Congenital thoracic vascular anomalies occur in the thoracic aorta and branch arteries, pulmonary arteries, thoracic systemic veins, and the pulmonary veins (Table 1). Technological innovations in magnetic resonance (MR) imaging and multidetector-row computed tomography (MDCT) have greatly advanced the noninvasive diagnosis of these anomalies in pediatric patients in recent years. From the neonate to the adolescent, high-resolution two-dimensional (2D) and three-dimensional (3D) MR imaging (MRI), noncontrast MR angiography (MRA), 3D contrast-enhanced MRA, and 3D MDCT angiography (CTA) datasets provide comprehensive multiprojectional, anatomic displays for interactive interpretation, treatment planning, and postoperative and postendovascular evaluation.

Effective use and interpretation of MRI-MRA (Fig. 1) and CTA (Fig. 2) for the evaluation of congenital thoracic vascular anomalies in pediatric patients require fundamental understandings of imaging techniques, anatomic embryology and characteristics, and underlying clinical pathophysiology. Imagers should have knowledge of strategies to optimize protocols to deliver accurate and safe cardiovascular imaging based on the suspected lesion(s) and the clinical stability of the patient. Equally important is the ability to adeptly use advanced postprocessing visualization techniques for image display, interpretation, and clinical management. This article assists the reader in these objectives. Imaging strategies and MR imaging-MR angiography/CTA techniques are reviewed, followed by a discussion on the commonly encountered thoracic congenital vascular anomalies, with emphasis on embryology, clinical manifestations, and characteristic imaging findings.

KEYWORDS

- Aortic arch anomalies
- Pulmonary artery anomalies
- Thoracic systemic venous anomalies
- Pulmonary venous anomalies
- CT angiography
- MR imaging
- MR angiography

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IMAGING STRATEGIES

Chest radiography, echocardiography, vascular ultrasound, esophagography, MRI-MRA, CTA, catheter angiography, or a combination thereof may be performed for diagnostic evaluation of congenital thoracic vascular anomalies. In the past, catheter angiography was regarded as the standard for angiographic evaluation of these disorders but, in the past decade, MR angiography and CTA have gradually replaced catheter angiography for diagnostic purposes. Currently, catheter-based angiography is reserved for endovascular interventions and obtaining direct hemodynamic measurements.

In most pediatric patients presenting with a suspected congenital thoracic vascular anomaly, a frontal (and preferably also a lateral) chest radiograph is the initial imaging modality. The chest radiograph is a fast and inexpensive means to obtain initial direct or indirect evidence for a congenital vascular lesion. Although the chest radiograph may not always yield the specific diagnosis, it is useful to exclude other potential causes, guide initial management, and direct selection of subsequent imaging for confirmation and characterization.

If cardiovascular symptoms predominate (eg, congestive heart failure, systemic hypoperfusion, cyanosis) transthoracic echocardiography (TTE) is usually obtained next. TTE, which requires no radiation, is readily performed and can assess morphology and function (eg, flow dynamics, pressure gradients). However, TTE is limited by the acoustic window (inversely related to patient age and size), acoustic impedance (air), operator skill, and the ability to visualize peripheral vascular segments (eg, pulmonary arteries, pulmonary veins, and supra-aortic branch arteries).

MR imaging-MR angiography or CTA are indicated based on the TTE findings and performance. State-of-the-art MRI-MRA with parallel imaging should be considered before CTA in pediatric patients, because it does not require radiation or iodinated contrast medium and can evaluate vascular hemodynamics as well as vascular morphology with high-resolution anatomic detail. MDCT angiography is indicated when MR imaging-MR angiography is not available, is contraindicated, is nondiagnostic, or has a high pretest probability for being nondiagnostic. CTA should be considered in the patient at high risk with sedation or anesthesia and when airway, lung parenchyma, and other noncardiovascular structures require more detailed imaging.

If respiratory symptoms predominate (eg, stridor, exercise intolerance, apnea, cyanosis, recurrent
upper and lower respiratory infections), MR imaging-MR angiography or CTA is performed following chest radiography per the guidelines discussed earlier. Echocardiography is indicated after a positive MR imaging-MR angiography or CTA to further evaluate cardiac morphology, assess function, and exclude other congenital cardiovascular lesions. In this clinical setting, CTA is advantageous to assess for tracheobronchomalacia, tracheal rings, and extrinsic tracheal compression. Paired inspiration-expiration MDCT angiographic techniques with controlled ventilation have proved

**Fig. 1.** Innominate artery compression on the trachea. A young child with a history of stridor underwent MR imaging-MR angiography for assessment of a possible vascular ring. (A) Sagittal 3D volume rendered image of the MR angiography shows a left aortic arch with normal 3 vessel branches and slightly horizontal course (arrow) of the innominate artery (IA). (B) Oblique MPR projection from a single-shot fast spin echo dark blood acquisition shows the relationship of the IA (long arrow) to the trachea (short arrows). (C–F) Transverse dark blood images show slightly more than 50% extrinsic compression on the trachea (arrow) by the IA (arrowhead), as the IA courses from left to right. LSCA, left subclavian artery; MPR, multiplanar reconstruction; RSCA, right subclavian artery.
to be a reliable means to diagnose clinically significant tracheomalacia and innominate artery compression on the trachea in pediatric patients. If gastrointestinal symptoms predominate (eg, feeding intolerance, failure to thrive, dysphagia, aspiration), an esophagram may be obtained following chest radiography. MRI-MRA or CTA is performed if the esophagram is positive, based on the guidelines discussed earlier. If symptoms persist following a negative esophagram and there remains a high index of suspicion for a thoracic vascular anomaly, consideration should be given to either MR imaging-MR angiography or CTA. Following a positive MRI-MRA or CTA, if not previously obtained, TTE may be indicated to assess for congenital heart structural abnormalities.

IMAGING TECHNIQUES

**MR Imaging and MDCT Protocols**

MR imaging and MDCT angiographic protocols are designed to accurately evaluate cardiac and vascular morphology, and to provide detailed assessments of the central airway and its relationship to cardiac chambers and vascular structures. To minimize the examination duration, only essential scan acquisitions should be used, because patients may become hemodynamically unstable or may develop respiratory distress. Before imaging, it is recommended to review the patient’s clinical presentation, prior medical history, prior imaging studies, current management, and other relevant clinical data. Such information helps in the selection of the most appropriate imaging modality and optimal acquisition and contrast injection protocols. Clinical review also helps to determine the most appropriate location and size for an intravenous catheter (eg, CEMRA, CTA) and whether sedation or anesthesia is required.

**MR imaging-MR angiography**

A standard MRI-MRA protocol for evaluation of pediatric thoracic vascular anomalies includes electrocardiogram-gated black-blood and bright-blood sequences in conjunction with an angiographic sequence. Black-blood and bright-blood MR imaging sequences provide comprehensive anatomic detail, in particular vessel course, caliber, and arterial branching or venous drainage pattern. Black-blood imaging is also applied to evaluate the central airway. Techniques for black-blood MR imaging typically include either single-shot fast spin echo with double inversion recovery or half-Fourier, single-shot fast spin echo with double inversion recovery. Bright-blood imaging in most current practices is achieved with 2D or 3D balanced steady-state free precession. Phase contrast (PC) MR imaging is an optional sequence that is used to evaluate flow direction and velocity, and assess vascular physiology.

