Spaghetti Junction: Know your way around mediastinal vascular anomalies

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Learning objectives

To describe the appearances and embryological origins of normal and normal variant mediastinal vascular anatomy on plain chest radiograph (CXR) with multidetector computed tomography (MDCT) correlation.

Characterising the embryological origins of these anomalies allows a detailed appreciation of their expected imaging appearances.

This educational exhibit demonstrates the imaging spectrum of mediastinal vascular anomalies that may be encountered in common practice, thereby enabling confident radiological diagnosis.

Background

Embryological Development of the aortic arch:

Congenital anomalies of the aortic arch and great vessels are reasonably frequently encountered in day to day radiological practice. It is important for the radiologist to accurately recognise these incidental imaging findings. Moreover such vascular anomalies can be symptomatic, particularly in the paediatric population, and may be associated with congenital heart disease. Early radiological diagnosis is paramount and may obviate the need for further clinical investigation.

The first arteries to appear are the right and left primitive aortae. Each has a ventral and dorsal portion. It is the ventral aortae which eventually fuse to form the aortic sac (fig 1). Thereafter, six pairs of arterial arches appear (fig 2), connecting the dorsal and ventral aortae. These aortic arches are also called the pharyngeal arch arteries due to their basket arrangement around the pharynx, begin to form during embryonic folding between day 22 and 24 of gestation. Selective regression and persistence of these arch vessels subsequently forms the great vessels of the head, neck and thorax.

Normally, the $1^{\text{st}}$, $2^{\text{nd}}$ and $5^{\text{th}}$ arches disappear relatively early.

The aortic sac remains connected to the $3^{\text{rd}}$, $4^{\text{th}}$ and $6^{\text{th}}$ arches.
• The 3rd arch forms the common carotid artery and first part of the internal carotid artery.
• The 4th arch forms the left aortic arch and right subclavian artery.
• The 6th arch forms the pulmonary arteries and ductus arteriosus.

An abnormal pattern of regression or persistence of arches leads to the numerous vascular anomalies to be discussed here.

**Embryological development of the systemic venous system:**

The main venous drainage system in the embryo is through paired cardinal veins which drain via a common cardinal vein into the sinus venosus, a structure which becomes incorporated into the right atrium.

The characteristic feature of formation of the vena caval system is anastomoses of the paired cardinal veins in such a way that blood is channeled from left to right.

As oblique anastomoses form between the anterior cardinal veins with regression of the proximal left anterior cardinal vein. The oblique anastomosis thus forms the left brachiocephalic vein, with the right anterior and common cardinal veins forming the right SVC.

Beneath the diaphragm a similar process occurs with anastomoses between the subcardinal veins forming the left renal vein, the right subcardinal vein forming the renal segment of the IVC and the sacrocardinal vein forming the infrarenal segment of the IVC.

**Embryological development of the pulmonary venous system:**

Venous drainage of the primitive lung is via a vascular plexus into the cardinal venous system. A single pulmonary vein then grows from the left atrium with a variable number of pulmonary veins subsequently forming.

**Mediastinal and hilar landmarks on the normal chest radiograph:**

(Fig 3, 4, 5)
In order for radiologists to identify mediastinal vascular anomalies on plain chest radiographs, it is important that the reader has a thorough understanding of the normal anatomy.

**The SVC (a)**

Seen as a vertical line continuous with the right heart border superiorly, outlined by air in the right lung.

**Para-aortic line (b)**

Created by the aortic arch and descending aorta abutted by air within the left lung.

**Aorto-pulmonary window (c)**

A "groove" created between the shadows of the aortic knuckle and pulmonary trunk, outlined by air in the left lung.

**The Hila (d)**

The hilar shadows are created primarily by the pulmonary arteries and pulmonary veins. The left and right hilar points are created by the lower lobe pulmonary arteries inferiorly (fig. 5 blue arrows) and a crossing upper lobe vessel, most commonly the superior pulmonary vein (fig. 5 yellow arrows) superiorly.

