

Review Article

Radiological Appearance of Congenital Aortic Abnormalities - Pictorial Essay

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Abstract

This is a review of most common congenital aortic abnormalities illustrated by clinical cases with typical CT and MRI findings.

ABBREVIATIONS

AC: Aortic Coarctation; APW: Aorto-Pulmonary Window; ASD: Atrial Septal Defect; BCA: Brachiocephalic Artery; CECT: Contrast-Enhanced Computed Tomography; CHD: Congenital Heart Disease; CT: Computed Tomography; DA: Ductus Arteriosus; DAA: Double Aortic Arch; LCCA: Left Common Carotid Artery; LScA: Left Subclavian Artery; MIP: Maximal Intensity Projection; MRI: Magnetic Resonance Imaging; PAH: Pulmonary Arterial Hypertension; PDA: Patent Ductus Arteriosus; RCCA: Right Common Carotid Artery; RScA: Right Subclavian Artery; RV: Right Ventricle; SSFP: Steady State Free Precession; TA: Truncus Arteriosus; TOF: Tetralogy of Fallot; VSD: Ventriculoseptal Defect.

INTRODUCTION

Embryological development of the great arteries is a complex process comprising multiple sequential stages of genesis and atresia of several intermediate embryonic structures, occurring during the fourth and fifth weeks of gestation [1]. Thus aortic arch, its major branches, right and left pulmonary arteries form as a result of asymmetrical regression of several of the six pairs of embryonic pharyngeal arches [1]. The two embryonic dorsal aortae recombine to become a single descending aorta. Separation of the embryonic truncus arteriosus by an ingrowing septum gives rise to ascending aorta and pulmonary trunk [1]. Given the complexity of embryogenesis of the great arteries, congenital abnormalities of the aorta and pulmonary arteries are relatively common. Some are benign and considered mere anatomical variants, others can be associated with significant pathology. Most commonly encountered congenital abnormalities of the aorta include abnormal configurations of the arch, abnormal communications between the aorta and central pulmonary arteries - so-called conotruncal abnormalities, and persistent structures of fetal circulation - e.g. patent ductus arteriosus. Another category of congenital aortic abnormalities relates to small diameter of the vessel, which could be focal or diffuse - e.g. aortic coarctation. This paper attempts to provide a concise review of radiological appearances of most common congenital

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- Williams syndrome

abnormalities of the aorta as encountered in radiological clinical practice.

Normal anatomy of the thoracic aorta

Normal aorta arises from the anatomical left ventricle. Aortic root is positioned posteriorly and to the right of the proximal pulmonary artery. After giving off the coronary arteries, which normally arise from the sinuses of Valsalva of the aortic root, the aorta extends cranially as a tubular structure without additional vascular branches. It then arches over the left main-stem bronchus to become the descending aorta. Left-sided aortic arch, i.e. to the left of the trachea, is the anatomical norm. Normally, three vessels arise from the aortic arch - first the brachiocephalic (innominate) artery, then the left common carotid artery and finally the left subclavian artery. The brachiocephalic artery branches into the right common carotid and right subclavian arteries. Vertebral arteries normally arise as the first branches from ipsilateral subclavian arteries. Normal computed tomography (CT) anatomy of the thoracic aorta and branches of the arch is illustrated in Figure (1).

Aortic anatomical variants

Some variations in the branching pattern of the aortic arch are common and are not associated with any pathology. These include origin of the left vertebral artery directly from the arch, as opposed to being from the left subclavian artery (Figure 2), and common origin of the brachiocephalic and left common carotid arteries (Figure 3). The latter is referred to as "bovine arch", although its appearance is very different from that seen in cows and other cattle [2].

Aberrant right subclavian artery

This is a relatively common abnormality seen in up to 2% of people undergoing CT or magnetic resonance imaging (MRI) of the chest [3]. In this condition, instead of arising from the right-sided brachiocephalic artery, the right subclavian artery (RScA) takes off directly from the aorta as the last branch of the arch.