Angiographic techniques include time of flight (TOF) MR angiography, PC-MR angiography, and multiphase (arterial and venous) 3D T1-weighted CEMRA. CEMRA is most frequently performed, whereas TOF-MR angiography and PC-MR angiography are reserved for when gadolinium is contraindicated. To maximize 3D displays, CEMRA slice thickness should not be greater than 1.5 mm. When parallel imaging techniques are applied with CEMRA, isotropic, submillimeter datasets can be obtained, yielding the highest possible spatial resolution and robust 3D structural displays on par with those from an MDCT angiogram.

**Fig. 2.** Tubular hypoplasia of the aortic arch. A neonate with congenital aortic stenosis underwent MDCT angiography to further define aortic arch anatomy. (A–C) 3D volume rendered images show moderate to severe tubular hypoplasia of the aortic arch (A, arrow) with a small patent ductus arteriosus (B, C; arrow).
Acquisitions may be in the coronal or sagittal plane, depending on the required anatomic coverage and breath-hold duration.

**MDCT angiography**

Because of its inherent dependence on radiation for generating images, low-dose helical and volumetric pediatric MDCT angiography protocols strive for only 1 core series, namely a single-phase angiographic scan. This single scan is acquired with a slice thickness of 0.5 to 1.5 mm and is synchronized with the arrival of contrast to generate arterial, venous, or equilibrium phase datasets. A noncontrast acquisition is a consideration in the postsurgical or endovascular patient to assess the presence, location, and integrity of high-density material that may degrade vascular interpretation or may be obscured by the contrast (e.g., metallic stents, surgical clips, embolization coils). Additional vascular phases should only be considered if image quality is suboptimal. Ultralow radiation dose volumetric, time-resolved, dynamic CTA with intermittent 2-second to 3-second data acquisitions in 10 to 15 seconds is a promising new technique that can be performed on a 320-channel MDCT scanner; similar to CEMRA, isolated arterial and venous phases can be acquired, providing temporal flow information and more direct imaging of vascular physiology with MDCT. With all pediatric CTA protocols, radiation dose reduction strategies should be used to achieve the lowest possible radiation exposure that will render an interpretable examination for the reader. These radiation dose reduction strategies include using the lowest possible voltage (e.g., 80 kVp), the lowest possible amperage (e.g., weight-based milliampere seconds), the minimum amount of coverage (with shielding of nontarget regions), and the shortest possible scan time (e.g., fastest scan rotation, high-pitch helical, wide-collimation helical, and volumetric MDCT techniques).

**Interpretation and Advanced Visualization**

MR imaging—MR angiography and CTA interpretation address vascular morphology and physiology, as detailed in Tables 2 and 3. Evaluation for associated cardiovascular and noncardiovascular abnormalities is imperative. Postoperative and endovascular evaluations should assess luminal patency and exclude aneurysms, pseudoaneurysms, and iatrogenic injury. For patients who have undergone stent placement, stent migration, fatigue, and disruption should be excluded.

Display and interpretation of thoracic MRA and CTA datasets are most effective when applying advanced workstation visualization techniques (Table 4). Techniques are selected in a complementary manner according to their strengths, using adjustable angiographic window and level settings, including a wide window setting to account for noise and high vascular contrast. Datasets are interrogated in real time with user-defined interaction of the techniques and related workstation tool functions. Alternatively, protocol-driven static postprocessed single and batch-serial images are generated for review along with the source images. The spatial detail of angiographic anatomy is best displayed using 3D volume rendering (VR) and 2D maximum intensity projection (MIP). Both require sliding thin slabs or prerendering editing to remove bone and other anatomic structures that may obscure vascular visualization. However, structural detail is assessed with the highest accuracy using 2D multiplanar reformations (MPR) and curved planar reformations (CPR). Although minimum intensity projection (MinIP) and ray sum (thick MPR) have limited applications in MR and CT angiography MinIP is useful with CTA datasets to show cardiac anomalies.

### Table 2

**Interpretive review: congenital thoracic arterial anomalies**

<table>
<thead>
<tr>
<th>Aorta</th>
<th>Aortic Arch</th>
<th>Ductus</th>
<th>Pulmonary Arteries</th>
<th>Vascular</th>
</tr>
</thead>
<tbody>
<tr>
<td>AsAo</td>
<td>DsAo</td>
<td>Transverse segment</td>
<td>Isthmus</td>
<td>Branch arteries</td>
</tr>
<tr>
<td>Location</td>
<td>Location</td>
<td>Location</td>
<td>Caliber</td>
<td>Number</td>
</tr>
<tr>
<td>Course</td>
<td>Course</td>
<td>Number</td>
<td>Contour</td>
<td>Order</td>
</tr>
<tr>
<td>Caliber</td>
<td>Caliber</td>
<td>Sidedness</td>
<td>Caliber</td>
<td>Preductal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Caliber</td>
<td></td>
<td>Juxtaductal</td>
</tr>
<tr>
<td>SVC</td>
<td>IVC</td>
<td>Innominate Vein</td>
<td>Coronary Sinus</td>
<td>Pulmonary Veins</td>
</tr>
<tr>
<td>----------</td>
<td>-----------</td>
<td>-----------------</td>
<td>----------------</td>
<td>-----------------</td>
</tr>
<tr>
<td>Sidedness</td>
<td>Sidedness</td>
<td>Sidedness</td>
<td>Presence</td>
<td>Number</td>
</tr>
<tr>
<td>Right</td>
<td>Right</td>
<td>Right</td>
<td>Caliber</td>
<td>Course</td>
</tr>
<tr>
<td>Left</td>
<td>Left</td>
<td>Left</td>
<td>Caliber</td>
<td>Atria</td>
</tr>
<tr>
<td>Course</td>
<td>Course</td>
<td>Course</td>
<td>Insertion</td>
<td>Ventricles</td>
</tr>
<tr>
<td>Caliber</td>
<td>Caliber</td>
<td>Flow direction</td>
<td>Patency</td>
<td>Septae</td>
</tr>
<tr>
<td>Insertion</td>
<td>Insertion</td>
<td></td>
<td>Defects</td>
<td>Flatting</td>
</tr>
</tbody>
</table>

*Abbreviations: IVC, inferior vena cava; SVC, superior vena cava.*
valves (eg, bicuspid aortic valve), airways, air trapping, and abnormal nonpulmonary air collections. Ray sum may be applied with MR angiography and CTA datasets to generate radiograph-like images for structural overview.

