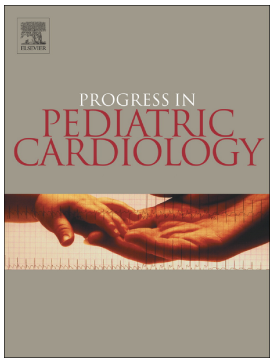

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Progress in Pediatric Cardiology

Multi-Modality Assessment of the Aortic Arch Branching and Vascular Rings

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Abstract

In congenital heart disease, echocardiography plays an essential role in the assessment of the aortic arch. The primary role of the cardiac sonographer in aortic arch evaluation, is to determine arch sidedness, branching pattern and the patency of the aortic arch. Aortic arch variations are commonly encountered and are often initially suspected by echocardiogram prior to additional confirmatory cross-sectional imaging. Herein, we will present a systematic approach for aortic arch assessment and discuss the role for advanced cardiac imaging (computed tomography angiography and magnetic resonance angiography) with vascular rings and common aortic arch branching abnormalities. Discussion of aortic coarctation and interrupted aortic arch, are beyond the scope of this article.

Keywords

Vascular ring

Aortic arch anomalies

Congenital heart disease

Echocardiography

Magnetic resonance imaging

Computed tomography angiography

Highlights

-vascular rings often need cross sectional imaging for confirmation

-Multiple different types of vascular rings exist

-vascular rings are commonly associated with other cardiac and genetic findings

-echocardiography is often the first line imaging tool by evaluating arch branching

Introduction

The aorta is the arterial conduit that allows oxygenated blood from the left ventricle to supply the brain and entire body. Anatomical variations in the aortic arch may lead to a diverse array of clinical symptoms. These may range from cardiogenic shock with critical coarctation or interrupted aorta; stridor or dysphagia with a vascular ring, or no symptoms in patients with a left aortic arch with an aberrant right subclavian artery. Echocardiography plays an essential role in the assessment of the aortic arch. Clinicians and cardiac sonographers need a systematic approach to the evaluation of an aortic arch. This should include assessment of the arch sidedness, branching pattern and the patency of the aortic arch. They should also be familiar with common aortic arch variations like vascular rings and arch branching abnormalities that can cause compression of the trachea or esophagus and lead to clinical symptoms. Some aortic arch anomalies may require advanced cardiac imaging (computed tomography angiography / magnetic resonance angiography) to make a confirmatory diagnosis or assess the relationship of other intrathoracic structures prior to surgical consideration. This review will present a systematic approach for aortic arch assessment and discuss the role for advanced cardiac imaging (computed tomography angiography and magnetic resonance angiography) with common aortic arch anomalies. Discussion of aortic coarctation and interrupted aortic arch, are beyond the scope of this article.

Embryological development of the aortic arch.

In order to understand the anatomical variations seen in the aortic arch, practitioners require a thorough understanding of the embryological development of the cardiovascular system. The heart is the first organ formed and starts beating around week four of gestation (1).

A primitive heart tube develops at week three of gestation with four unique segments that are destined for differentiation: primitive atria (atria) – primitive ventricle (ventricles) – bulbus cordis (smooth portion of ventricles and infundibulum) - truncus arteriosus (great arteries). Six primitive aortic arches develop from the trunco-aortic sac at the superior portion of the heart tube, with a pair of branches (right and left) traveling within each arch and ending in the dorsal aorta. All six pairs of arches are not simultaneously present; they develop and regress at different stages. While the 1st and 2nd arches form the arteries of the face and ears, and the 5th arch usually disappears, while the paired 3rd, 4th, and 6th arches as well as the paired 7th intersegmental arteries contribute to the creation of a primitive embryologic double aortic arch system (2, 3). The easiest way to understand aortic arch anomalies is to consider every aortic arch as originating as a double arch and then undergoing spontaneous regression of some aspect of the ring to allow for the arch anatomy seen. Figure 1 demonstrates double aortic arch anatomy with paired right and left arches encircling the esophagus and trachea. The illustration line breaks indicates regression of that arch segment. For example, a break at line 1 will result in a left aortic arch. This was originally described by Dr. Jesse E. Edwards - Edwards' double aortic arch model (4).