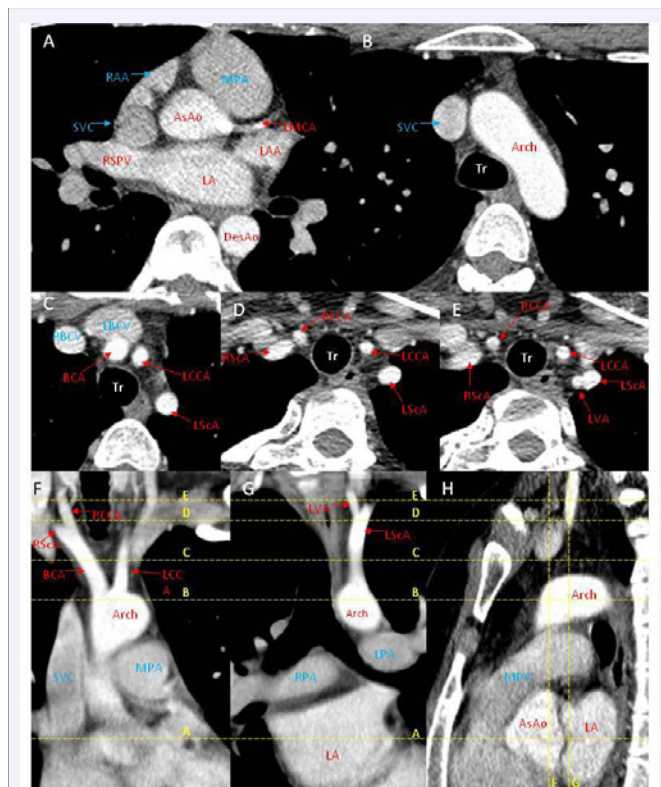


Figure 1 Normal anatomy of the thoracic aorta as seen on CECT on axial (A-E), coronal (F,G) and sagittal (H) images. Dashed yellow lines and yellow letters indicate the levels of the axial images A-E and of the coronal images F and G. In A and H, normal position of the proximal ascending aorta (AsAo) relative to the main pulmonary artery (MPA) - posterior and to the right, and normal position of the descending aorta (DesAo) - left of the midline, are shown. In B, normal position of the aortic arch - to the left of the trachea (Tr) is shown. C shows normal branching pattern of the three arch vessels, which arise in the following order - brachiocephalic artery (BCA), left common carotid artery (LCCA) and left subclavian artery (LSCA), also demonstrated on coronal images in F and G. D shows branching of the brachiocephalic artery into right subclavian artery (RSCA) and right common carotid artery (RCCA), also shown in F. E and G demonstrate origin of the left vertebral artery (LVA) from the left subclavian artery. Additional abbreviations: SVC - superior vena cava, LA - left atrium, LAA - left atrial appendage, RAA - right atrial appendage, RSPV - right superior pulmonary vein, LBCV - left brachiocephalic vein, RBCV - right brachiocephalic vein, LPA - left pulmonary artery, RPA - right pulmonary artery.

It then courses behind the esophagus (Figure 4). It is commonly associated with dilatation of its origin, known as Kommerell's diverticulum - a remnant of the contralateral fourth dorsal aortic arch. On lateral chest radiographs aberrant vessel may be seen as an opacity posterior to the trachea (Figure 4A). Although generally considered a benign variant, the aberrant RSCA can sometimes be associated with dysphagia thought to be due to compression of the esophagus, known as "dysphagia lusoria" [3].

Double aortic arch

Failure of normal involution of one of the embryonic fourth dorsal aortic arches during the 5th gestational week results in double aortic arch (DAA), which encircles the trachea and esophagus in a vascular ring (Figure 5). The left-sided portion of the DAA has a course similar to that of the normal single left aortic arch. The right arch moiety courses to the left behind the

esophagus and joins the left arch. The descending aorta is usually on the left side. The subclavian and carotid arteries arise from their respective arches as separate vessels. This gives the classical appearance of the "4 vessels sign" on axial imaging of the thorax just above the level of the aortic arch (Figure 5C), as opposed to the normal 3 vessels (BCA, LCCA and LSCA) seen in normal single left arch configuration. The right and left arches of the DAA may be symmetric, but usually the right arch is larger, extends more cranially and is more posterior than the left arch.