### Table 4
Cardiovascular advanced visualization techniques

<table>
<thead>
<tr>
<th>Display</th>
<th>Principal Use</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>MPR</td>
<td>2D Structural detail</td>
<td>Structural overview</td>
<td>Limited spatial perception</td>
</tr>
<tr>
<td>CPR</td>
<td>2D Structural overview</td>
<td>Single anatomic display</td>
<td>Operator dependent</td>
</tr>
<tr>
<td>Ray sum</td>
<td>2D Structural overview</td>
<td>Slice through dataset in axial, coronal, sagittal, and oblique projections</td>
<td>Loss of structural detail with increased slab thickness</td>
</tr>
<tr>
<td>MIP</td>
<td>2D Structural overview</td>
<td>Slice through dataset in axial, coronal, sagittal, and oblique projections</td>
<td>Anatomic overlap (vessels, bone, viscera) with increased slab thickness</td>
</tr>
<tr>
<td>MinIP</td>
<td>2D Structural overview</td>
<td>Slice through dataset in axial, coronal, sagittal, and oblique projections</td>
<td>Anatomic overlap</td>
</tr>
<tr>
<td>VR</td>
<td>3D Structural overview</td>
<td>Slice through dataset in axial, coronal, sagittal, and oblique projections</td>
<td>Dependent upon opacity-transfer function</td>
</tr>
</tbody>
</table>

**Abbreviations:** CPR, curved planar reformation; MinIP, minimum intensity projection; MIP, maximum intensity projection; MPR, multiplanar reformation; VR, volume rendered.

development; blood passes from the primitive heart to paired dorsal aorta. Subsequent development of the thoracic aorta, aortic arch branch arteries, pulmonary arteries, and ductus arteriosus occurs during the fourth to eighth weeks of life, beginning with the growth of 6 paired pharyngeal aortic arches (PAA), which bridge the aortic sac to the dorsal aortae through the pharyngeal pouches. Normal development leads to a left-sided aortic arch (LAA) and descending aorta; aortic arch branching (in order) consists of the brachiocephalic artery, the left common carotid artery (LCCA), and the left subclavian artery (LSCA) (see Fig. 1). The ductus arteriosus is left sided, extending from the proximal left pulmonary artery (LPA) to the aortic isthmus, the segment that is between the LSCA and the proximal descending aorta. To achieve this morphology, the aortic arches along with the dorsal aortae and the seventh intersegmental arteries, undergo selective involution and differential growth of persisting structures, as detailed in Table 5.3

THORACIC AORTA
Obstructive Aortic Arch Lesions

**Tubular hypoplasia**
Tubular hypoplasia of the aortic arch (THAA) is a congenital anomaly in which the transverse aorta is reduced in caliber in a short segment such that antegrade flow is reduced. Borders of the aorta in THAA are smooth, without focal narrowing (see Fig. 2). Obstructive physiology may be present in affected patients depending on the length of hypoplasia and the pressure gradient (Fig. 3). It can occur in isolation or be associated with other left-sided obstructive lesions (LSOLs), including congenital mitral stenosis (MS), mitral atresia (MA), hypoplastic left heart syndrome (HLHS), aortic stenosis (AS), aortic atresia (AA), interrupted aortic arch (IAA), and coarctation of the aorta (COA).

The hemodynamic theory can explain the pathogenesis of aortic arch hypoplasia and its association with concomitant congenital heart defect (CHD) lesions. Normal development and size of the aortic arch is dependent on flow dynamics. In normal fetal circulation, half of the combined ventricular blood volume flows from the right ventricle to the main pulmonary artery, whereas the other half flows from the left ventricle to the ascending aorta. Most the pulmonary blood volume flows across the ductus arteriosus to the descending aorta. The majority of the ascending aortic blood volume flows to the coronary circulation and the supra-aortic arteries, resulting in approximately 15% of blood volume flowing across the isthmus and a fetal isthmus that measures up to 70% to 75% the size of the ascending aorta caliber. Any event or lesion that results in decreased antegrade blood flow will lead to less vascular stimulation for growth of the fourth aortic arches and smaller than expected size of the aortic arch and isthmus.

MR imaging—MR angiography and CTA findings of THAA include diffuse smooth narrowing of the

### Table 5
Embryologic origins and development of the left aortic arch

<table>
<thead>
<tr>
<th>Structure</th>
<th>Outcome</th>
<th>Vascular Derivatives</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pharyngeal aortic arches</td>
<td></td>
<td></td>
</tr>
<tr>
<td>First</td>
<td>Near complete involution</td>
<td>Ipsilateral ECA</td>
</tr>
<tr>
<td>Second</td>
<td>Near complete involution</td>
<td>Ipsilateral ECA</td>
</tr>
<tr>
<td>Third</td>
<td>Bilateral persistence</td>
<td>Ipsilateral CCA and ICA</td>
</tr>
<tr>
<td>Fourth</td>
<td>Left: dominant persistence</td>
<td>Left: midaortic arch</td>
</tr>
<tr>
<td></td>
<td>Right: partial persistence</td>
<td>Right: BCA, RSCA</td>
</tr>
<tr>
<td>Fifth</td>
<td>Never forms or involutes</td>
<td>None</td>
</tr>
<tr>
<td>Sixth</td>
<td>Left: dominant persistence</td>
<td>Left</td>
</tr>
<tr>
<td></td>
<td>Right: partial persistence</td>
<td>Ventral: ipsilateral central PA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dorsal: ductus arteriosus</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Right</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ventral: Ipsilateral central PA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dorsal: ductal involution</td>
</tr>
</tbody>
</table>

| Seventh segmental artery   | Bilateral persistence       | Left: LSCA           |
|                            |                             | Right: LSCA          |

| Dorsal aortae              | Left: dominant persistence  | Left: distal aortic arch |
|                            | Right: involutes            | Right: none           |

**Abbreviations:** BCA, brachiocephalic artery; CCA, common carotid artery; ECA, external carotid artery; ICA, internal carotid artery; PA, pulmonary artery.
transverse aorta. Narrowing may involve the entire aortic arch or only a portion (typically midaortic arch to distal aortic arch). Search should be made for associated left-sided obstructive lesions. If MR imaging-MR angiography is performed, phase contrast imaging is essential to document the presence of a gradient. Treatment is only indicated when there is a hemodynamically significant gradient (eg, >15–20 mm Hg). If diagnosis is made by CTA, echocardiography can subsequently be performed to determine the gradient.

Fig. 3. Tubular hypoplasia of the aortic arch with endovascular stent placement. An adolescent with asymmetric upper extremity hypertension underwent MDCT angiography. (A–C) Initial diagnostic 3D volume rendered images show mild tubular hypoplasia of a mildly elongated transverse aortic arch (A, B; long arrow) with compensatory LSCA enlargement (B, C short arrows) for collateral flow. Catheter angiography confirmed a significant pressure gradient that subsequently led to endovascular stent placement. (D–F) 3D volume rendered CTA images after stent placement show overlapping uncovered stent placement (long arrow) extending from the LCCA to just past the LSCA. Note the persistent residual LSCA enlargement (short arrow). LCCA, left common carotid artery.
** Interruption of the aortic arch**

Interruption of the aortic arch is a rare, hemodynamically critical left-sided obstructive conotruncal anomaly in which there is discontinuity of the aortic arch, resulting in systemic perfusion that is dependent on the ductal and/or aortic branch artery. IAA accounts for 0.4% to 1.3% of CHD lesions and 7% of CHD lesions presenting with significant physiologic compromise. Arch interruption may occur at 1 of 3 levels with variable frequency: (1) type A, distal to the LSCA origin (26%); (2) type B, between the origins of the LCCA and LSCA (72%); and (3) type C, proximal to the LCCA, between the origins of the innominate and left common carotid arteries (2%). Associated CHD lesions include patent ductus arteriosus (PDA), ventricular septal defect (VSD; isolated and multiple), atrial septal defect (ASD), left ventricular outflow tract obstruction (LVOTO), such as hypoplasia and subaortic, valvular, and supravalvular aortic stenosis, aortopulmonary window, truncus arteriosus, transposition of the great arteries (TGA), double outlet right ventricle, and aortic arch anomalies.

