalities that can cause compression of adjacent structures are innominate artery compression of the trachea and pulmonary sling (Backer and Mavroudis 2000b). The innominate artery compression syndrome is caused by a more distal and posterior takeoff of the innominate artery, typically from a normal left-sided aortic arch. In pulmonary sling syndrome,

(**b**) distal to the stent the aorta is unobstructed (asterisk). *Ao* aorta. Courtesy Dr. AM Spijkerboer, Amsterdam, The Netherlands

the left pulmonary artery branches from the right pulmonary artery and courses between the trachea and the oesophagus on its way to the left lung (Fig. 8) (Backer and Mavroudis 2000b). Distal origin of the right subclavian artery from a left-sided aorta (lusoria artery) can cause dysphagia, referred to as dysphagia lusoria, because of the retro-oesophageal course of the right subclavian artery (Fig. 9) (Levitt and Richter 2007).

Abnormalities of the trachea are frequently seen in relation to vascular rings; especially pulmonary slings are associated with complete tracheal rings. Furthermore, associated congenital cardiac defects are reported and are risk factors for mortality in patients referred for correction of a vascular ring (Suh et al. 2012).

3.3.1 Presentation and Diagnosis

The presentation of vascular rings depends on the severity of tracheo-oesophageal compression. Respiratory symptoms include stridor, wheezing, cough and recurrent respiratory infections. In



Fig. 8 Pulmonary artery sling. CT angiography in a 2-year-old boy with pulmonary artery sling. (a) The left pulmonary artery (LPA) originates distally from the right pulmonary artery (RPA). (b) The LPA (*arrow*) encircles the trachea (asterisks) and courses in front of the oesopha-

gus. (c) The tracheobronchial tree is malformed with mild proximal right main bronchus stenosis (*arrow*), as well as narrowing and compression of the left main bronchus (*arrowhead*)



Fig. 9 Aberrant right subclavian artery. CT angiography in a 76-year-old female patient with aortic aneurysm after ascending aorta replacement. Aberrant right subclavian artery (RSA), also called lusoria artery as incidental finding (*arrow* **a** and **b**). The RSA artery arises as the last

adults, symptoms of oesophageal compression are more common. Regurgitation of indigested food, bloating and chest pain, especially with chunky foods, should trigger an evaluation for vascular rings and related disorders (Levitt and Richter 2007).

Chest radiographs may raise suspicion of a vascular ring as abnormalities are present in more than 95% of patients (Browne 2009). Double or right aortic arches are typically appreciated on the posterior-anterior projection. Tracheal narrowing in close relation to the aortic knob suggests airway compression. A pulmonary sling can cause tracheal hypoplasia as well as bronchial tree branching abnormalities (Browne

artery from the aortic arch, with a retro-oesophageal course to the right axilla (c). Lusoria artery is usually asymptomatic but may cause dysphagia. *RSA* right subclavian artery, *RCC* right carotid communis artery, *LCC* left carotid communis artery

2009). Transthoracic echocardiography is frequently successful in the evaluation of vascular rings or related cardiac abnormalities. In the suprasternal views the aortic arch sidedness and all branching vessels need to be demonstrated. In the past, diagnosis of vascular ring was predominantly made with barium oesophagography. Posterior indentation suggested compression by an aberrant subclavian artery, with a left aberrant subclavian artery causing an indentation pointing towards the left shoulder. Anterior impression of the oesophagus can be caused by a pulmonary sling, while bilateral indentation on the anteroposterior view suggests double-aortic arch (Browne 2009). Currently barium oesophagography has been largely replaced by tomographic techniques such as CT and MRI. Both MRI and CT have sensitivities approaching 100% to detect vascular rings. Some centres prefer MRI (Kir et al. 2012; Smith et al. 2015), while others advocate CT due to its superior spatial resolution and evaluation of the airways (Fig. 10) (Browne 2009; Etesami et al. 2014; Leonardi et al. 2015). The MRI protocol may encompass black-blood imaging in the axial or oblique coronal view to assess vascular anatomy, the tracheobronchial tree and the oesophagus. Contrast-enhanced 3D MRA or 3D SSFP sequences yield the anatomy of the vascular system (Dillman et al. 2011; Smith et al. 2015). If contrast-enhanced MR angiography or CT angiography is used, the contrast bolus must be adjusted for the suspected lesion, i.e. either the aortic arch or the branch pulmonary arteries in suspected pulmonary sling (Dillman et al. 2011).