DAA is the most common type of complete vascular ring. The severity of potential associated symptoms of esophageal and tracheal obstruction is variable. Most symptomatic patients present with stridor, recurrent respiratory infections or dysphagia within the first year of life. Occasionally, the vascular ring is loose and is discovered incidentally in asymptomatic adults. DAA is rarely associated with congenital heart disease [4].

Right aortic arch with mirror-image branching

Occasionally, the aortic arch is located on the right side of the trachea, secondary to failure of normal partial involution of the embryonic double arch. On frontal chest radiographs of individuals with right-sided aortic arch, the indentation of the trachea will be seen on the right side (Figure 6A). The descending

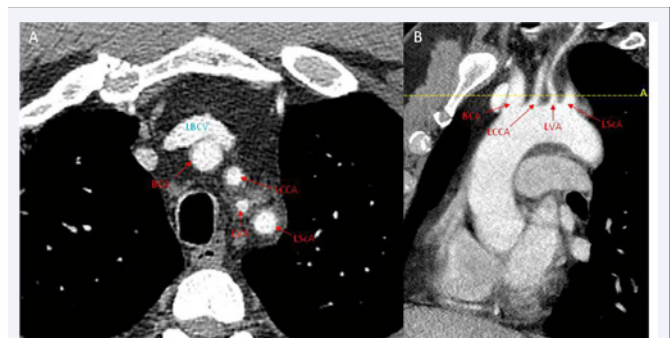


Figure 2 Origin of the left vertebral artery from the arch. CECT axial (A) and sagittal oblique (B) images demonstrate anatomical variant of a left vertebral artery (LVA) arising directly from the aortic arch, between the left common carotid (LCCA) and left subclavian (LSCA) arteries. Dashed yellow line shows the level of the axial image.

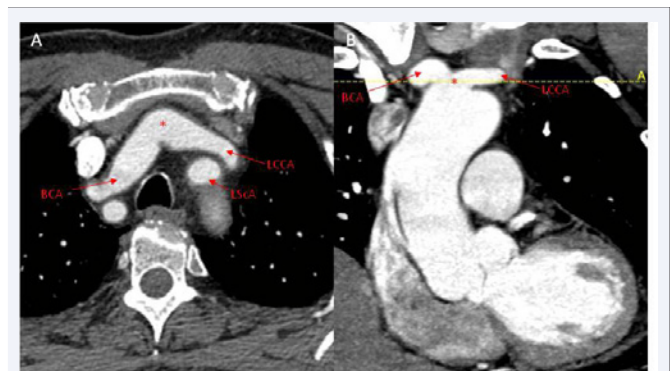


Figure 3 Bovine arch. Axial (A) and coronal (B) images of a CECT of the chest demonstrate common origin of the brachiocephalic (BCA) and left common carotid (LCCA) arteries from a short common trunk (*) - the so-called "bovine arch". Dashed yellow line shows the level of the axial image.



Figure 4 Aberrant right subclavian artery. Lateral chest radiograph (A) demonstrates a round retrotracheal opacity (outlined arrow). This corresponds to the aberrant right subclavian artery (RScA) shown on axial CECT (B), which arises as the last vessel of the arch and courses posterior to the esophagus (Eso). Coronal reformations (C-E) show abnormal order of the vessels arising from the arch - RCCA, LCCA, LScA and RScA. Dashed yellow lines show approximate level of the coronal images.