Evidence indicates that type A IAA occurs secondary to decreased antegrade hemodynamics from an underlying CHD lesion (eg, LVOTO lesions, left to right shunts), with presumed left fourth aortic arch involution. By distinction, type B IAA results from genetically altered neural crest emigration. In patients affected with type B IAA, approximately 50% to 82% have a 22q11.2 deletion (most commonly Tbx1 haploinsufficiency) in which abnormal neural crest development and migration lead to an impaired epithelial remodeling of the fourth pharyngeal arch and derivatives of the third and fourth pharyngeal pouches. This condition accounts for the variably associated concomitant cardiovascular (eg, other conotruncal CHD lesions) and noncardiovascular phenotype expression (eg, DiGeorge syndrome, Velo-Cardio-Facial syndrome, conotruncal anomaly face syndrome). IAA is also associated with CHARGE syndrome, in which CHD7 gene mutation (8q12.1 chromosome) contributes to abnormal neural crest and fourth PAA epithelial morphogenesis.

Affected patients usually present clinically within the first few days of life and typically not more than 2 weeks of age. As with other critical LSOLs, the neonate with IAA may appear normal at birth and with no clinical cyanosis. However, as the ductus closes, systemic blood flow decreases and pulmonary venous pressure rises; congestive heart failure, respiratory distress, systemic hypoperfusion, pallor, and decreased organ function may ensue. Prompt hemodynamic stabilization, diagnosis, and surgical revision are mandatory for survival. Radiographically, IAA (and other LSOLs) shows pulmonary venous congestion, with the heart size ranging from normal to markedly increased. If a septal defect is present with left to right shunting, there may be a component of increased pulmonary vascularity. A narrow mediastinum in these patients indicates thymic aplasia (eg, DiGeorge syndrome). Echocardiography can usually define IAA with reliable accuracy. MR imaging-MR angiography or CTA may be indicated to further define the level of interruption, aortic branch arteries, and/or associated anomalies, which is key information for preoperative planning.

**Coarctation of the aorta**

Coarctation of the thoracic aorta is defined as a focal, eccentric, obstructive narrowing involving the aortic isthmus (Fig. 4). It accounts for 1.8% to 9.8% of CHD, with most studies showing an incidence of 5% to 6%. COA has a slight male predominance (1.2–2.3:1). Most cases occur sporadically, but both environmental factors and genetic causes may contribute. COA is characterized as preductal, juxtaductal, or postductal, based on its anatomic relationship to the ductus arteriosus. Preductal COA predominates in children less than 1 year of age, whereas the postductal type is more common in children greater than 1 year and in adults.

The narrowing in COA results from abnormal fibromuscular ductal tissue encircling the aorta. The ductal theory hypothesizes that, as the LSCA migrates cephalad through differential growth of the dorsal aorta, the ductal ostium from the sixth arch is pulled into the aorta, forming the circumferential sling. Obstruction at the isthmus develops when there is postnatal constriction of the ductus arteriosus. The hemodynamic theory facilitates a greater understanding of the pathogenesis and lends an explanation to the occurrence of COA with associated cardiovascular lesions. Congenital lesions with decreased antegrade flow in the ascending aorta (eg, LSOLs, left to right shunts), result in reversal of blood flow across the fetal isthmus, altering the branch point angulation, and accentuating LSCA cephalad migration, increasing the possibility for developing COA. In contrast, right-sided obstructive cardiac lesions (eg, right ventricular outflow tract obstruction, pulmonary stenosis, and pulmonary atresia) protect against coarctation, because there is dominant antegrade isthmus flow.

The primary physiologic sequelae of coarctation is increased left ventricular afterload and decreased systemic perfusion with activation of the
sympathetic and renin-angiotensin systems, resulting in increased blood pressure. In neonates and infants with COA (similar to IAA), systemic perfusion is dependent on maintaining patency of the ductus arteriosus. Prostaglandins are initiated early in the clinical course to maintain this patency. Once the ductus closes, the inability to rapidly develop collateral blood flow and counter the

Fig. 4. COA. A young child with upper extremity hypertension underwent CTA examination following echocardiographic diagnosis of COA. (A, B) 3D volume rendered images show severe narrowing involving the isthmic portion of the aorta (arrow). (C, D) 3D volume rendered images show well-developed collateral pathways, including internal mammary (C, long arrows), thoracodorsal (C, short arrows), superficial paraspinal (D, long arrows), and parascapular (D, short arrows) pathways. (E, F) 3D volume rendered images show intercostal (arrowhead) and mediastinal paraspinal (arrows) collateral arteries.
rising afterload may lead to left heart dysfunction with chamber enlargement and congestive heart failure. Pulmonary hypertension (with right heart dysfunction), renal insufficiency, and systemic shock may also develop. With isolated COA, when sufficient compensatory collateral flow develops to supply blood distal to the obstruction, clinical presentation is delayed until later in life. Primary collateral pathways include the subclavian, internal mammary, intercostal, cervical, scapular, and thoracodorsal arteries (see Fig. 4). The presence of collateral arteries and a pressure gradient are the distinguishing features between coarctation and pseudocoarctation (Fig. 5). In cases of pseudocoarctation, there is absence of a pressure gradient and collateral arteries.

Associated congenital cardiovascular abnormalities may occur in 44% to 84% of patients with COA; most of these patients present by 2 years of age.\textsuperscript{30,32,41} Commonly associated abnormalities include PDA, bicuspid aortic valve, left to right shunts (eg, ASD, VSD), LSOLs, and TGA. Syndromes and genetic disorders associated with COA include Shone complex, PHACE syndrome, Williams syndrome, Noonan syndrome, Turner syndrome (45 XO karyotype), trisomy 13, and trisomy 18.

Early diagnosis and intervention are essential to minimizing morbidity and mortality in pediatric patients with COA. Untreated COA in neonates and infants has a poor prognosis (50% mortality) without urgent surgical intervention.\textsuperscript{42} Milder forms of COA may take years or decades to become symptomatic. However, the long-term effects of systemic hypertension from aortic coarctation may lead to late cardiovascular complications, reducing life expectancy compared with the general population. Chronically increased blood pressure...
pressure results in left ventricular hypertrophy with congestive heart failure in early to midadulthood. The increased blood pressure can lead to an increased incidence of premature coronary artery disease, ischemic heart disease, cerebral vascular accidents (eg, ischemia, hemorrhage), aortic valvular disease (eg, stenosis or insufficiency related to a bicuspid aortic valve), aortic root dilation, aortic aneurysms (eg, proximal to a coarctation), acute aortic disease (eg, dissection, rupture), and bacterial endocarditis.