**Para-azygos line (e)**

Created by the azygos arch traversing superior to the proximal right main bronchus. This should not be misinterpreted as pathological lymph node enlargement.

**Right paratracheal stripe (f)**

Created by the right wall of the trachea outlined by air within its lumen medially and air within the right lung laterally.

**Anterior junction line (g)**

Created by four layers of pleura adjacent to one another where the two lungs abut each other anteriorly, below the level of the manubrium.

**Posterior junction line (h)**
Created by four layers of pleura adjacent to one another where the two lungs abut each other posteriorly, extending from the level of the aortic arch to the clavicles.

**Azygo-oesophageal line (i)**

Created where the right lung abuts the oesophagus and azygos vein on the right.

**Right and left paravertebral stripes (j)**

Created by the interface between lung and the paravertebral soft tissues. The left paravertebral line should be seen on most adequately penetrated chest radiographs. The right paravertebral line is not usually seen until age-related spinal osteophytes from and displace the mediastinal pleura of the right lung laterally.

Images for this section:
**Fig. 1:** Simplified embryological diagram of the left and right primitive aortae. Initially these are composed of dorsal and ventral portions which subsequently fuse, as demonstrated here, to form the aortic sac. This is continuous with the endocardial tube.

**Fig. 2:** Simplified embryological diagram illustrating the six paired pharyngeal arch arteries which connect the dorsal and ventral aortae. Sequential regression with selective persistence of arch vessels results in the formation of the major arteries in the head, neck and thorax.
Fig. 3: Normal CXR: The numerous mediastinal lines and stripes are indicated here. Please see text for labels and explanation.
Fig. 4: Normal coronal MDCT maximum intensity projection (MIP) of the mediastinal vessels. See text for labels.
Fig. 5: Normal MIP coronal MDCT reformat showing the hilar vascular anatomy. See text for labels.
Aortic Arch Anomalies:

Right Aortic Arch

There are several subtypes of right aortic arch. The two of most clinical importance will be discussed here:

1. Right aortic arch with aberrant left subclavian artery
2. Mirror image right aortic arch

A right aortic arch is formed when the embryological right dorsal aorta fails to regress, with abnormal regression of the left 4\textsuperscript{th} arch and left dorsal aorta.

Right aortic arch with aberrant left subclavian artery:

This anomaly is seen in approximately 1 in 1000 of the population and is associated with a 5-10\% incidence of congenital heart disease, which usually manifests as a simple defect such as an atrial septal defect (ASD).

This is essentially a mirror image of left arch with aberrant right subclavian (see below), with the sequence of great vessels arising from the arch from left to right being left common carotid, right common carotid, right subclavian and aberrant left subclavian. There are usually no brachiocephalic (innominate) arteries, although a right brachiocephalic may be seen (fig 6), which divides in the normal way into the right common carotid and right subclavian arteries.

A frequent variant is a diverticulum of Kommerell, a small retro-oesophageal outpouching which represents a remnant of the left dorsal aorta. It is the origin of the aberrant left subclavian artery (fig 7, 8). This pattern is often associated with a left ligamentum or ductus arteriosus connecting the left pulmonary artery to the diverticulum, thus forming a vascular ring.
The proximal descending aorta descends on the right and typically crosses the midline at the level of the lower mediastinum, where it continues its descent through the diaphragmatic hiatus in the normal fashion.

On CXR the right aortic arch and right descending aorta can be identified. An opacity projected over the left superior mediastinum representing the diverticulum of Kommerell may also be seen (fig 9).

**Mirror image right aortic arch:**

This is a less commonly encountered anomaly that is associated with congenital heart disease in 98% of cases. These most frequently take the form of cyanotic heart disease with Tetralogy of Fallot being the most frequent. Diagnosis is therefore usually made in the neonatal period.

As with the aberrant left subclavian artery subtype there is a right aortic arch however, the descending aorta tends to cross the midline more proximally before continuing its course caudally on the left.

The configuration of the aortic arch vessels is a "mirror image" of normal anatomy such that the first branch (from right to left) is the left brachiocephalic artery, the second branch is the right common carotid artery and the third comprises the right subclavian artery (fig 10, 11). There is no aortic diverticulum.