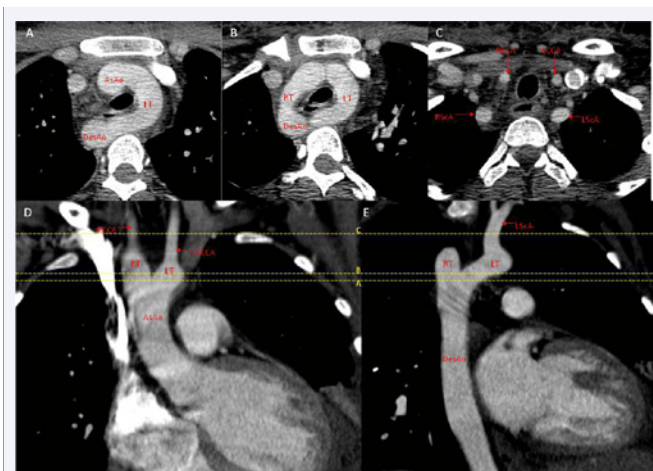


Figure 5 Double aortic arch. Axial (A-C) and oblique coronal (D,E) images of a CECT demonstrate two aortic arches - right (RT) and (LT), completely encircling trachea and esophagus. C demonstrates a classical "four vessel sign" - instead of the expected three arteries (BCA, LCCA and LScA) arising from the arch at this level, 4 vessels (RScA, RCCA, LCCA and LScA) are seen due to the absence of the BCA.

aorta is usually on the right. Right aortic arch is a relatively common anomaly, occurring in approximately 0.05% of people [5].

If the configuration of the right aortic arch branching is symmetrical to that of the normal left arch, with the first branch of the arch being the left-sided BCA, followed by RCCA and RScA, then the pattern is referred to as "mirror-image" (Figure

6). Right aortic arch with mirror-image branching is frequently associated with congenital heart disease (CHD), most commonly tetralogy of Fallot (TOF). Therefore, incidental finding of this aortic abnormality should prompt screening for congenital heart abnormalities.

Right aortic arch with aberrant left subclavian artery

In this anomaly the branching pattern of the right-sided aortic arch is symmetrical to the left aortic arch with aberrant right subclavian artery (Figure 7). As in the latter case, it can be associated with Kommerell's diverticulum. Contrary to the right aortic arch with mirror-image branching, right aortic arch with aberrant LScA is not associated with increased risk of CHD.

Patent ductus arteriosus

Ductus arteriosus (DA) is a normal fetal structure which connects the central pulmonary arteries and the aorta, and together with foramen ovale is part of the mechanism allowing the oxygenated blood from the placenta to bypass the nonfunctioning fetal lungs. DA undergoes spontaneous closure and atresia shortly after birth to become a remnant ligamentum arteriosum. If still patent later in life, it is referred to as patent

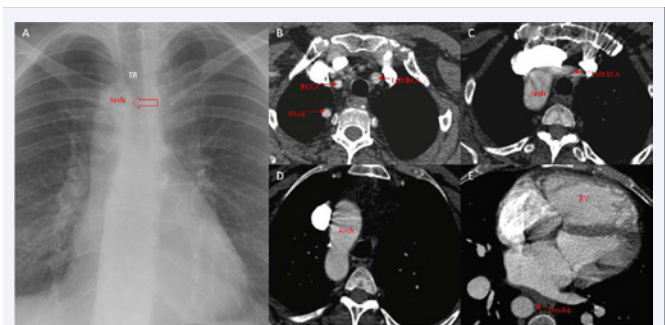


Figure 6 Right aortic arch with mirror-image branching pattern. (A) Postero-anterior chest radiograph demonstrates right sided aortic arch, indenting the right wall of the distal trachea (Tr) (outlined arrow). CECT (B-C) shows that the aortic arch is located to the right of the trachea (D) and that descending thoracic aorta is to the right of the midline (E). The first vessel of the arch is a left brachiocephalic artery (C), which then branches into LCCA and LScA. Second and last branches of the arch are RCCA and RScA, respectively (B). Note right ventricular (RV) hypertrophy (E) in this patient with remote surgery for TOF repair.