In the normal left aortic arch with normal branching, the right aortic arch undergoes spontaneous regression after the right subclavian artery (Figure 1 with line break 1) leading to a persistent left aortic arch with a first branch as an innominate artery (right subclavian artery and right carotid artery), second branch as the left carotid artery and third branch as the left subclavian artery. In this arrangement, the embryological remnants of the right 3rd arch give rise to the right common carotid artery. The right subclavian artery is derived from contributions from the right 4th arch, the right 7th intersegmental artery, and the right dorsal aorta. The left

3rd arch gives rise to the left common carotid artery, while the left 4th arch develops into the definitive aortic arch between the left common carotid and the left subclavian artery. The ductus arteriosus arises from paired 6th arches and dual patent ductus arteriosus can persist if the right 6th arch does not regress appropriately. Therefore, in the typical left aortic arch, dissolution of the right 6th aortic arch (ductus arteriosus) and the right dorsal aorta distal to the origin of the right 7th intersegmental artery (precursor to the right subclavian artery), results in the typical branching pattern (2,5). When specific arches fail to regress, they can persist into the post-natal period, giving rise to a multitude of variants that will be further discussed below.

Multi-modality Assessment of the Aortic Arch

a. Echocardiography

Echocardiography can non-invasively provide complete visualization of the aortic arch within the thoracic cavity. To adequately image the aortic arch, imaging in the supraclavicular region, especially within the suprasternal notch, is the preferred location. This can be accomplished by placing the child in a supine position with the neck hyperextended. Placing a rolled towel or pillow under the patient's shoulder blades can provide comfort for the patient while exposing the suprasternal notch when imaging. A systematic approach should be taken by all sonographers when evaluating an aortic arch:

1. Define the arch sidedness
2. Define the branching pattern of the aortic arch
3. Assess the patency of the aortic arch

Step 1: The sonographer should define the arch sidedness. The arch is either rightward or leftward relative to the trachea and the main-stem bronchus. In the normal branching pattern, the

first branch (innominate artery or brachiocephalic trunk /artery) usually travels to the side opposite to that of the aortic arch. Imaging for arch sidedness and branching begins in the suprasternal notch views. Place the transducer in the supraclavicular (suprasternal notch) region with the transducer indicator at the 3 o'clock position (towards patients left side) in order to obtain an orthogonal – short axis view of the aorta. Start inferiorly to show the ascending aorta, superior vena cava, right pulmonary artery, and left atrium. From this position sweep or angle superiorly towards the head to demonstrate branching pattern. Following the course of the first vessel off the aortic arch until branching is demonstrated is the maneuver of choice. Branching of the first arch vessel to the right, indicates a left aortic arch. Branching of the first arch vessel to the left indicates a right aortic arch. Another maneuver is a tracheal sweep. From a suprasternal long axis view, a tracheal sweep can be performed to establish arch sidedness. A left aortic arch can be established by starting rightward and slowly sweeping left. In the setting of a left aortic arch, the trachea and its rings will be visualized prior to visualizing the entirety of the aortic arch. In the setting of a right aortic arch, the aortic arch will be visualized prior to visualizing the trachea. Additionally subcostal imaging is another useful view in identifying the position of the descending aorta relative to the spine. There are some exceptions such as the retro-esophageal innominate artery that can arise as the last branch from the aortic arch. In some patients, the origin of both carotid arteries may be close to each other, and thus making it difficult to determine (6).

Step 2: Define the branching pattern of the aortic arch. The left aortic arch lies to the left of the trachea and courses over the left main-stem bronchus. In the classical anatomical configuration, the aortic arch (AA) is left sided and the most common branching pattern is as follows: first the innominate artery (brachiocephalic trunk), then the left common carotid artery,

and finally the left subclavian artery from right to left. The brachiocephalic trunk branches into the right subclavian artery and right common carotid artery (Fig.2A, B, and C). This branching pattern occurs in 65–94% of cases and is the classic "normal" branching pattern (7). There are two frequent anatomical variants of the left arch. Approximately 30% of the population may have a "bovine arch" (8). This occurs when the innominate artery and left common carotid artery have a common origin from the aortic arch (Fig.3A, B). Another common variant is a separate origin of the left vertebral artery directly from the transverse arch proximal to the left subclavian artery. The left vertebral artery usually arises from the left subclavian artery. This arrangement creates the appearance of four vessel arising from the aortic arch (innominate – left carotid – vertebral – left subclavian arteries) versus the traditional three vessels (innominate – left carotid – left subclavian arteries) as seen in Figure 4 (6).