Definitive management of COA requires surgical (eg, resection with interposition graft) or endovascular (eg, angioplasty with stent placement) repair. Age at the time of coarctation repair has a predictive value for operative mortality (2%–41%, highest in infants <1 year old), and residual hypertension (12.5%–21%, lowest when operated between 1 and 5 years old). To optimize surgical outcome and minimize potential future cardiovascular risk, elective repair is recommended in early childhood (1–5 years of age) and should not be delayed past 10 years of age. Hypertension at the first postoperative evaluation and the development of postoperative paradoxical hypertension are risk factors for chronic hypertension (residual or recurrent) and acquired cardiovascular disease. Patients should be followed closely after repair for possible recurrent coarctation, progression of associated cardiac defects, and development of acquired cardiovascular disease.

Chest radiography has a low to moderate sensitivity for detection of COA, dependent on the age of the patient, the degree of narrowing, and the presence of associated cardiac defects. In the neonate and infant, the heart is typically enlarged and pulmonary venous congestion is present. In the older child to young adult with isolated COA, typical imaging findings include a prominent aortic arch and proximal descending aorta silhouette with a figure-of-3 contour, inferior rib scalloping and/or sclerosis, and a normal or mildly enlarged heart size. Confirmatory diagnosis of COA in neonates and infants is most often made with TTE, which has a sensitivity of 94% to 98% and a positive predictive value as high as 98% and 100% for neonates and infant, respectively. In older children to young adults, CEMRA (sensitivity 98%, specificity 99%) is superior to TTE for depicting COA and the complete thoracic arterial system. PC-MR imaging is applied to measure the gradient across the obstructive narrowing and quantify collateral flow. MDCT angiography is a highly sensitive imaging modality with an accuracy as high as 100%. As with MRA, collateral pathways should be described in detail because their number and extent directly correlate with the severity of disease.

### Aortic Arch Anomalies

Aortic arch anomalies (AAA) are congenital vascular abnormalities involving development of the primitive aortic arches and their derivatives, accounting for 0.5% to 1.6% of CHD lesions. Men have a slightly greater prevalence than women (1.2:1). Four percent of patients with these anomalies may have 22q11.2 deletion, whereas 35% of those with 22q11.2 deletion may have an isolated aortic arch anomaly. Common AAA in pediatric patients include vascular rings, pulmonary artery slings (PAS), and innominate artery compression (IAC). The most common symptomatic AAA are a double aortic arch (49%, range 36%–72%) and right aortic arch with a left ligamentum/ductus arteriosum (28%, range 8%–49%), followed by IAC (10%, range 3.3%–27%), left aortic arch with an aberrant right subclavian artery (8%, range 1.7%–20%), and pulmonary sling (5%, range 1.8%–12.5%). Associated CHD lesions occur in 18% (range 12%–32%) of patients with AAA, including VSD, ASD, PDA, right-sided obstructive lesions, and COA.

The unifying characteristic of these disorders is secondary vascular compression on the central airway, the esophagus, or both. Depending on the type of lesion, severity of compression, and the presence of comorbid cardiovascular and noncardiovascular congenital disease, affected patients present with variable degrees of respiratory and gastrointestinal symptoms during the neonatal period, infancy, childhood, or young adulthood. Respiratory symptoms are more prevalent among infants and young children, whereas esophageal symptoms are more common among older children, adolescents, and adults. In addition to extrinsic compression of the central airway, concomitant intrinsic tracheomalacia may occur in up to 53% of pediatric patients with AAA, affecting the presentation and clinical management of respiratory symptoms.

### Vascular rings

A vascular ring occurs when the trachea and/or esophagus are surrounded and compressed by vessels (eg, aortic arch or arches, aortic arch branch arteries, pulmonary branch arteries) and the ductus or ligamentum arteriosum. A vascular ring may be complete or incomplete. Vascular rings result from abnormal persistence and involution of primitive brachial arch segments, most commonly the third, fourth, and sixth arches. They may occur with a normal left-sided arch,
a double aortic arch (DAA), a right-sided aortic arch (RAA), and a cervical aortic arch (CAA). Classification of vascular rings is based on Edwards’ hypothetical embryologic double aortic arch model.

**Left aortic arch** The normal LAA, left-sided descending aorta, and left-sided ligamentum arteriosum are formed by regression of the right and persistence of the left fourth arches, eighth dorsal aorta segments, and sixth dorsal arches, respectively. Two main anomalous patterns may occur. The first is an aberrant right subclavian artery (Fig. 6), which results when right fourth arch regression occurs between the right common carotid and right subclavian arteries, rather than distal to the right subclavian artery. The aorta gives rise to the right common carotid, the left common carotid, the left subclavian, and the right subclavian arteries. The right subclavian artery courses retroesophageal. Two subdivisions are possible, namely a left-sided or right-sided ligamentum arteriosum. If there is a left-sided ligament, no vascular ring occurs; the aberrant right subclavian artery has smooth caliber throughout its course. However, a rare right-sided ligament completes a true vascular ring; it forms by persistence of the right dorsal sixth arch and passes from the right pulmonary artery (RPA) to the descending aorta after the retroesophageal segment. The second anomalous pattern with a LAA is a circumflex right descending aorta. With this entity, distal aortic arch and proximal descending aorta course posterior to the esophagus and trachea. It may occur with normal arch branching or an aberrant right subclavian artery. The vascular ring is completed by the ligament passing between the RPA and the descending aorta after the retroesophageal segment.

**Double aortic arch** A double aortic arch (Fig. 7) results from persistence of both the right and left fourth arches and the dorsal aorta, forming a complete vascular ring. Right dominance occurs in 66% (range 37%–81%), left dominance in 16% (range 10%–20%), and codominance in 17% (range 3%–53%). The left arch may be atretic with a fibrous segment distal to the take-off of 1 or both of the left arch branch arteries. Rarely, the right arch may be atretic. The right arch typically supplies the right common carotid and brachiocephalic arteries, whereas the left supplies the left common carotid and brachiocephalic arteries. The descending aorta is often on the left but the proximal descending aorta can be on the right or midline. The ligament is usually left sided. Less commonly, it can be right sided or bilateral. Pulmonary arteries are typically normal.

