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REVIEW ARTICLE

Congenital anomalies of the pulmonary arteries: an imaging overview

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ABSTRACT

Congenital pulmonary artery anomalies represent a diverse group of abnormalities, ranging from asymptomatic incidental findings to causes of sudden cardiac death. While some may be recognized in childhood, others may be found incidentally in adulthood. We review the clinical and imaging findings in patients with congenital anomalies of the pulmonary arteries, including valvular and perivascular anomalies as well as abnormal narrowing, course and communications of the pulmonary arteries. We also discuss the role of various imaging modalities in the evaluation of these patients. It is vital to be aware of the key radiologic manifestations and associated haemodynamic consequences in these conditions in order to facilitate accurate diagnosis and prognostic stratification.

INTRODUCTION

Congenital pulmonary artery (PA) anomalies range from asymptomatic incidental findings to causes of sudden cardiac death. Some may be recognized in childhood while others may be found incidentally in adulthood. We review the clinical and imaging findings in patients with congenital anomalies of the pulmonary arteries. Specifically, we will start with valvular and perivascular anomalies and subsequently discuss abnormal narrowing, course and communications of the pulmonary arteries (Table 1). We discuss the role of various imaging modalities in the evaluation of these patients and describe the imaging findings related to surgical and interventional procedures performed for the management of these patients.

Embryonic development of the aortic arches, from which the pulmonary arteries arise, is intricate and complex, leaving many opportunities for disruptions along the way. By approximately, Day 29 of embryonic development, all six pairs of aortic arches are present. The pulmonary arteries and ductus arteriosus arise from the sixth pair of aortic arches with some possible contribution from the fourth arch as well. Specifically, the ventral sixth arches become the proximal right and left main pulmonary arteries. The dorsal right sixth arch involutes and the dorsal left sixth arch becomes the ductus arteriosus. By Day 50, the truncus

arteriosus divides into the aorta and pulmonary trunk and the right and left pulmonary arteries have become part of the vascular plexus of the lung buds.

Research has suggested that PA development continues in the neonatal period and can be altered in the presence of abnormal pulmonary blood flow. In the newborn, the pulmonary trunk and main pulmonary arteries are believed to be similar to the aorta with regard to diameter, mural thickness and elastic fibers. When the pulmonary pressures and resistance drop after birth, the elastic fibers of the central pulmonary arteries become irregular and interrupted and the overall wall thickness is less than that of the aorta. Lobar and distal pulmonary arteries maintain continuous mural elastic fibers. In cases of pulmonary hypertension, as present in left-to-right shunts, the central pulmonary arteries are thought to retain their dense elastic fibers. Conversely, in low-flow states, as present in tetralogy of Fallot (TOF), the central pulmonary arteries demonstrate more pronounced thinning and elastic fiber interruptions.¹

Valvular and perivalvular anomalies

Overview of valvular and perivalvular anomalies
Pulmonic stenosis is congenital in 95% of cases and represents roughly 10% of congenital heart disease. There is no race or gender predilection. Valvular stenosis is the

Table 1. Overview of congenital pulmonary artery anomalies

VALVULAR AND PERIVALVULAR ANOMALIES
Isolated dysplastic or dome-shaped pulmonary valve
Absent pulmonary valve syndrome
Quadricuspid and bicuspid pulmonary valve
ABNORMAL NARROWING
PIPA
PA-VSD
PA-IVS
Pulmonary branch stenosis
ANOMALIES OF COURSE
Pulmonary sling
Partial anomalous left pulmonary artery
ABNORMAL COMMUNICATIONS
ALCAPA
PAVF
PDA

ALCAPA, Anomalous left coronary artery from the pulmonary artery; PIPA, Proximal interruption of the pulmonary artery; PA-VSD, Pulmonary atresia with ventricular septal defect; PA-IVS, Pulmonary atresia with intact ventricular septum; PAVF, Pulmonary arteriovenous fistula; PDA, Patent ductus arteriosus;

most common type of congenital pulmonic stenosis, followed by sub- and supravulvar types. The pulmonic valve leaflets are either thickened or partially fused, narrowing the lumen and limiting flow through the right ventricular outflow tract. Degree of pulmonic stenosis is determined by the pressure gradient across the valve. Mild pulmonic stenosis (peak gradient <30–40 mmHg) is usually asymptomatic. Moderate (40–60 mmHg) or

severe (>60 mmHg) pulmonic stenosis can result in systemic venous congestion, mimicking congestive heart failure.²

Pulmonic stenosis is often an isolated lesion, however some associations exist. Subvalvular pulmonic stenosis and bicuspid pulmonic valve are associated with TOF.³ Valvular pulmonic stenosis is associated with Noonan syndrome. Supravulvar pulmonic stenosis is associated with Williams syndrome (Williams-Beuren syndrome), similar to supravulvar aortic stenosis.