In patients with a double-aortic arch it is important to identify the smaller arch as this dictates the surgical approach, with the smaller arch being ligated. The four-vessel sign refers to the symmetrical branching of the arch vessels from the aortic arch and is suggestive of a doubleaortic arch (Fig. 11). If one of the arches is atretic or if a ductal ligament remains, diagnosis of vascular ring may be difficult. However, secondary signs may help diagnosing the vascular ring (Gould et al. 2015). Presence of a diverticulum in relation to the aortic arch or a right aortic arch with a left descending aorta should raise the suspicion of an atretic segment with double-aortic arch morphology (Fig. 12). The presence of a diverticulum of Kommerell, from which the subclavian artery arises, is also an indication for the presence of vascular ring with a ductal ligament connecting the diverticulum to the pulmonary artery, most commonly in the setting of a right aortic arch (Gould et al. 2015). Furthermore, a circumflex retro-oesophageal course of the aortic arch or the presence of a ductal dimple at the proximal descending aorta contralateral to the arch is an indicative of a vascular ring (Gould et al. 2015).

3.3.2 Treatment and Follow-Up Imaging

In symptomatic patients surgery is indicated (Backer et al. 2005; Slater and Rothenberg 2016). Long-term survival after vascular ring correction is excellent and comparable to the general population (Ruzmetov et al. 2009). However, recurrent compression symptoms either on the oesophagus or the trachea may occur. The diagnostic workup is similar to initial diagnosis of vascular ring.



Fig. 10 Tracheal narrowing due to double-aortic arch. CT angiography in a 14-year-old male patient with double-aortic arch and recurrent pulmonary infections, coronal reconstructions. (a) Soft-tissue setting showing

vascular ring with large right arch and small left arch. (b) Lung setting showing mild tracheal narrowing at the level of the vascular ring



Fig. 11 Double-aortic arch with attretic segment. 14-yearold male patient with recurrent pulmonary infection. CT angiography showing complete vascular ring: Doubleaortic arch with large right arch and small left arch with an attretic segment (fibrous strand) in connection to the descending aorta. The ring causes tracheal narrowing. Left upper panel above arch level with the typical four vessel sign (**a**), right upper panel at arch level and narrowing of trachea (**b**), left lower panel below arch level (**c**), right lower panel at descending aorta level that had median course (**d**). AA ascending aorta, DA descending aorta, LBA left brachiocephalic artery, LCC left carotid communis artery, LSA left subclavian artery, RCC right carotid communis artery, RSA right subclavian artery

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Fig. 12 Double-aortic arch with atretic segment. 7-month-old female patient, admitted to the intensive care unit with an upper airway tract infection and a history of multiple severe cyanotic incidents. CT angiography showing complete vascular ring: Double-aortic arch with large right arch and small left arch causing tracheal narrowing (a). An atretic segment (*, fibrous strand, confirmed at surgery) is connecting the left arch to the descending aorta (b). Panels (c) and (d): volume-rendered depictions of the double-aortic arch with symmetric branching of the arch vessels causing the typical four vessel sign. *Ao* aorta, *DAa*

artery; LSA left subclavian artery; *R and RAo* right aortic arch, *RCA* right carotid artery, *RSA* right subclavian artery; *SCV* superior caval vein, *Tr* trachea. *marks the location of the atretic segment closing the vascular ring. 14-year old male patient with recurrent pulmonary infection. Volume rendered movie of CT angiography showing a double aortic arch with large right arch and small left arch with an atretic segment (fibrous strand) in connection to the descending aorta. See chapter for further details.

4 Congenital Pulmonary Artery Abnormalities

4.1 Pulmonary Artery Stenosis

Congenital stenosis of the main pulmonary arteries may occur as isolated lesion or may be related to complex congenital heart defects, such as in tetralogy of Fallot (Fig. 13), or several other syndromes such as Williams(–Beuren), Noonan and Alagille (Pierpont et al. 2007). Pulmonary stenosis may also occur as important complication following congenital heart disease surgery, e.g. after arterial switch operation for transposition of the great arteries (Morgan et al. 2017).