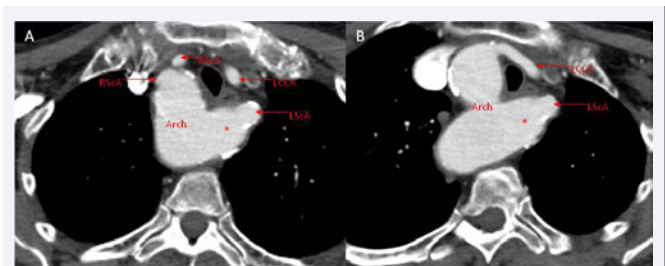


Figure 7 Right aortic arch with aberrant left subclavian artery. Axial (A) and axial oblique (B) images of CECT show right aortic arch. The first branch of the arch is left common carotid artery (LCCA), followed by RCCA and RScA. The last vessel arising from the arch is the aberrant left subclavian artery (LScA). There is a bulbous dilatation at the origin of the LScA (*), so-called "Kommerell's diverticulum".

ductus arteriosus (PDA) (Figure 8). It constitutes a left-to-right shunt and may result in pulmonary arterial hypertension (PAH) and right heart failure [6]. If symptomatic, it can be treated either surgically or endovascularly (Figure 8D).

Aortic coarctation

Aortic coarctation (AC) refers to congenital stenosis of a portion of the aorta due to an intraluminal ridge formed by a mixture of smooth muscle and fibro-elastic tissue [7]. Pre-ductal AC is usually symptomatic at birth and is therefore surgically treated in infancy, and seldom encountered in adults. Post-ductal AC can remain occult or asymptomatic, and be detected later in life usually secondary to systemic arterial hypertension work-up or incidentally. On imaging AC presents as a shelf-like structure in the lumen of the aorta associated with focal luminal stenosis, most commonly in the proximal descending aorta (Figure 9). If hemodynamically significant, it can lead to development of collateral vessels. If the collateral blood flow is via intercostal arteries, then rib notching due to erosions of the undersurface of the ribs by dilated vessels can be seen on thoracic imaging.

AC is often associated with CHD, of which bicuspid aortic valve is the most common. Intracranial arterial aneurysm is another known association with aortic coarctation, and patients with AC should be screened with cerebral CT angiography. AC is often seen in patients with Turner syndrome.

Treatment can be surgical with repair at the site of coarctation or with bypass grafts (Figure 10), or endovascular with dilatation and stenting (Figure 9C). Postsurgical aneurysm at the site of coarctation repair is a relatively common complication (Figure 11).

Aorto-pulmonary window

Ascending aorta and pulmonary trunk arise from a common embryonic vascular structure (truncus arteriosus) which gradually develops an internal septum from conotruncal ridges, separating ascending aorta from pulmonary artery (PA). If normal developmental mechanism fails, abnormal communication between the aorta and PA may persist. When both ascending aorta and pulmonary trunk form together with separate aortic and pulmonic valves, but feature a communication between their respective lumens, the condition is known as aorto-pulmonary window (APW) or aorto-pulmonary septal defect (Figure 12). It is classified in four different types depending on the location of the inter-arterial communication and its extent. APW constitutes a left-to-right shunt and, as such might result in PAH and right heart failure [8].

Truncus arteriosus

Truncus arteriosus (TA) is another consequence of failure of normal segregation of the embryonic primitive common arterial trunk into mature ascending aorta and pulmonary artery, i.e. a conotruncal anomaly. Contrary to the aorto-pulmonary window, TA features a single ventriculo-arterial valve, which can have 2 to 4 cusps, and a single arterial trunk supplying both systemic and pulmonary arterial circulations [9]. Different types of TA are known and at least 2 classification systems exist. Most common type of TA is one where aorta and main PA arise from a common trunk. (Figure 13) shows an example of TA, where right and left pulmonary arteries arise separately from posterior aspect of a



Figure 8 Patent ductus arteriosus. Axial (A,B) and sagittal oblique (C) images of CECT show a channel (*) between the aortic arch (Ao) and left pulmonary artery (LPA). Note markedly enlarged pulmonary trunk in B, secondary to long-standing left-to-right shunt. Follow-up chest radiograph (D) shows an Amplatzer-type occluder device in the expected location of the ductus arteriosus (outlined arrow).