Again a right aortic knuckle is seen on CXR with an anomalous course of the proximal descending aorta.

A further clue to the presence of a right aortic arch may sometimes be encountered during fluoroscopy, where there is displacement of the oesophagus toward the left. Tracheal deviation toward the left is seen on CXR in both subtypes.

**Left aortic arch with aberrant right subclavian artery**

This occurs in 0.5% of the population and is the commonest true congenital arch anomaly (excluding bovine aortic arch variant: an anatomic variant of aortic arch great vessel branching). It is a consequence of complete involution of the right fourth aortic arch and the cranial dorsal aorta, which normally give origin to the right subclavian artery. The right seventh intersegmental artery normally becomes confluent with the right fourth arch however, in this case it persists in its attachment to the distal aorta.
The aberrant right subclavian artery (ARSCA) is the fourth and final branch of the aortic arch. It arises medially from the aorta and travels towards the right axilla behind the oesophagus and trachea. It does not form a complete vascular ring.

Although usually an incidental finding, ARSCA can present in children and adults with difficulty swallowing or 'dysphagia lusoria' - a result of the vessel abutting the posterior oesophagus.

CXR shows an apparent right paratracheal mass or an opacity coursing obliquely from right to left. MDCT confirms its presence with the 'four artery sign' (fig 12, 13): Right common carotid artery, left common carotid artery, left subclavian artery and ARSCA. A diverticulum of Kommerell can also occur with ARSCA. This is a small dilatation at the origin of the vessel which represents a remnant of the persistent aortic arch.

**Double Aortic Arch**

Double aortic arch is a rare congenital anomaly which results from persistence of both right and left fourth aortic arches and right and left dorsal aortas. In most cases there is a dominant posterior right arch, and a small anterior left arch with a left-sided descending aorta, although variations to this are possible.

Double aortic arch is the commonest form of a vascular ring: a congenital abnormality in which anomalous development of the aortic arch and its tributaries results in complete encirclement of the trachea and oesophagus, often causing compressive symptoms. The second commonest ring is a right aortic arch with a retro-oesophageal left subclavian artery and a left ligamentum arteriosum or patent ductus arteriosus connecting the left subclavian to the pulmonary artery, thus stretching anterior to the trachea. Another non-aortic vascular ring is the pulmonary artery sling (see below).

Each arch gives rise to it’s respective common carotid and subclavian arteries; there are no brachiocephalic arteries. There is usually a left-sided ductus arteriosus.

CXR shows a large right-sided arch displacing the trachea to the left (fig 14); this may be confused for a paratracheal mass. There is a smaller aortic knuckle on the left, and a symmetrical appearance to the great vessels within the superior mediastinum. The lateral radiograph (fig 14) demonstrates increased retrotracheal opacity, tracheal narrowing and anterior bowing. A barium study classically shows a 'reverse S' configuration of the oesophagus caused by bilateral vascular impressions.
Definitive anatomical evaluation is best achieved by helical MDCT (fig 15) or MRI.

**Systemic Venous Anomalies:**

**Persistent Left Superior Vena Cava/Double SVC**

Persistent left superior vena cava (SVC) occurs in approximately 0.3-0.5% of the general population and is the most commonly occurring venous anomaly within the thorax. It results when normal embryological regression of the left anterior and common cardinal veins fails to occur, often with failure of formation of the left brachiocephalic vein.

In the most common subtype both the left and right SVCs are present (double SVC, fig 16, 17, 18) with or without a bridging brachiocephalic vein; a third occurring without the latter. In double SVC, the left vessel is usually larger. In 80-90% of cases the persistent left SVC drains directly into the coronary sinus with no consequent effect on haemodynamics. Of the remaining 10-20%, the majority will drain directly into the left atrium with a resultant right-to-left shunt.