The flow jet through the narrowed pulmonic valve is generally directed toward the left PA, resulting in post-stenotic dilation. Classic chest radiograph and CT demonstrate enlargement of the pulmonary trunk and left PA, but normal caliber right PA (Figure 1). Right ventricular hypertrophy may also be apparent as the right ventricle (RV) is subject to elevated pressure overload related to the stenotic valve. Thickening of the pulmonic valve can also be seen. MRI is useful in evaluating the morphology of the pulmonic valve, assessing the direction and size of the flow jet, and quantification of the pressure gradient across the valve using phase contrast imaging.²

Isolated dysplastic or dome-shaped pulmonary valve

The two most common types of congenital isolated pulmonary stenosis are a dome-shaped pulmonary valve (accounting for

Figure 1. Congenital pulmonic stenosis. (A) Frontal chest radiograph in a young adult showing classic asymmetric enlargement of the left PA and main PA (circle). (B) Contrast-enhanced sagittal CT showing congenital pulmonic stenosis with large aneurysmal dilatation of the main PA (*) resulting in displacement of the LAD coronary artery (arrow). (C) Same patient as (B) status post pulmonary valve replacement and resection of PA aneurysm. Notice resolved displacement of the LAD (arrow). LAD, left anterior descending; PA, pulmonary artery.

Figure 2. Absent pulmonary valve in a 19-day-old infant. (A) Contrast-enhanced axial CT shows marked enlargement of the pulmonary arteries (*), compression of the bronchial structures (arrows) and left lower lobe collapse. (B) Virtual bronchoscopy and volume rendered image of the airway in the same patient showing severe narrowing of the carina and right mainstem bronchus (arrows).

40–60% of cases) and dysplastic pulmonary valve (accounting for 20–30%). Dome-shaped pulmonary valve is the result of commissural fusion with two to four raphe, creating a funnel shape and small round orifice. Dysplastic pulmonary valve demonstrates thickened, immobile cusps with myxomatous thickening limited to the free edge of the leaflets. Both will appear as enlarged main and left pulmonary arteries on chest radiograph and CT. Supravalvular narrowing at the sinotubular junction may be seen in association with both dome-shaped and dysplastic valves. Dome-shaped valves can be treated successfully with balloon valvuloplasty, indicated in symptomatic patients with a peak gradient greater than 50 mmHg. Dysplastic pulmonary valves more often require replacement, sometimes with a transannular patch if there is associated annular or pulmonary trunk hypoplasia.⁴

Absent pulmonary valve syndrome

Absent pulmonary valve syndrome (APVS) is a rare anomaly where rudimentary or absent pulmonary valve leaflets results in severe pulmonic regurgitation with to-and-fro flow between the RV and PAs. This to-and-fro pattern leads to massively dilated branch pulmonary arteries, which can compress the tracheo-bronchial tree leading to severe bronchomalacia (Figure 2). Nearly, all cases present with a malaligned ventricularseptal defect (VSD) and therefore, APVS is often considered a variant of TOF. Additionally, there is almost always absence of a patent ductus arteriosus (PDA), thought to contribute to the pathogenesis of APVS by preventing exit of blood from the PA into the aorta, thus causing increased PA pressure and pulmonic regurgitation, which interrupts normal valve development.^{5,6}

If the foetus survives to term, the physiology and clinical presentation is essentially identical to TOF, although the hypoxia usually has a cardiac and airway component.⁵ Specifically, hypoxia is often due to both a right-to-left shunt through the VSD and a ventilation–perfusion mismatch related to severe malacia.^{5,7} Nevertheless, APV syndrome is extremely rare and is seen in only about 3–6% of patients with TOF.^{5,8} CT is the