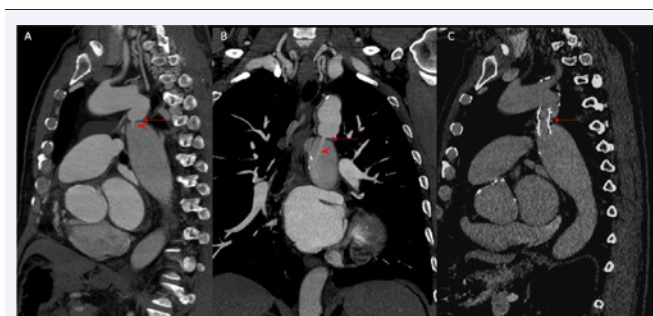


Figure 9 Post-ductal aortic coarctation. Sagittal (A) and coronal (B) reformations of a CECT demonstrate marked narrowing of the proximal descending aorta and severe luminal stenosis due to shelf-like ridge (arrows) associated with severe impediment of blood flow inferred from the jet of contrast across the narrowing (arrowheads). Follow-up CECT sagittal image performed with a different technique (C) shows status post dilatation and placement of an endovascular stent (arrow) across the coarctation with improved luminal caliber.

common arterial trunk (type II according to Collett and Edwards TA classification).

Hemitruncus

Hemitruncus is a condition where one of the pulmonary arteries arises from the ascending aorta, whereas the other pulmonary artery arises normally from the pulmonary trunk. Most of the reported cases of hemitruncus are those where the right PA arises from the aorta - right hemitruncus (Figure 14). It is a rare abnormality and is detected in early infancy, as it is usually fatal, if left untreated [10].

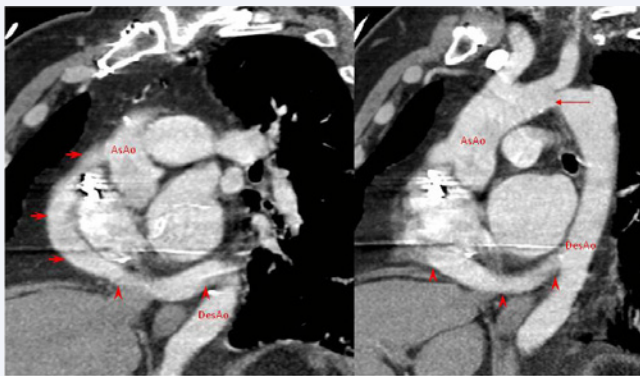


Figure 10 Sagittal oblique reformations of a CECT show an extra-anatomic bypass graft (arrowheads) extending from the proximal ascending aorta (AsAo) to the distal descending aorta (DesAo), coursing underneath the heart, for palliation of the post-ductal coarctation (arrow).

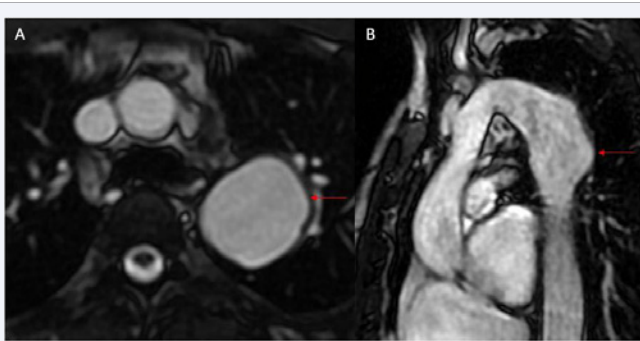


Figure 11 Aneurysm at the site of post-ductal aortic coarctation repair. Axial (A) and sagittal oblique (B) SSFP MRI images demonstrate aneurysm in the proximal descending aorta in a patient status post remote repair of aortic coarctation.