In practice the diagnosis is usually made incidentally, typically during cardiovascular imaging or via MDCT thorax performed for other reasons (fig 19). CXR demonstrates widening of the mediastinum, often more so on the left. A wide aortic shadow with a venous crescent extending from the left border of the aortic arch to the middle of the left clavicle is also a sensitive finding (fig. 20). CT or MRI provide a definitive diagnosis with an enhancing vessel anterolateral to the aortic arch (fig 21).

**Azygos continuation of the Inferior Vena Cava**

This anomaly occurs secondary to abnormal embryological development of the hepatic segment of the inferior vena cava (IVC). The prevalence is approximately 0.6% in the general population.

‘Absence’ of the hepatic segment of the IVC results when the right subcardinal vein fails to make a connection with the liver, thus draining via the supracardinal vein. In the adult, the hepatic segment of the IVC is often not entirely absent, but is in fact hypoplastic, draining directly into the right atrium. The anatomical infrahepatic IVC therefore terminates cranial to the level of the renal veins, where it drains directly into the azygos vein. It is the azygos vein which then enters the thorax via the diaphragmatic hiatus (fig 22, 23).
Within the thorax the anatomy of the azygos vein is unaltered however it is of enlarged calibre due to the increased volume of blood that it contains.

With this anomaly an additional finding is that each gonadal vein drains directly into the ipsilateral renal vein. There is an association with severe congenital heart disease, asplenia and polysplenia syndromes however the majority of cases occur in isolation.

It is paramount for the reading radiologist to identify this anomaly on cross sectional imaging as the enlarged azygous arch and enlarged azygous vein at the diaphragmatic hiatus can be misdiagnosed as a paratracheal mass and retrocrural adenopathy respectively.

Double IVC with hemiazygos continuation is rare. If present however, it must be recognised to avoid misdiagnosing the aberrant vessel as a left mediastinal mass. As is the case with azygos continuation of the IVC, identifying this venous anomaly is important if a patient is due to undergo procedures such as cardiac catheterisation. Simplified, the embryological basis for double IVC with hemiazygos continuation centres upon failed development of the right pre-renal IVC.

**Azygous lobe**

An azygous lobe is a not uncommon congenital variation of the right upper lobe resulting from an anomalous lateral course of the azygous vein within a pleural septum. It is not a true pulmonary lobe as it does not have its own artery, vein or bronchus. It is most commonly an incidental finding with no associated morbidity (fig 18, 24).

**Pulmonary Vascular Anomalies:**

**Pulmonary artery agenesis**

Absence of the left or right main pulmonary artery (PA) is a very rare condition, caused by abnormal development of the 6th pharyngeal arch. If absent on the left, severe cardiac malformations such as Tetralogy of Fallot are commonly encountered.

Typical CXR findings include a small ipsilateral lung and hilum with resultant volume loss in the ipsilateral hemithorax, including elevation of the hemidiaphragm and ipsilateral mediastinal shift (fig 25). The lung itself appears hyperlucent with diminished pulmonary
vascular markings. There is hyperinflation of the contralateral lung. No expiratory air-trapping is demonstrable in the ipsilateral lung, an important distinguishing feature from the more commonly encountered Swyer-James syndrome. On contrast enhanced MDCT (fig 26) absence of the affected PA is readily observed in conjunction with hypertrophy of the systemic vessels (e.g. the intercostal and internal mammary arteries) within the ipsilateral hemithorax.

**Pulmonary artery sling (aberrant left pulmonary artery)**

This congenital anomaly forms 10% of all vascular rings and arises from an abnormality of the 6th pharyngeal arch. Its embryological basis is incompletely understood however there are theories that the left PA fails to develop with a compensatory branch forming from the posterior aspect of the right PA (instead of the pulmonary trunk). This aberrant branch passes over the right mainstem bronchus and then from right to left, between the trachea and oesophagus, as it courses towards the left hilum resulting in compression of the distal posterior trachea and right main bronchus.