Step 3: Once arch sidedness and branching have been established, the arch should be interrogated for patency. Traditional two dimensional imaging in the standard suprasternal orthogonal and sagittal planes (short and long axis windows) should be completed along with color Doppler and spectral Doppler interrogation. These are important to evaluate the direction of flow, pattern of flow and site of any anomalies within the arch. To obtain the "candy cane view" of a left aortic arch in the long axis view (sagittal plane), the transducer is placed in the suprasternal notch with the indicator at the 12-1 o'clock position (Figure 2B). This will allow the sonographer to assess the patency of the aortic arch. Complete assessment at our institution, includes two-dimensional imaging, color Doppler, pulse Doppler and continuous wave Doppler in the ascending, transverse and descending aorta. In addition, all patients should have two-dimensional imaging, color Doppler, pulse Doppler of the abdominal aorta from the subcostal view to assess for hemodynamically significant coarctation. This would typically be manifested

by a dampened systolic pulse and/or continuous wave Doppler pattern with continuous flow into diastole.

Although echocardiography is the most universally accepted form of evaluating the aortic arch anatomy, there are some limitations. These include an inability to adequately detect atretic segments without visible flow and difficulty in to visualizing the airway and inability to fully assess the related intrathoracic structures (trachea and esophagus). Additionally, many pediatric patients are unable to cooperate with imaging from the suprasternal notch due to their age, fear or developmental delays, leading to suboptimal windows and the inability to acquire images sufficient for complete assessment of the aortic arch and its branches.

b. Advanced cardiac imaging

Computed tomography angiography and magnetic resonance angiography provide three-dimensional visualization of the vascular anatomy, which can accurately assess the aortic arch in two and three dimensions. This allows clinicians the ability to accurately detect arch anomalies, understand the relationship of adjacent intrathoracic structures (trachea and esophagus) and gives detailed anatomic surveillance for interventional or surgical planning if indicated. Figure 2C demonstrates an example of a normal left aortic arch by computed tomography showing the arch coursing leftward of the trachea. When determining which modality to use, one must consider availability of the modality, expertise with using the modality and the associated risks and benefits. While computed tomography angiography carries risks associated with iodinated contrast administration and exposure to ionizing radiation, the modality is more available than magnetic resonance imaging and has the benefit of rapid image acquisition. The speed of image acquisition with computed tomography angiography decreases the need for sedation, which carries its own risks and has a greater impact on the pediatric population as they can be agitated,

irritable and poorly compliant with adequately obtaining the study. In a patient who cannot remain immobile for magnetic resonance angiography and is not a suitable candidate for sedation or anesthesia, the rapid scan time of computed tomography angiography would be most beneficial. A further strength of computed tomography angiography is the higher spatial and temporal resolution as compared to magnetic resonance imaging which produces accurate visualization of the airways. In cases of suspected tracheo-bronchomalacia or complete tracheal rings in association with a vascular ring or sling, computed tomography angiography is the modality of choice (9).

In comparison to computed tomography angiography, magnetic resonance imaging allows for simultaneous multi-planar imaging of associated cardiac abnormalities and can be used to evaluate the trachea, albeit with less spatial resolution compared to computed tomography angiography. Additionally, magnetic resonance imaging can assess the area of concern using multiple different sequences for confirmation. This benefit is an advantage over computed tomography angiography where the image can be degraded by motion or inaccurate bolus timing. Cardiac magnetic resonance angiography is not associated with iodinated contrast administration or exposure to ionizing radiation and may be safer in patients with renal dysfunction or those that require serial studies. Despite these benefits, magnetic resonance angiography is not a risk-free modality for vascular imaging. The magnetic resonance imaging scanner can induce anxiety or claustrophobia, is noisy, can require periodic breath holding to optimize image quality and typically can last 30-60 minutes depending on the imaging protocol used. As such, magnetic resonance angiography examinations for aortic anomalies can increase the need for sedation in younger pediatric patients (5).