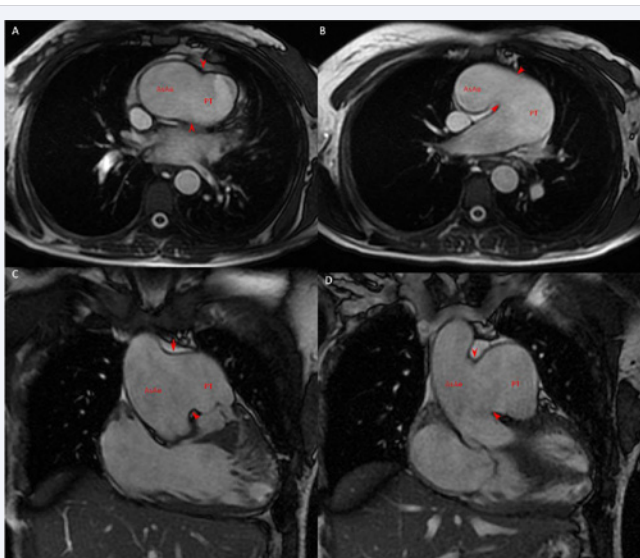


Figure 12 Aorto-pulmonary window. Axial (A,B) and oblique coronal (C,D) SSFP images of cardiac MRI demonstrate a large communication (arrowheads) between the ascending aorta (AsAo) and pulmonary trunk (PT), which starts just above the level of the pulmonic valve. Both aortic and pulmonic valves are present.

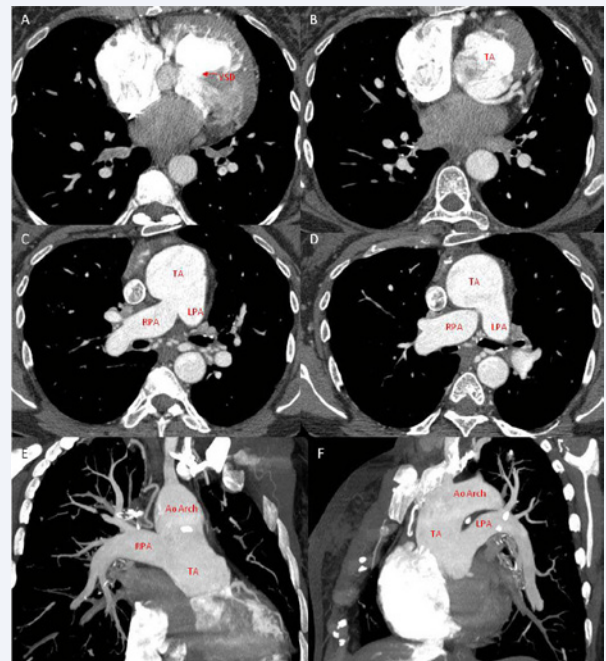


Figure 13 Truncus arteriosus type II. Axial (A-D), coronal oblique MIP (E) and sagittal oblique MIP (F) images of a CECT demonstrate a large ventricular septal defect (VSD in A), a common arterial trunk (TA) straddling the VSD (B). The pulmonary trunk is absent, and right (RPA) and left (LPA) pulmonary arteries arise from the posterior aspect of the TA, which continues distally as aortic arch (AoArch).

Transposition of great arteries

As its name implies, transposition of great arteries (TGA) refers to congenital abnormality where aorta arises from the anatomical right ventricle and pulmonary artery from the anatomical left ventricle, i.e. ventriculo-arterial discordance (Figure 15). If the abnormality is associated with atrio-ventricular discordance (left atrium drains into the right ventricle, and right atrium drains into the left ventricle), then normal systemic and pulmonary circulations are maintained, and the condition is referred to as “congenitally corrected TGA” or “L-loop TGA”. If associated with atrio-ventricular concordance, TGA, then referred to as D-loop, leads to cyanosis and eventually to cardiac and respiratory failure. In live newborns D-TGA is inevitably associated with ASD, VSD or PDA, as it would be otherwise incompatible with life. D-TGA is treated surgically either with atrio-ventricular baffles (Mustard or Senning procedures) or with arterial switch [11].

Williams syndrome (WS)

This is a genetic disorder characterized by facial abnormalities, developmental delay, cognitive impairment, and calcium metabolism abnormalities. With respect to the aorta, WS is associated with small aortic caliber (Figure 16), occasional supravalvular aortic stenosis, and rarely with middle aortic syndrome - progressive narrowing of the abdominal aorta and its visceral branches [12].