**Right aortic arch** A right aortic arch occurs from persistence of the right, and regression of the left, fourth arches and eighth dorsal aorta segments, respectively. A vascular ring may occur depending on the level of the left fourth arch resorption, the origins and course of the ligamentum arteriosum and the descending aorta, or a combination thereof. Three possible patterns of a RAA may occur. The first is an aberrant LSCA/brachiocephalic artery. In this case, left fourth arch regression occurs between the left carotid

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**Fig. 6.** Left aortic arch with an aberrant right subclavian artery. A young child underwent CTA for suspected pulmonary embolism. *(A, B)* 3D volume rendered images show a left aortic arch with an aberrant right subclavian artery (RSCA; arrow). The RSCA arises from the aorta as the fourth aortic branch artery, after the right common carotid artery (RCCA), LCCA, and LSCA.
and subclavian arteries. The aorta gives rise to the left common carotid, the right common carotid, the right subclavian, and the left subclavian arteries; the LSCA courses retroesophageal (Fig. 8). Regression before the LCCA results in an anomalous left brachiocephalic artery. The right carotid artery becomes the first branch followed by the right subclavian artery and the left brachiocephalic artery. The left brachiocephalic artery courses retroesophageal with subsequent branching into the left common carotid and subclavian arteries. In both types, when the ligament is right sided, no vascular ring is present. The left subclavian or brachiocephalic artery has a regular caliber throughout its course. When the ligament is left sided, a complete vascular ring is present; the ligament courses between the LPA and the proximal descending aorta, via the aberrant LSCA or brachiocephalic artery (Fig. 9). The retroesophageal segment of the left subclavian or brachiocephalic artery is dilated with a diverticulum of Kommerell in 15% to 21% of cases.60,62

The second pattern with a RAA is mirror image branching. In this condition, left fourth arch regression occurs distal to the LSCA and the aorta gives rise to the left brachiocephalic, the right common carotid, and the right subclavian arteries. In most instances, no vascular ring is present because the descending aorta and ductus/ligamentum arteriosum are ipsilateral. The right ligament passes between the right descending aorta and the RPA. Less commonly, the ductus is contralateral. In this instance, the ligamentum usually passes between the brachiocephalic artery and the LPA without formation of a vascular ring. Rarely, however, the ligamentum arises from the proximal descending aorta and takes a retroesophageal course to the LPA, creating a vascular ring. Although the ligament may not be visualized, a small leftward-facing dimple may be present on the proximal descending aorta, at the take-off of the ligament, indicating the anomalous ligament.

The third pattern with a RAA is circumflex left descending aorta. With this entity, the descending aorta courses posterior to the esophagus and then descends on the left side, analogous to a left aortic arch with a right descending aorta. A circumflex left descending aorta can occur with an anomalous LSCA, an anomalous left brachiocephalic artery, and mirror image branching. A vascular ring is completed by a left ligamentum arteriosum coursing between the descending aorta and the LPA.

**Cervical aortic arch** A CAA occurs when the aortic arch is positioned above the thoracic inlet. This condition results when the third arch is the basis for aortic arch development, rather than the fourth. CAA may occur with both right and left third arches, leading to the potential formation of left-sided, right-sided, and double AAA. Most commonly, it occurs on the right side with a persistent right third aortic arch and right dorsal aorta.68
Pulmonary artery sling
PAS is a condition in which the LPA typically arises from the RPA and courses between the trachea and esophagus toward the left lung (Fig. 10). PAS develops as a result of proximal left sixth arch involution. Although right and left ligaments are possible, only a left ligament leads to a complete vascular ring. In this instance, the left ligament connects between the main or RPA and the left descending aorta. PAS is often associated with concomitant central airway anomalies and acquired abnormalities, including tracheal rings, right upper lobe tracheal bronchus, and tracheomalacia. Relief of symptoms requires transposing the LPA and correction of potential central airway anomalies or acquired abnormalities.

Innominate artery compression
IAC on the trachea occurs when the anterior crossing innominate artery extrinsically compresses the upper to midanterior trachea, often in an oblique manner (see Fig. 1). Although IAC does not constitute a traditional vascular ring, it can cause significant respiratory symptoms, particularly in infants and young children, as discussed.

Fig. 8. Right aortic arch with an aberrant LSCA. A young child with recurrent aspiration and respiratory distress underwent MR angiography for evaluation of a vascular ring. (A–C) 3D volume rendered images show a right aortic arch with an aberrant LSCA (short arrow), a diverticulum of Kommerell (long arrow), and a left ligamentus arteriosum resulting in a complete vascular ring. (D, E) Transverse dark blood images show an approximate 50% tracheal compression (asterisk) by the retroesophageal LSCA (arrow).
in an article by Lee and colleagues elsewhere in this issue. IAC often results when there is accentuated horizontal angulation of the ascending aorta and compensatory angulation of the aortic arch and branch arteries. MRI-MRA or CTA can directly depict the innominate artery caliber and course as well as the degree of tracheal narrowing, which may vary from a shallow asymmetric indentation to marked anterior-posterior compression of greater than 50%. IAC has a high association with intrinsic tracheomalacia, which should be considered when choosing the imaging modality, selecting the MRI-MRA or CTA protocol, and interpreting the examination.1

Pulmonary Arterial Anomalies

In addition to PAS, congenital pulmonary arterial anomalies include hypoplasia, agenesis (Fig. 11), and stenosis. These anomalies most often occur in association with complex conotruncal congenital heart lesions, such as pulmonary valve stenosis, pulmonary valve atresia, tetralogy of Fallot, and TGA.69 Isolated pulmonary arterial hypoplasia and agenesis, in the absence of complex CHD, are often associated with pulmonary hypoplasia and hypogenetic lung syndrome (Fig. 12). Isolated pulmonary arterial stenosis (PAS) is rare. Such stenoses most commonly involve the central main and branch pulmonary arteries, but
Fig. 11. Pulmonary artery agenesis. A neonate with mild respiratory distress underwent CTA following an abnormal chest radiograph. (A) Frontal chest radiograph shows dextroposition of the heart and mild right lung oligemia with asymmetric left greater than right pulmonary vascularity. (B) 3D volume rendered image shows RPA agenesis. Pulmonary blood flow (PBF) is thus from the main pulmonary artery (MPA) to the LPA. (C) Coronal MIP image shows right pulmonary arterial reconstitution at the lobar level from systemic to pulmonary collateral arteries (arrows).

Fig. 12. Hypogenetic lung syndrome with partial pulmonary venous return to the IVC (scimitar syndrome). An adolescent with respiratory exercise intolerance underwent MDCT angiography. (A) Frontal chest radiograph shows increased left pulmonary vascularity and an enlarged heart with either dextrocardia or dextroposition of the heart. (B) 3D volume rendered image confirms dextroposition of the heart. (C) Coronal inverse MinIP image shows hypoplasia of the right lung with absence of the right upper lobe bronchus. (D) Coronal MIP image shows hypoplasia of the RPA (long arrow) and partial anomalous pulmonary venous return of most of the right lung into the IVC via 2 venous channels (short arrows). (E) Oblique thin-slab MIP image shows the hypoplastic RPA with compensatory LPA enlargement related to dominant PBF. (F, G) Cardiac 4-chamber (F) and short-axis (G) multiplanar reformations show an enlarged right atrium (RA) and right ventricle (RV) related to the right heart volume overload. LA, left atrium; LV, left ventricle. (H, I) 3D volume rendered images show the anomalous right lung venous drainage (short arrows) into the IVC. A small native right middle lobe vein (long arrow) drains directly into the LA.
may occur in peripheral segments (eg, lobar and segmental divisions). Lesions may be diffuse with long segments of disease, resembling hypoplasia, or may be focal or multifocal with poststenotic dilatation. Possible underlying causes for PASt include congenital rubella, Williams syndrome, Noonan syndrome, Alagile syndrome, Ehlers-Danlos syndrome, and cutis laxa. The common pathologic finding is abnormal development of the elastic tissue of the media and increased collagen and fibrous tissue. Secondary intimal proliferation occurs with the risk of localized thrombosis. Progressive and long-standing PASt increases right ventricular afterload (pressure), leading to right ventricular hypertrophy, strain on the tricuspid valvular apparatus, and potentially tricuspid insufficiency and right heart failure.