Figure 3. Bicuspid and quadricuspid pulmonary valve. (A) Phase contrast MR in a patient with TOF demonstrating a bicuspid pulmonary valve (arrow). (B) Double oblique images from a contrast-enhanced CT shows a quadricuspid pulmonary valve (arrow) incidentally noted in a 67-year-old male. Note the asymmetry in size, with a smaller right posterolateral leaflet (arrowhead). TOF, tetralogy of Fallot.

primary imaging modality used to evaluate branch PA dilation and associated airway compromise.⁸

Quadricuspid and bicuspid pulmonary valve

The incidence of both quadricuspid pulmonic valve and bicuspid pulmonary valve is between approximately 1 in 1000 and 1 in 20,000, noted incidentally on autopsy studies (Figure 3).^{9,10} Quadricuspid pulmonic valve rarely presents with clinical complications and congenital cardiac anomalies are reported in only 4% of cases.⁹ Conversely, bicuspid pulmonary valve is rarely an isolated finding and is usually associated with other congenital cardiac anomalies, most often a TOF.¹¹

Abnormal narrowing

Proximal interruption of the pulmonary artery

Proximal interruption of the pulmonary artery (PIPA), also known as unilateral absence or atresia of a branch PA, is a rare anomaly with a prevalence of approximately 1 in 200,000.¹² There is no gender predilection. There are two believed aetiologies of PIPA. The first is failure of the sixth aortic arch (which form extrapulmonary/mediastinal pulmonary arteries) and lung bud development (which forms intrapulmonary pulmonary arteries, beyond the level of the hila), usually leading to total lung atresia on the affected side. The second is thought to be related to ductal origin of a PA that results in proximal interruption of that vessel when ductal tissue regresses at birth.^{12–14}

PIPA is invariably unilateral and, in most cases, the interrupted PA lies on the opposite side of the aortic arch. Interruption of the right PA is more common and typically occurs in isolation. Interruption of the left PA is usually associated with right aortic arch and congenital cardiac anomalies, most commonly TOF. Although the affected PA abruptly terminates at the hilum, the intrapulmonary vascular network remains intact. Intrapulmonary arteries continue to develop as they receive oxygenated blood via systemic collateral vessels, either bronchial or transpleural (intercostal, internal mammary, subclavian, or innominate arteries). Collateral vessels respond to chronic pulmonary ischaemia and decreased pulmonary blood flow via smooth muscle wall hypertrophy and dilation to direct more oxygenated blood to the affected lung.¹⁵ In order to reflect this preservation

Figure 4. PIPA in two patients. (A) Contrast-enhanced axial CT images show left PIPA, with associated right aortic arch and multiple bronchial and intercostal collaterals (arrows). The left PA is completely atretic and not visualized. (B) Lung windows in the same patient as (A) demonstrate thickened, serrated pleura, subpleural bands, and interlobular septal thickening (arrow) due to transpleural collaterals. (C) Repaired right PIPA with a main pulmonary trunk to branch PA graft. A proximal stenosis of the graft developed requiring stenting (arrow), but severe stenosis of the distal anastomosis of the shunt is now present (arrowhead). PA, pulmonary artery; PIPA, proximal interruption of the pulmonary artery.

of the intrapulmonary artery supply, the term “interruption” is favoured over “absence” of the PA.¹⁶

Chest radiograph appearance varies from total atresia of the affected hemithorax to volume loss with secondary compensatory hyperinflation of the contralateral lung. The contralateral hilum appears enlarged compared to the affected hilum reflecting that the contralateral PA receives the entire output of the RV. These findings may be challenging to distinguish from hypogenetic lung syndrome, which should show the classic scimitar appearance of the anomalous pulmonary vein.

CT demonstrates absence of the mediastinal portion of the PA with ipsilateral pulmonary hypoplasia and resultant deviation of the mediastinum to the affected side (Figure 4). Fine reticulation of the affected lung, subpleural parenchymal bands, and serrated pleural thickening, all reflecting hypertrophied and dilated collateral vessels, are easily apparent on CT. Contrast-enhanced CT directly illustrates the hypertrophied collateral vessels.^{16,17}

Presentation is variable. Patients with isolated interruption of the right PA often present incidentally as adults or in the setting of haemoptysis. Haemoptysis is due to rupture of systemic collateral vessels.¹⁵ It occurs in 10–20% of cases, ranging from minor and self-limited to life-threatening. Haemoptysis, recurrent pulmonary infections, or pulmonary hypertension often require intervention. Transcatheter embolization and pneumonectomy are potential treatment options, although the former is usually considered first line given its lower risk profile.^{16,17}