CONCLUSION

Congenital aortic abnormalities are relatively common. Most frequently encountered anomalies, such as bovine arch and left

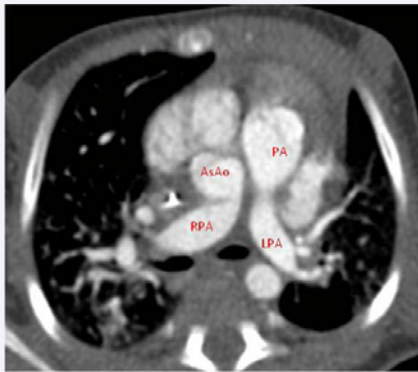


Figure 14 Right hemitruncus. CECT in a newborn male demonstrates left pulmonary artery (LPA) originating from the main pulmonary artery (PA), and right pulmonary artery (RPA) originating from the ascending aorta (AsAo).

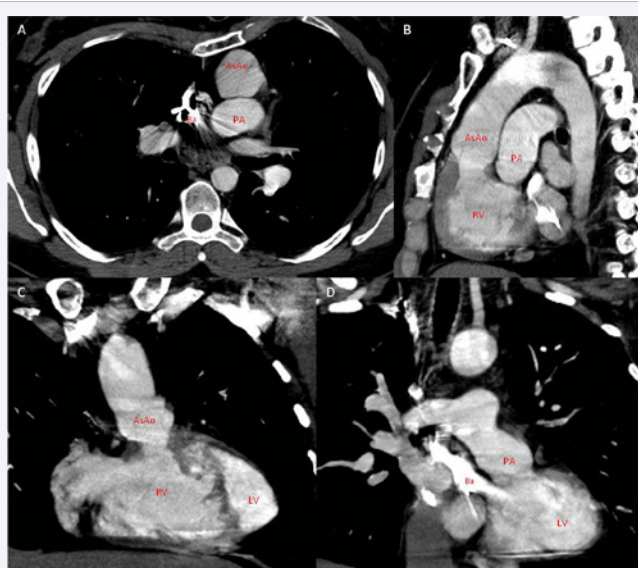


Figure 15 D-transposition of great arteries. Axial (A), sagittal (B) and coronal oblique (C, D) images of CECT demonstrate the ascending aorta (AsAo) arising from the right ventricle (RV) and the pulmonary artery (PA) arising from the left ventricle (LV). The patient had a Mustard procedure at a young age; the contrast-filled right atrial-to-left ventricular baffle (Ba) is seen in A and D.

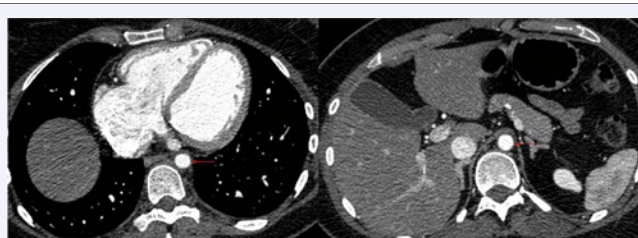


Figure 16 Williams syndrome. Axial CECT shows small diameter of the descending aorta (arrows).

vertebral artery arising from the arch, are considered normal variants. Others, such as double aortic arch and aberrant origin of the subclavian artery, can be associated with symptoms due to compression of the esophagus and trachea. Right aortic arch with mirror image branching is frequently associated with CHD. Abnormal communications between the aorta and pulmonary arteries, such as PDA, aorto-pulmonary window, TA and hemitruncus constitute left-to-right shunts and can lead to PAH and heart failure. D-TGA results in cyanosis and is usually surgically treated in infancy. Adult patients post TGA repair or with congenitally-corrected TGA might be occasionally seen in daily radiological practice. Aortic coarctation and Williams syndrome can be associated with hemodynamically significant aortic stenosis and necessitate surgery or endovascular treatment.

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