VENOUS SYSTEMS
Systemic Venous Anomalies

Normal thoracic systemic venous anatomy consists of bilateral subclavian (SCV) and brachiocephalic (BCV) veins, draining to the right superior vena cava (RSVC) and then into the sinus venosus portion of the right atrium. Embryologically, these veins develop from the paired anterior cardinal and common cardinal veins. A communicating vein forms from the superior transverse capillary plexuses and directs blood from the left to right anterior cardinal vein. The entire right anterior cardinal vein normally persists, forming the right SCV (RSCV) and right BCV (RBCV) as well as the right internal jugular vein. The right SVC (R SVC) develops from the right anterior cardinal and
common cardinal veins, which enter the sinus venosus. In distinction, the left anterior cardinal vein undergoes near complete atrophy below the level of the communicating vein, forming the ligament of the left vena cava. A residual central venous segment forms the left superior intercostal vein, which drains the second and third intercostal spaces. The remainder of left anterior cardinal vein persists to form the left SCV and internal jugular vein. The communicating vein becomes the left BCV (LBCV) and enlarges to accommodate increased left to right flow. The LBCV courses obliquely downward superior to the aortic arch and anterior to the supra-aortic branch arteries, joining with the RBCV to form the RSVC. The left common cardinal vein is incorporated into the coronary venous anatomy, becoming the oblique vein of the left atrium (vein of Marshall), draining into the coronary sinus of the right atrium.3

Anomalies of these systemic veins are rare. Systemic venous anomalies may occur in isolation or may be associated with cardiac disease, in particular CHD and arrhythmias. Three of the more common systemic venous anomalies are (1) a persistent left SVC (LSVC) with a native RSVC; (2) a LSVC with mirror image venous drainage (Fig. 13); and (3) a retroaortic left BCV (RA-LBCV, Fig. 14). Clear understanding of these systemic venous anomalies can facilitate planning and placement of central venous catheters, hemodynamic monitoring devices, and cardiac pacemaker and cardioverter-defibrillator leads, as well as assessment of their respective positioning on chest radiographs following placement. Recognition of a LSVC on echocardiography, MRI-MRA, or CTA is important for cardiac preoperative planning and surgical management, including cardiopulmonary bypass and procedures for CHD (eg, cavopulmonary shunts).

**Persistent left superior vena cava**

A persistent left superior vena cava (LSVC) has a prevalence of 0.1% to 0.5% in the general population.70,71 It occurs more frequently in patients with CHD, with a reported prevalence of 1.3% to 5%.72–74 Commonly associated CHD anomalies include septal defects (ventricular, atrial, and atrioventricular), tetralogy of Fallot, pulmonary atresia, bicuspid aortic valve, AS, aortic coarctation, PDA, and anomalous pulmonary venous return.72,74 Extracardiac congenital anomalies may be found in 60% of pediatric patients with a persistent LSVC. Commonly associated disorders include VACTERL (vertebral anomalies, anal atresia, cardiovascular anomalies, tracheoesophageal fistula, esophageal atresia, renal (kidney) and/or radial anomalies, limb defects), trisomy 21, 22q11 deletion, CHARGE (coloboma of the eye, central nervous system anomalies, heart defects, atresia of the choanae, retardation of growth and/or development, genital and/or urinary defects, ear anomalies and/or deafness) syndrome, and Turner syndrome.74 Embryologically, a persistent LSVC results from persistence of the left anterior cardinal vein. Concurrent persistence of the right anterior and common cardinal veins yields bilateral SVCs with or without a communicating brachiocephalic vein (ie, bridging vein).

**Left superior vena cava with mirror image venous drainage**

Involution of the right common cardinal vein and the central anterior cardinal vein, along with

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**Fig. 13.** Persistent left SVC with mirror systemic venous drainage. A young child with a known ASD underwent MDCT angiography after abnormal chest radiograph. (A) Frontal chest radiograph shows a prominent left paramediastinal shadow (arrows). (B) Coronal MIP from a direct CT venogram (CTV) shows a persistent LSVC draining into the coronary sinus (CS). (C) Coronal 3D volume rendered image obtained during the delayed CTV phase shows mirror image systemic venous drainage with a right brachiocephalic vein (RBCV) draining to the LSVC. The LSVC is again noted to drain to the CS.
persistence of the communicating vein and the left anterior cardinal vein, results in mirror image thoracic systemic venous drainage (ie, right BCV and an LSVC). In most instances, the LSVC drains into the coronary sinus and the coronary sinus enlarges to accommodate flow.74 Coronary sinus dilatation may potentially lead to left atrioventricular valve inflow obstruction, which in turn may lead to cardiac arrhythmias and/or sudden death. In addition, abnormal cardiac impulse formation and conduction may arise from abnormal morphologic development of the sinoatrial node, atrioventricular node, and bundle of His (eg, lengthening).70 Rarely, the LSVC drains into the left atrium (ie, complete unroofing of the coronary sinus), leading to interatrial communication, right to left shunting, and cyanosis.74

**Retroaortic left brachiocephalic vein**

The retroaortic LBCV is found in 0.5% to 0.6% of patients with CHD75,76 and in only 0.02% of patients who do not have CHD.76 It courses posterior to the ascending aorta, underneath the aortic arch, and anterior to the central main and right pulmonary arteries, to join the RSVC at or below the ostial confluence of the ayzygous vein. Although it results in neither physiologic nor hemodynamic sequelae, a retroaortic left brachiocephalic vein is associated with congenital heart disease, including right side obstructive lesions (tetralogy of Fallot, pulmonary atresia), truncus arteriosus, and AAA (eg, IAA, RAA, CAA). Embryologically, it occurs when the developing communicating vein anastomoses with the right inferior transverse capillary plexus.75

**Pulmonary Venous Anomalies**

The lung buds, developing from the foregut, initially drain via a venous plexus into the cardinal venous system.3 A common pulmonary vein (CPV) arises from the dorsum of the left atrium and anastomoses with the venous plexus. Connections to the cardinal venous system involute with subsequent direct pulmonary venous drainage into the left atrium. CPV absorption into the wall of the left atrium results in variable pulmonary vein ostia and branching patterns, with a standard of single, bilateral superior and inferior pulmonary vein trunks.

Abnormal development of the CPV (eg, incomplete resorption) and its anastomosis with the primitive venous plexus, along with persistent connections to the systemic cardinal veins, give rise to partial anomalous pulmonary venous return (PAPVR) or total anomalous pulmonary venous return (TAPVR; 0.7%–3.2% of CHD).8,26–28 Both PAPVR and TAPVR result in a left to right shunt with partial (PAPVR) or complete (TAPVR) admixture of deoxygenated and oxygenated blood. PAPVR and TAPVR may be associated with other
congenital heart diseases, including heterotaxy; atrial, ventricular, and atrioventricular septal defects; tetralogy of Fallot; and COA.\(^77\) Anomalous pulmonary venous return should be distinguished from pulmonary veins, which have anomalous peripheral connections and/or an aberrant course (eg, aberrant meandering vein) within the lungs, before normal drainage into the left atrium.\(^78\) In most instances, echocardiography with gray scale and Doppler interrogation depicts the number, location, and course of pulmonary veins; detects the direction of venous blood flow; and excludes flow obstruction. MRI-MRA or CTA may be required when echocardiography cannot identify all veins or when more comprehensive evaluation is required following the diagnosis of PAPVR or TAPVR. MR imaging offers the advantage of quantifying the shunt ratio (eg, Qp/Qs) using phase contrast imaging. CTA is advantageous for its superior visualization of the central airway and lung in the evaluation of associated pulmonary developmental anomalies.