Pulmonary atresia with ventricular septal defect

Pulmonary atresia is estimated to occur in 1 out of every 10,000 live births.¹⁸ Pulmonary atresia with ventricular septal defect (PA-VSD) is group of malformations characterized by a lack of continuity between the RV and the pulmonary arteries and associated with a VSD. Concomitant intracardiac defects in PA-VSD vary and include TOF, double-outlet RV, or transposition of the great arteries. The most severe form of TOF, where severe infundibular stenosis results in complete obstruction of

the pulmonary valve is considered a subset of PA-VSD. Right ventricular hypertrophy is typically present due to elevated pressures from RV outflow obstruction. Genetic factors are contributory, with incidence markedly increased in siblings of those with TOF. Infants usually present with cyanosis and hypoxia or heart failure.^{19,20}

The main and branch pulmonary arteries are usually atretic or hypoplastic, may have complex connections and may not be contiguous with each other. Total disconnection of the PA from the RV necessitates the presence of extracardiac sources of pulmonary blood supply, either a PDA or major aortopulmonary collateral arteries (MAPCAs). MAPCAs are believed to be persistent foetal primitive intersegmental arteries originating from the descending aorta. They connect to the native pulmonary arteries at the lobar or subsegmental level to provide pulmonary blood flow. Unlike typical bronchial arteries, they have elastic walls like the PA and aorta, do not provide mediastinal or spinal cord branches, do not connect to intercostal arteries, and anastomose with the pulmonary arteries near the hilum, not at the lung periphery. Three types of PA-VSD exist classified by source of pulmonary blood supply: (A) native pulmonary arteries supplied by a PDA, (B) blood supply by both native pulmonary arteries and MAPCAs, (C) absent native pulmonary arteries with only MAPCAs as pulmonary blood supply.^{19,20}

CT allows direct visualization and accurate characterization of the branch PA anatomy and delineates the pulmonary blood supply, which is necessary to determine surgical approach. One should describe the length of pulmonary atresia, presence of branch PA confluence, sizes of the main, right and left PAs at the origin and hilum, and presence of any branch PA stenosis. One should also detail the native and collateral blood flow provided to each lung including number, origin, and course of MAPCAs (Figure 5). MAPCAs can develop aneurysmal dilation or stenosis. Stenoses protect the pulmonary vasculature distal to the stenosis from future pulmonary hypertension. If a PDA is present, MAPCAs usually will not coexist in the same lung. The

Figure 5. PA-VSD in a 5-year-old girl. (A) Contrast-enhanced axial CT images show pulmonary atresia (arrow) and MAPCAs (arrowheads). Notice the large caliber of the aorta, a common finding in patients with TOF spectrum (*). (B) 3D volume rendered image demonstrating the large MAPCAs (arrows). 3D, three-dimensional; MAPCAs, major aortopulmonary collateral arteries; PA-VSD, pulmonary atresia with ventricular septal defect; TOF, tetralogy of Fallot.

branch PA at the site of PDA insertion is at high risk of stenosis and subsequent isolation.²⁰

Gadolinium-enhanced three-dimensional MR angiography has also been shown to be a fast and accurate way to assess the PA anatomy in both adults and children while avoiding radiation.^{21,22} Additionally, fast spin echo with double inversion recovery sequences can accurately depict MAPCAs and central pulmonary arteries, phase-shift velocity mapping can be used to quantify shunt sizes, and steady-state free precession (SSFP) cine cardiac imaging allows evaluation of cardiac function, volumes, and mass.²¹ Nevertheless, there are limitations to performing MRI in paediatric patients, particularly those that are critically ill, who often require deeper sedation and may have surgical clips, vascular grafts/stents or pacemakers that either are contraindicated for MRI or would limit the study due to artefact.²³

Pulmonary atresia with intact ventricular septum

Pulmonary atresia with intact ventricular septum (PA-IVS) occurs in approximately 4.2 per 100,000 live births. There is no known genetic association. Newborns present with cyanosis, worsening over the first 48 h of life due to closure of the ductus arteriosus.²⁴