At the most severe end of the spectrum of pulmonary artery obstruction is interruption of a pulmonary artery, where the lung is supplied with blood through collateral arteries or a persistent duct that connects to the hilar arterial vessels. This is a rare condition that may be found incidentally in adults, suspected on chest radiograph or found



Fig. 13 Hypoplastic pulmonary arteries with stenoses. 26-year-old female patient with tetralogy of Fallot. Gadolinium-enhanced MR angiography, showing severely hypoplastic main and branch pulmonary arteries (**a**).

by cross-sectional imaging (Castaner et al. 2006). Patients with pulmonary valve atresia often are burdened with severe arborisation abnormalities and multiple peripheral pulmonary stenoses. Peripheral pulmonary artery stenosis, i.e. stenosis in the lobar or segmental/subsegmental pulmonary arteries, has been recognised in adults as a cause of pulmonary hypertension (Tonelli et al. 2015).

4.1.1 Presentation and Diagnosis

Primary presentation of an isolated pulmonary stenosis during adulthood is rare. artery Echocardiography allows evaluation of the main as well as the proximal left and right pulmonary arteries from the parasternal short axis. Luminal narrowing or interruption can be seen on greyscale imaging; turbulence on colour-Doppler imaging marks the location of stenosis, especially if the acoustic windows are limited. Using pulsed or continuous-wave Doppler and applying the modified Bernoulli equation an estimation of the gradient across the stenosis can be obtained. Both MRI and CT can evaluate the morphology of the pulmonary vasculature in great detail. MRI 3D SSFP tech-

Panel (**b**) demonstrates residual, non-unifocalised aortopulmonary collateral arteries (*arrows*). *MPA* main pulmonary artery, *LPA* left pulmonary artery, *RPA* right pulmonary artery

niques or 2D cine SSFP acquisitions in the (oblique) axial plane, or MR angiography (Fig. 13), can be used to image the pulmonary arteries. Phasecontrast velocity mapping can be used to quantify the flow velocities across a stenosis. It should be noted that the resultant velocities are lower than those obtained by pulsed-wave or continuous-wave Doppler ultrasound, unless the imaging slice is at the exact location of the fastest flow jet. This technique is also useful for the quantification of relative pulmonary blood flow, which is typically shifted towards the lung with the unobstructed arterial supply. In patients with severe pulmonary artery narrowing or with an arterial stent, flows in the pulmonary veins may more accurately represent the ipsilateral lung perfusion. Nuclear imaging can also be used to assess differential lung perfusion in case of branch pulmonary artery stenosis, although the technique is increasingly replaced by phasecontrast MRI (Sridharan et al. 2006). Some centres prefer detailed pulmonary fluoroscopic angiography for the planning of treatment, especially in the case of syndromal peripheral pulmonary stenosis (Monge et al. 2013).

4.1.2 Treatment and Follow-Up Imaging

Isolated pulmonary artery stenosis and even longer segment hypoplasia can be treated percutaneously, by either balloon angioplasty or stent placement, or surgically. Morbidity and mortality are low (Monge et al. 2013; van Gameren et al. 2006). Restenosis may occur and warrants close monitoring with echocardiography or cross-sectional imaging modalities. When stents are used for the relief of pulmonary stenosis, evaluation by echocardiography or MRI can be difficult, while CT can provide optimal information on stent integrity and in-stent stenosis (Fig. 14).