It is the only vascular anomaly that courses between the trachea and oesophagus in addition to representing the only vascular ring that results in asymmetric aeration of the lungs (fig 27). CXR demonstrates either hyperinflation or collapse/consolidation of the right lung due compression of the right main bronchus. This in turn will determine whether the normal left lung is compressed or hyperinflated. There is leftward deviation and right-sided compression of the distal trachea, with an 'inverted-T' configuration to the low carina. The aberrant PA may be confused with a right paratracheal or suprahilar mass. The left hilum appears lower than normal due to the more caudal termination of the aberrant vessel. A lateral radiograph demonstrating a mass between the trachea and oesophagus should always lead to suspicion of the presence a PA sling. MDCT confirms the anomalous origin of the left PA from the right PA (fig 28).

PA sling is associated with (i) congenital heart disease including ASD, PDA, VSD and persistent left SVC and (ii) airway anomalies such as anomalous origin of the right main bronchus, tracheomalacia and complete tracheal rings - which can potentiate severe airway obstruction.

**Anomalous venous drainage of the pulmonary veins**

Anomalous venous drainage of the pulmonary veins can either be total or partial, with several subtypes existing in each case. Embryologically, failure of the primitive common pulmonary vein to connect with the pulmonary venous plexus results in persisting connections between the common pulmonary vein and one or more of: the SVC, left
vertical vein or portal vein. Abnormal development of the septum primum can result in direct communication between the pulmonary veins and right atrium.

1. **Partial anomalous pulmonary venous return (PAPVR)**

PAPVR is a rare congenital anomaly whereby typically one of the pulmonary veins drains directly into the right atrium (fig 29), IVC or SVC. At most only mild right cardiac dilatation and engorgement of the pulmonary arteries occurs and pulmonary vascularity is normal. Therefore, CXR is of little use in diagnosis.

2. **Total anomalous pulmonary venous return (TAPVR)**

TAPVR is a congenital anomaly whereby the pulmonary veins return blood to the right side of the heart via a single common vein draining into either the right atrium, coronary sinus or a systemic vein. Three subtypes are recognised;

- **Type 1 (supracardiac):** The most common subtype, in which the single common vein typically drains into a persistent left SVC or "vertical vein" in the superior mediastinum. This in turn drains into the left brachiocephalic vein and thus into the right atrium via the right SVC.

- **Type 2 (cardiac):** The second most common subtype, in which the single common vein drains directly into the coronary sinus or right atrium.

- **Type 3 (infracardiac):** In this subtype the single common vein courses through the diaphragmatic hiatus and typically drains into the portal vein. Drainage into the IVC is less common.

Types 1 and 2 overload the right side of the heart with resultant dilatation of the right atrium, right ventricle and pulmonary arteries. Communication with the left side of the heart usually occurs via an ASD. The classic "snowman/figure-of-8" appearance on CXR is seen in the **type 1** anomaly due to the persistent left SVC or vertical vein which causes prominence of the left superior mediastinum. CXR appearances of the **type 2** anomaly resemble those of transposition of the great vessels: cardiomegaly with a narrow superior mediastinum. The haemodynamics of the **type 3** anomaly differ from the other types and result in pulmonary venous congestion with no enlargement of the right side of the heart. This occurs secondary to high vascular resistance in the anomalous infracardiac vein.
The anomalous vessels associated with PAPVR and TAPVR are clearly demonstrated on cross-sectional imaging.

**Situs Anomalies and Heterotaxy Syndromes:**

Although situs anomalies are rare, they are important to recognise since there is a high association with congenital cardiac defects, particularly in heterotaxy.

"Situs" refers to the position of the atria and viscera relative to the midline in the context of congenital heart disease. Situs solitus is the normal configuration of organs and major vessels. Those with situs inversus have a "mirror image" arrangement of the normal configuration (fig 30), such that the organs and viscera that normally appear on the right are found on the left, and vice versa. Heterotaxy (or situs ambiguous) refers to a complex spectrum of visceral malposition and dysmorphism, with variable atrial arrangement. During embryogenesis the primitive heart and major vessels form during the first 20-30 days. It is disruption of this process that results in heterotaxy syndromes.

The anatomy encountered in a patient with heterotaxy can be extremely variable however two recognised syndromes exist: asplenia (right isomerism) and polysplenia (left isomerism).