PA-IVS represents a spectrum of right ventricular anomalies, most commonly resulting in a hypoplastic (small) thick-walled (hypertrophied) RV (Figure 6). The RV is thought to be hypoplastic because there is no VSD present to provide flow to the RV cavity promoting growth. This is in contrast to PA-VSD in which shunting across the VSD allows for RV cavity growth. The RV becomes hypertrophied in PA-IVS due to elevated RV pressures from RV outflow obstruction. RV outflow obstruction is typically due to muscular atresia (overgrowth of the infundibulum and apex) and/or valvular atresia with well-developed infundibulum. Additionally, the tricuspid valve is often hypoplastic and dysplastic, which correlates to RV size. A minority of patients with severe tricuspid regurgitation may have a dilated and thin-walled RV and right atrial enlargement due to volume

Figure 6. PA-IVS status post Glenn and Fontan repair. (A) Contrast-enhanced axial CT images show a hypoplastic and hypertrophied RV (*), intact ventricular septum (arrow), (B) marked hypoplasia of the main PA (arrow), and (C) coronary sinusoids (arrow). Coronary sinusoids (ventriculo-coronary connections) may form due to the RV hypertension, may be associated with coronary stenosis, and contribute to morbidity. PA, pulmonary artery; PA-IVS, pulmonary atresia with intact ventricular septum; RV, right ventricle.

overload. Prognosis and treatment depends on the severity of RV hypoplasia.^{24,25}

Importantly, unlike TOF with pulmonary atresia, branch PAs and ductus arteriosus are frequently normal. In fact, blood flow to the PAs is dependent on a PDA. Blood returning to the right atrium passes through a patent foramen ovale to the left heart and into the pulmonary circulation via PDA.^{24,25}

PV-IVS is also associated with coronary circulation anomalies such as sinusoids/fistulae, coronary stenosis, or atresia. Intramyocardial sinusoids are channels within the RV wall that communicate directly within both the coronary arteries and ventricular cavity (Figure 6). They are thought to occur due to elevated RV pressures during development and are distinguished from ventriculocoronary fistulae because they traverse a capillary bed. RV to coronary artery fistulae are seen in 45–55% of patients and usually involve both coronary arteries. Right ventricular dependent coronary circulation is seen in 8–23% of patients and occurs due to proximal coronary artery stenosis or atresia with communication of the distal coronary to the RV cavity to preserve perfusion. In these patients, elevated RV pressures must be maintained in order to sustain coronary perfusion. Fistulae can develop stenosis due to elevated pressures and therefore progress to right ventricular dependent coronary circulation.²⁵

Peripheral pulmonary stenosis

Peripheral pulmonary stenosis (PPS) is actually a form of supra-valvular pulmonary stenosis (SVPS), found in less than 1% of patients with congenital heart disease.²⁶ SVPS is seen in Alagille syndrome, Williams syndrome, Ehlers-Danlos syndrome, and post-rubella syndrome.²⁷ In fact, PPS is a phenotypic hallmark of both Alagille and Williams syndromes.²⁶ Adults with repaired TOF commonly have PPS, often secondary to palliation with a modified Blalock-Taussig shunt in infancy and the frequently encountered acutely angulated LPA (Figure 7).

Pulmonary stenosis can cause RV hypertension leading to hypertrophy, systolic and diastolic dysfunction, and eventual ventricular failure. Surgical treatment of branch PA stenosis is difficult and catheter-based angioplasty with or without stenting is preferred.²⁸ CT or MR angiography can play an important role

Figure 7. Pulmonary branch stenosis in a patient with TOF. (A) Axial MR angiography shows severe left PA stenosis (arrow). (B) Fused phase contrast and magnitude image shows region of interest over the left PA (arrow). Phase contrast imaging can be used to assess differential flow. This patient had 88% right PA and 12% left PA flow. PA, pulmonary artery; TOF, tetralogy of Fallot.

in identifying and delineating the location and extent of PPS. If MRI is being performed, phase contrast should be performed of both pulmonary arteries to quantify flow. Additionally, if stents have been placed for PPS repair, close attention should be paid on follow up studies to exclude stent thrombosis or pseudoaneurysm formation. Cardiac CT or MRI can also show additional findings such as RV hypertrophy or dysfunction and potential systemic collateral supply to the lung distal to the focal stenosis.^{29,30}