4.2 Pulmonary Arteriovenous Malformations

In adults, pulmonary arteriovenous malformations (PAVMs) are increasingly recognised with an estimated prevalence of 1 per 2630 in the general population (Nakayama et al. 2012). The most common cause of PAVMs is hereditary hemorrhagic telangiectasia. Also, AVMs may occur in complex congenital heart disease (Marianeschi et al. 1998). Furthermore, patients with cyanotic heart disease are at substantial increased risk of developing PAVMs, especially in lungs of a subset of patients with so-called single-ventricle circulations, that do not receive blood that has



Fig. 14 Stented left pulmonary artery. CT angiography in an 11-year-old male patient with tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries, after unifocalisation of the collaterals, insertion of pulmonary valve homograft and stent in the proximal left pulmonary artery. (a) The axial reconstruc-

tion shows a calcified homograft, a dilated right pulmonary artery (RPA) and stented LPA. (c) Long-axis view and (d) short-axis view of the LPA, showing intimal hyperplasia, creating in-stent obstruction (*arrows*). Also, presence of thrombus material (dotted *arrows* in **b**, and **c**)



Fig. 15 Pulmonary arteriovenous malformation. 35-yearold patient with a Fontan-type circulation. Pulmonary arteriovenous malformation in the right lung (*circle* and

passed through the liver (Fig. 15), when the liver veins are not incorporated within the Fontan circulation.

Most PAVMs present in the lower lobes. A simple PAVM is fed by a single pulmonary artery, whereas in complex PAVMs more arteries are involved. The efferent vessels drain directly into the pulmonary veins (Cartin-Ceba et al. 2013).

4.2.1 Presentation and Diagnosis

PAVMs often become symptomatic in the first three decades of life, depending on their number and size (Cartin-Ceba et al. 2013). Symptoms and signs include dyspnoea, cyanosis, chest pain, haemorrhage (either hemoptesis or haemothorax) and stroke (Shovlin 2014). Of note, pregnancy is recognised as a period of increased risk for complications caused by PAVMs, such as haemorrhage, pulmonary emboli and myocardial infarction (Shovlin 2014). Chest radiography can reveal round or oval nodules especially in the lower lobes. Of course, nodules on the chest radiograph can have numerous causes and additional imaging is necessary for diagnosing PAVM. Echocardiography can be used to detect the right-to-left shunt via PAVMs with the use of contrast-enhanced echocardiography, either with agitated saline (gas microbubbles) or albumin. Arrival of any gas microbubbles on the left side of the heart, especially after only three to four cardiac cycles, indicates intrapulmonary shunting (Cartin-Ceba et al. 2013). However, this phe-

arrows in **a**) in the anterobasal segment of the right lower lobe (**b**) and (**c**). *AVM* arteriovenous malformation, *a* artery, *v* vein

nomenon can also be seen in normal lungs and in up to 8% of the general population at rest (Shovlin 2014). Radionuclide imaging, although not commonly used in patients with suspected PAVMs, can also be used to detect and approximate the right-to-left shunting (Cartin-Ceba et al. 2013; Shovlin 2014).

Currently, CT is considered the non-invasive reference standard for diagnosing PAVM, and to assess the localisation, number, size, extent and suitability for intervention (Gill et al. 2015). Both afferent and efferent vessels can be visualised. Imaging is timed when the contrast bolus is estimated to reach the pulmonary arteries. MRI has been used for the diagnosis of PAVMs, albeit with lower spatial resolution compared to CT. When using MR, contrast-enhanced MR angiography is the preferred technique, particularly using time-resolved approaches or ECG gated (Maki et al. 2001; Ohno et al. 2002; Schneider et al. 2008). Cardiac catheterisation is currently less frequently employed for diagnosing PAVMs, but is used for their percutaneous treatment.

4.2.2 Treatment and Follow-Up Imaging

Percutaneous embolisation is the treatment of choice in PAVMs with low morbidity and mortality. Occlusion is generally recommended, even in asymptomatic patients, to prevent complications. After embolisation, recanalisation and collateralisation can occur and current guidelines recommend CT scans at 6–12 months post-intervention and subsequently at intervals of 1–5 years (Shovlin 2014).

Conclusions

Imaging of the thoracic systemic and pulmonary arteries in adults most commonly involves evaluation of post-treatment sequelae. Understanding the underlying pathology and possible complications that may occur during follow-up is essential for selecting the appropriate imaging modality and subsequent execution of the examination. Echocardiography is the first-line imaging modality to assess the vascular morphology, but for detailed assessment of the thoracic systemic and pulmonary arteries CT and MRI are often necessary. The main advantages of CT over MRI are the short acquisition times and the option to evaluate intravascular stent patency. MRI, on the other hand, does not depend on ionising radiation and offers functional information.

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