In **asplenia** there are trilobed lungs (bilateral right lungs) with bilateral minor fissures, eparterial bronchi (the left and right main bronchi are located superior to their corresponding main pulmonary artery) and bilateral systemic atria. Below the diaphragm there is a midline liver, absent spleen and a variable location of the stomach.

In **polysplenia**, there are bilobed lungs (bilateral left lungs), hyparterial bronchi (the left and right main bronchi are located inferior to their corresponding main pulmonary artery) and bilateral pulmonary atria. Below the diaphragm there is a midline liver and multiple spleens situated along the greater curvature of the stomach. These can occur in variable locations.

**Images for this section:**
Fig. 6: Right aortic arch with aberrant left subclavian artery. Coronal MDCT MIP image through the anterior thorax demonstrating the position of the right brachiocephalic artery (a), aberrant left subclavian artery (b), diverticulum of Kommerell (c) and right common carotid artery (d).
Fig. 7: Right aortic arch with aberrant left subclavian artery: Axial MDCT shows a right sided aortic arch. Image (a) shows the diverticulum of Kommerell (arrow), a small out-pouching from which the left subclavian artery (image b) arises from. (c) Both the ascending (red star) and descending (blue star) aortas are right-sided.
**Fig. 8:** Right aortic arch with aberrant left subclavian artery: Axial MDCT image through the superior mediastinum in a different patient demonstrates the right aortic arch (black arrow) and the aberrant left subclavian artery arising from the diverticulum of Kommerell (white arrow).
Fig. 9: Right aortic arch with aberrant left subclavian: PA and lateral CXR showing the posterolateral impression of the right-sided aortic arch on the trachea (yellow arrows). The normal left aortic knuckle is absent however note a further opacity projected over the left paraspinal line in the superior mediastinum representing the diverticulum of Kommerell (white arrow). This lies at the junction of the right arch and descending aorta and is the origin of the aberrant left subclavian artery, the fourth branch of the aortic arch.
**Fig. 10:** Mirror-image right aortic arch: Contrast enhanced axial MDCT images showing the right aortic arch (star). Superiorly the great vessels are a mirror image of the normal sequence: (a) right subclavian artery, (b) right common carotid artery, (c) left brachiocephalic artery.
**Fig. 11:** Mirror-image aortic arch: Coronal MDCT reformat showing a right-sided aortic arch with 'mirror image' great vessels: (a) right subclavian artery, (b) right common carotid artery, (c) left brachiocephalic artery. This is a rare anomaly.
**Fig. 12:** Left aortic arch with aberrant right subclavian artery: Axial MDCT images showing the retro-oesophageal course of the ARSCA (yellow arrows) as it course across the mediastinum from left to right.
**Fig. 13:** Aberrant right subclavian artery: Coronal MIP MDCT reformat with angulation demonstrates the normal origin of the left subclavian artery (white arrow) from the left aortic arch and the aberrant right subclavian artery (yellow arrow) coursing from its posterior origin towards the right.
Fig. 14: Double aortic arch: PA CXR shows a right-sided arch (white arrow) and a left-sided arch (yellow arrow) both causing an indentation on the tracheal wall. The double arch forms a vascular ring around the trachea, with variable degrees of compressive symptoms. The lateral radiograph demonstrates increased retrotracheal opacification (star) with tracheal narrowing (arrow).
Fig. 15: Double aortic arch: Axial MDCT image through the superior mediastinum demonstrating left and right aortic arches (white arrows) which form a vascular ring around the trachea and oesophagus. Caudally, the arches unite to form a common descending aorta.
**Fig. 16:** Double SVC: Sequential axial MDCT images demonstrate bilateral symmetrical SVCs. The right sided vessel drains normally into the right atrium, whereas the left vessel drains into a dilated coronary sinus.
Fig. 17: Double SVC: Axial and coronal reformat MDCT images showing equal calibre bilateral SVC's (arrows). The right SVC drains normally into the right atrium. In contrast the left SVC drains into the coronary sinus. Rarely alternative drainage of the left SVC into the left atrium results in a symptomatic right-to-left shunt.