Anomalies of course

Pulmonary sling

Pulmonary sling occurs due to an embryologic failure of formation of the sixth aortic arch leading to anomalous origin of the left PA. The left PA arises from the posterior aspect of the right PA and courses over the right mainstem bronchus, forming a sling behind the trachea and anterior to the oesophagus, in order to reach the left hilum.³¹

The outcome of patients with pulmonary sling depends extent of tracheal abnormality and presence or absence of additional cardiac anomalies. Type I pulmonary sling demonstrates compression of the trachea and right mainstem bronchus by the pulmonary sling, presence or absence of complete cartilaginous rings and a normal tracheobronchial branch pattern. It may be associated with tracheobronchomalacia, unilateral pulmonary hyperinflation, or tracheal bronchus. In Type II pulmonary sling, there is extensive airway abnormalities, and thus has a higher morbidity and mortality. There is absence of the normal carina, and instead there is a pseudocarina at the level of T6, resulting in the anomalous PA also being more caudal. There are long segment complete cartilaginous rings and diffuse airway narrowing. Associated abnormalities include absence of the right upper lobe and a bridging right mainstem bronchus arising from the left.^{32,33} The combination of pulmonary sling and complete tracheal cartilage rings is known as the ring-sling complex. Patients typically present within the first few weeks of life with severe obstructive symptoms, including stridor, respiratory distress, or pneumonia.^{34,35}

Figure 8. Pulmonary sling with complete tracheal cartilaginous rings in a 6-month-old infant. (A) Contrast-enhanced CT shows the left PA (arrow) originating from the right PA and coursing between the trachea and oesophagus. (B) Volume rendered images in the same patient show a long segment tracheal narrowing (arrow) and a low T-shaped carina (arrow-head). PA, pulmonary artery.

Frontal and lateral chest radiographs demonstrate asymmetrical hyperinflation with or without tracheal narrowing. In patients with Type II anatomy, the mainstem bronchi have a horizontal course resulting in a low T-shaped carina. CT or MRI delineates the relationship of the airways and blood vessels, shows a rigid “O”-ring trachea without a pars membranacea in cases of complete tracheal rings, provides three-dimensional reconstruction, virtual bronchoscopy and demonstrates dynamic airways changes (Figure 8).^{34–36}

Pulmonary sling is repaired surgically with median sternotomy and cardiopulmonary bypass. Surgeons ligate the anomalous left PA and reimplant it into the main PA anterior to the trachea. Complete tracheal rings are repaired by tracheal resection or end-to-end anastomosis, tracheal autograft, pericardial patch or slide tracheoplasty.³⁵

Partial anomalous left pulmonary artery

Partial anomalous left pulmonary artery is extremely rare, with only approximately seven cases reported in the literature. There is normal bifurcation of the pulmonary trunk but anomalous origin of a branch of the left PA from the right PA (Figure 9). The anomalous left PA may pass anterior and inferior to the trachea without airway obstruction or pass between the trachea and oesophagus with the resultant sling causing airway compression. It may be associated with congenital heart disease.^{37–39}

Abnormal communications

Anomalous left coronary artery from the pulmonary artery

Anomalous left coronary artery from the pulmonary artery (ALCAPA) also known as Bland-White-Garland syndrome, occurs in 1 in 300,000 live births and accounts for 0.2 to 0.5% of congenital heart disease.⁴⁰ Its counterpart ARCAPA is even rarer. It results either due to malrotation of the spiral septum dividing the truncus or malpositioning of the coronary buds.⁴¹

ALCAPA is tolerated in foetal and early neonatal period because of high pulmonary arterial pressures allowing antegrade flow

Figure 9. Partial anomalous left PA. (A) Contrast-enhanced axial CT showed the left lower lobe PA (arrow) arising from the right PA and coursing posterior to the trachea, creating a sling. (B) 3D volume rendered images show resultant narrowing of the trachea (arrow). Case courtesy Monica Ocampo, MD. 3D, three-dimensional; PA, pulmonary artery.

through the left coronary artery (LCA). However, at about 6 weeks of age, the PA pressures decrease leading to a “coronary steal” phenomenon in which oxygenated blood from the LCA drains into the lower pressure PA system. In children, ALCAPA is one of the most common causes of myocardial ischaemia and infarction and 90% die within the first year if left untreated. In adult type ALCAPA, the LCA still fails to supply the myocardium but significant collateral circulation from the right coronary artery to LCA is present. Nevertheless, collateral flow is usually not sufficient and chronic left ventricular subendocardial ischaemia results. 80–90% of these patients will have sudden cardiac death.⁴⁰