**Partial anomalous pulmonary venous return**

In PAPVR, 1 or more (but not all) of the pulmonary veins (segmental, lobar, or main central trunk) drain directly into systemic veins (eg, SVC, inferior vena cava [IVC], SCV, BCV, azygous vein), right atrium, or coronary sinus. Hemodynamic sequelae reflect the degree (eg, number and size of anomalous veins) and duration of shunting. When the shunt is significant, flow across the right heart and pulmonary circulation is increased (eg, increased pulmonary blood flow [PBF]). The right cardiac chambers and pulmonary vasculature (arteries and veins) are enlarged because of volume overload. Pulmonary hypertension and right heart failure may subsequently develop. Increased left atrial pressure may lead to pulmonary venous congestion; initially acyanotic, patients may become cyanotic. The right pulmonary veins are anomalous twice as often as the left. Right upper lobe venous drainage into the SVC is the most common type of PAPVR (Fig. 15) and may be associated with a sinus venosus defect (Fig. 16). The second most frequent type of PAPVR is left pulmonary venous drainage to the LBCV. The third most common form is anomalous drainage from the right lung to the IVC with an intact atrial septum. This condition may be associated with more complex pulmonary developmental anomalies in the spectrum of congenital pulmonary venolobar syndrome, including bronchopulmonary sequestration (BPS), scimitar syndrome (see Fig. 12), and horseshoe lung. Most associated BPS are extralobar with venous drainage into azygous or hemiazygous veins. In scimitar syndrome, also known as hypogenetic lung syndrome, an anomalous CPV (scimitar vein) drains a portion or the entire lung into the IVC either above or below the diaphragm. Alternatively, drainage may occur into hepatic veins, portal veins, azygous vein, coronary sinus, or right atrium. The right lung is almost exclusively involved and has variable hypoplasia versus partial agenesis, associated with dextroposition of the heart and ipsilateral decreased pulmonary perfusion relative to the left. Additional associated anomalies include bronchogenic cyst, BPS, horseshoe lung, accessory diaphragm, and congenital diaphragmatic hernia. Pulmonary arteries to the affected lung may have variable hypoplasia or agenesis, with or without systemic arterial supply in the absence of an associated sequestration.

**Total anomalous pulmonary venous return**

In TAPVR, all pulmonary veins have anomalous drainage. Affected patients typically present in the neonatal period. An obligatory ASD or patent foramen ovale is often present, leading to a right to left shunt and cyanosis. Veins may drain via a common vein into systemic veins or the right atrium (via the coronary sinus). Alternatively, veins may first drain into a venous confluence and then to a common draining vein. Depending on the level of anomalous connections, TAPVR drainage may be categorized as supracardiac (type I, Fig. 17), infracardiac (type II), intracardiac (type III), or mixed (Figs. 18 and 19). In the common form of type I TAPVR, the pulmonary veins are most commonly drained by a left ascending vertical vein to the LBCV and then the SVC, resulting in increased PBF, cardiomegaly, and a wide mediastinum radiographically. Other sites of supracardiac systemic connection are the SVC (right or left, see Fig. 17) and azygous vein. In type 2 TAPVR, the anomalous pulmonary veins drain into the coronary sinus (see Fig. 18B–D), leading to increased PBF, cardiomegaly, and a narrow mediastinum radiographically. Venous flow in types I and II TAPVR is unobstructed. Affected neonates often develop right heart volume overload, pulmonary hypertension, and right heart failure. In type 3 TAPVR (see Fig. 19B, C), anomalous veins drain via a descending vertical vein into the portal, hepatic, or mesenteric venous systems, with flow obstruction at or below the diaphragm, pulmonary venous congestion, a normal to small heart size, and a narrow mediastinum radiographically. This type constitutes a neonatal cardiopulmonary emergency because there is diminished cardiac output, poor systemic perfusion, and even greater cyanosis. The mixed type consists of combinations of types I to III TAPVR.
Fig. 15. Partial pulmonary venous return with intact atrial septum. An adolescent with exertional chest pressure and mild hypoxia underwent MDCT angiography. (A) Frontal chest radiograph shows moderate cardiomegaly with prominent central pulmonary vascularity. (B) Four-chamber multiplanar reformation shows an enlarged RA and RV with flattening of the interatrial and interventricular septae, related to the right heart volume overload. (C, D) 3D volume rendered images show right upper lobe segmental pulmonary veins (arrows) draining to the SVC (asterisk) with all other pulmonary veins draining into the LA.

Fig. 16. Partial pulmonary venous return with sinus venosus defect. Cardiac MR imaging was performed in a young child with exertional shortness of breath and mild hypoxia. (A, B) Bright-blood MR angiography MIP images show right upper lobe anomalous pulmonary venous return into the SVC, above (arrows) or at the cav-oatrial junction (arrowhead). A septal defect is present along the sinus venosus portion of the interatrial septum (asterisk), leading to communication between the RA and LA.
Fig. 17. TAPVR (Supracardiac Type I). A neonate with complex congenital heart disease underwent MDCT angiography. (A, B) 3D volume rendered images show bilateral anomalous drainage of all pulmonary veins (arrows) into a left SVC (LSVC). Blood then flows to the CS.

Fig. 18. TAPVR (mixed type I and II). A neonate with anomalous pulmonary veins diagnosed by echocardiography underwent low-dose MDCT angiography. (A–D) Variable thick, MIP images show that the left upper pulmonary veins drain into the LBCV (A, short arrow), whereas the left lower (B, D; arrowheads) and all right lung pulmonary veins (B, C; long arrows) drain to a retrocardiac confluence (B, asterisk). From the common confluence, blood drains (C, D, asterisks) into the CS.
SUMMARY

Diagnostic imaging is crucial in the evaluation of thoracic congenital arterial and venous anomalies in pediatric patients. Although clinical assessment may provide insight into the possible diagnosis, imaging is usually necessary to confirm the diagnosis. Initial imaging algorithms using noninvasive modalities often begin with chest radiography followed by echocardiography. State-of-the-art MRI-MRA and MDCT angiography with advanced 3D visualization are essential in these algorithms not only for diagnosis but also treatment planning and postoperative and postendovascular evaluation. Selection between MRI-MRA and CTA should be based on their respective advantages, balanced by the suspected or known congenital vascular anomaly and patient’s clinical presentation and hemodynamic stability. Understanding the embryologic basis and anatomic characteristics of thoracic congenital vascular anomalies assists in their recognition with MRI-MRA and MDCT angiography in pediatric patients.

REFERENCES


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