Fig. 18: Double SVC and azygous lobe: PA CXR shows bilateral linear para-tracheal vascular opacities representing right (yellow arrows) and left (white arrows) SVC's. Note the lateral course of the azygous vein (curved arrow) resulting in an azgous lobe.
Fig. 19: Persistent left SVC: Coronal MDCT reformat shows the persistent left-sided SVC containing a central venous catheter. In this case the vessel drains directly into the coronary sinus without haemodynamic compromise.
Fig. 20: Persistent left SVC: CXR shows increased linear opacification of the superior mediastinum on the left (yellow arrows), with a double contour in the region of the proximal descending aorta (white arrows) in keeping with a left-sided SVC.
Fig. 21: Persistent left SVC: Axial MDCT image through the superior mediastinum demonstrating the anomalous course of the SVC on the left (arrow). Note in this case the right SVC is hypoplastic and cannot be seen.
**Fig. 22:** Azygous continuation of the IVC: Coronal MDCT reformat demonstrates a dilated azygous vein (yellow star). This is in direct continuation with the suprarenal IVC, bypassing a hypoplastic hepatic IVC as it traverses through the diaphragmatic crus. Note the similar calibre to the adjacent descending aorta (white star).
**Fig. 23:** Azygos continuation of the IVC: Contrast-enhanced axial MDCT image through the lower thorax demonstrating a dilated azygos vein (arrow) and absence of the intrahepatic IVC.
Fig. 24: Azygous lobe: A common incidental anatomical variant resulting from a more lateral course of the azygous vein (arrow). This is not a 'true' pulmonary lobe as it does not receive its own artery, vein or bronchus.
**Fig. 25:** Absent left pulmonary artery: Chest radiograph demonstrating a small left hilum, left hemithorax volume loss and incidental cardiomegaly.
**Fig. 26:** Absent left pulmonary artery: Contrast enhanced axial and coronal MDCT MIP images showing the solitary main right pulmonary artery arising from the pulmonary trunk with absence of the left pulmonary artery. Note the volume loss within the left hemithorax.
Fig. 27: Pulmonary artery sling: A neonatal CXR shows asymmetrical aeration of the lungs with hyperinflation of the right lung due to air-trapping and reduced aeration of the left lung as a result of compressive atelectasis. Note the splayed 'inverted T' configuration of the carina. There is cardiomegaly. The patient is intubated.
**Fig. 28:** Pulmonary artery sling: Axial contrast-enhanced CT shows an aberrant origin of the left pulmonary artery (black arrows) from the right main pulmonary artery (red arrow) instead of the main pulmonary artery (star). As it travels towards the left lung it passes between the trachea (small low attenuation structure) and the oesophagus (containing an NG tube in this case). It thus forms a 'sling' around the trachea resulting in upper airway obstruction.
Fig. 29: Partial Anomalous Pulmonary Venous Return (PAPVR): Cardiac-gated coronal and axial MDCT images showing anomalous drainage of the right superior pulmonary vein (yellow arrows) into the right atrium (black arrow) instead of the left atrium.
Fig. 30: Situs inversus: Axial MDCT shows a right aortic arch with a right-sided descending thoracic aorta (yellow arrows), left SVC (white arrow) and the pulmonary trunk just to the right of the midline (red arrow). Coronal reformat MIP image demonstrating dextrocardia in association with complete situs inversus.
Conclusion

Mediastinal vascular anomalies can provide significant diagnostic challenges. Understanding embryological origins for both normal and normal variant mediastinal anatomy is helpful for recognition of these developmental anomalies.

These vascular anomalies frequently present as an incidental finding in adults on CXR and MDCT thorax but have huge potential implications for patient management with respect to planned vascular intervention as well as thoracic surgery. Furthermore, in the paediatric population early radiological identification can avoid delays in diagnosis of congenital cardiac disease.

Knowledge of the wide spectrum of imaging appearances that may be encountered, together with their associations, is crucial for confident and early radiological diagnosis.

Personal Information

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