Chest radiograph may show left ventricular enlargement from chronic myocardial ischaemia and pulmonary oedema. Cardiac CT and MRI allow direct visualization of the LCA origin directly off the main PA (Figure 10). In the adult form, there are extensive

Figure 10. ALCAPA in a 51-year-old female. Contrast-enhanced axial CT images show the left coronary artery (arrow) arising from the pulmonary trunk and a large caliber both the right (arrowhead) and left coronary arteries. ALCAPA, anomalous left coronary artery from the pulmonary artery.

collaterals between the right coronary artery and LCA, which are both markedly dilated and tortuous. MRI depicts well the reversal of flow at the origin of the LCA into the main PA as a jet entering the main PA on SSFP cine images, quantifiable on phase-contrast images. MR can also show left ventricular wall motion abnormalities, myxomatous degeneration and ischaemic dysfunction of the papillary muscles with associated mitral insufficiency and prolapse, and subendocardial late gadolinium enhancement representing infarction. Evidence of infarction may be predictive of sudden cardiac death and surgical repair should be considered.⁴⁰

Pulmonary arteriovenous fistula/malformation

Pulmonary arteriovenous fistulas (PAVF), also known as pulmonary arteriovenous malformations, are direct communications between pulmonary arteries and pulmonary veins without intervening pulmonary bed (Figure 11). This results in a right-to-left shunt. Rarely, a PA may communicate with a central pulmonary vein or with the left atrium.^{42,43} Prevalence is 2 to 3 per 100,000 live births. Greater than 80% of PAVFs are congenital.

Neonates present with central cyanosis, exertional dyspnoea, and decreased arterial oxygen saturation. PAVFs can result in heart failure or systemic and cerebral emboli if left untreated. Additionally, approximately 70% are associated with hereditary haemorrhagic telangiectasia (Osler-Rendu-Weber syndrome).⁴⁴

Patent ductus arteriosus

PDA is defined as persistent patency of the ductus arteriosus after birth. PDA connects the proximal descending aorta below the origin of the left subclavian artery to the roof of the main PA near the orifice of the left PA. If there is concomitant RV outflow obstruction, the PDA will usually insert into the proximal left PA.⁴⁵ Isolated PDA accounts for approximately 10–12% of congenital heart anomalies.⁴⁶

The haemodynamic significance of a PDA is dictated by the degree of left-to-right shunt, which determined by the PDA size and the difference between systemic and pulmonary vascular resistance. This shunt results in increased pulmonary blood flow manifested on radiograph as increased heart size and increased pulmonary vascularity.

Chest radiograph classically shows left atrial and left ventricular enlargement secondary to left-to-right shunting via the PDA, which results in pulmonary overcirculation and left heart volume overload.⁴⁷ However, radiograph findings can vary if associated cardiac anomalies are present. Transthoracic echocardiography is typically sufficient for PDA evaluation with CTA or MRI usually only required if there are concomitant findings of complex heart disease or pulmonary hypertension. CT angiography can directly depict a PDA with contrast jet indicating a patent shunt. MRI SSFP cine sequences can also show dephasing of blood verifying flow through a patent PDA, although it may cause visual underestimation due to the short TE. Qp/Qs ratio can be calculated, quantitatively representing the severity of the shunt.⁴⁶

Figure 11. PAVF/PAVM in a 55-year-old male. Contrast-enhanced axial (A–C) and sagittal (D) CT images show a left PA to left upper pulmonary vein fistula (arrows). PAVF, pulmonary arteriovenous fistula; PAVM, pulmonary arteriovenous malformation.

PDA is treated with indomethacin, which inhibits prostaglandins. If indomethacin is unsuccessful, then surgical ligation is performed or endovascular occlusion is performed.⁴⁸

CONCLUSION

Congenital PA anomalies represent a diverse group of conditions, ranging from asymptomatic incidental findings to causes of sudden cardiac death. Radiologists should be aware of the key radiologic manifestations, particularly given that many of these anomalies may be incidentally found in adults. This review provides radiologists with a framework for understanding

the wide range of congenital PA anomalies, knowledge of key imaging features, and an overview to the role of various imaging modalities.

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