Congenital variants and anomalies of the aortic arch

A complete and thorough assessment of the aortic arch is essential in all pediatric patients to exclude critical forms of congenital heart disease with coarctation; and to detect aortic arch anomalies that may lead to vascular rings or non-ring vascular compression of the esophagus or trachea. Identification of these anomalies can have important implications for prognosis and management during surgical and percutaneous interventions. The prevalence of aortic arch malformations varies between 1–2% among the general population (10). Vascular rings are formed when vessels (or their atretic portions) completely encircle the trachea and esophagus, resulting in the potential for airway and/or esophageal compression (8). Clinically this may present as stridor or noisy breathing in infants, recurrent lung infections, respiratory distress, dysphagia (swallowing difficulty), or choking, especially when swallowing large chunks of meats or other solid foods. Aortic arch anomalies can occur in isolation or may co-exist with various forms of congenital heart disease or chromosomal abnormalities. Approximately 12 percent of patients with vascular rings will have associated congenital heart disease with the most common forms being ventricular septal defect and tetralogy of Fallot. DiGeorge syndrome is a microdeletion of chromosome 22q11.2 and characterized by a high incidence of midline malformations such as conotruncal anomalies and thymic hypoplasia (10, 11).

Left aortic arch with aberrant right subclavian artery

A left aortic arch with an aberrant right subclavian artery (Fig. 5) is the most common congenital arch anomaly with a prevalence of 0.5%–2% (11). Figure 1, a line break at 2 results in left arch with anomalous subclavian artery. This anomaly results from regression of the right arch (between the right common carotid and right subclavian arteries) including the right ductus arteriosus. The distal right dorsal aorta (rather than the right fourth arch) becomes the proximal right subclavian artery, forming the retroesophageal portion in which the right subclavian artery

originates from the descending aorta, distal to the left subclavian. As it courses toward the right arm, the aberrant vessel travels behind the trachea and esophagus. This can be seen by echocardiography and often is confirmed by magnetic resonance imaging (Fig.6A and 6B). The prevalence of an aberrant right subclavian increased to 26–34% in individuals with Down syndrome and other chromosomal defects (12).

These patients are usually asymptomatic but rarely may present with dysphagia. Sometimes, they are incidentally detected on endoscopy or barium swallow as posterior indentation on the esophagus. Usually no surgical intervention is required.

Imaging begins with determining arch sidedness from the suprasternal notch with the indicator at 3 o'clock. If bifurcation of the first vessel cannot be demonstrated, an aberrant origin of the subclavian is suspected. The aberrant vessel can often be visualized coursing parallel and postero-inferior to the first arch branch (see table 1). Additionally, Rotating the transducer to the sagittal plane with the indicator at ~1 o'clock and sweeping to the patient's right, can display the aberrant vessel. The aberrant right subclavian arises from the descending aorta crossing the midline, and courses from caudal left to cranial right. Color imaging can be useful to visualize the pulsatile flow in the aberrant vessel. The vessel should be confirmed with color Doppler interrogation.

Right aortic arch with mirror image branching

A right aortic arch with mirror image branching is the result of regression of the left fourth branchial arch between the left ductus and dorsal aorta. Figure 1 a line break at 4 results in right arch mirror image branching. The branching pattern is the mirror opposite of the left aortic arch with normal branching pattern (Fig. 7A). This anomaly typically is not associated with the formation of a complete vascular ring as the ductus typically arises from the innominate artery in

the setting of a right aortic arch, thus not forming a ring. If, however, a left sided ductus arises from the descending thoracic aorta and courses posterior to the esophagus to join the left pulmonary artery, then this results in a rare configuration consistent with a vascular ring (Fig. 7B). Right aortic arch with mirror image branching can be seen in association with other forms of congenital heart disease such as: truncus arteriosus, tricuspid atresia, and tetralogy of Fallot with or without pulmonary atresia (13). There is a strong prevalence of chromosomal anomalies such as 22q11.2 deletion syndromes with a right aortic arch (14). Patient's with isolated cases of a right aortic arch with mirror image branching are asymptomatic and usually do not require any treatment (6). They may be incidentally detected on chest x-ray with an aortic knob on the right of the trachea, pulsatile compression in the right side of the trachea on bronchoscopy and /or posterior indentation on the right side of the esophagus by endoscopy or barium swallow.

Imaging begins with suprasternal short axis views. Start inferiorly to show the ascending aorta, superior vena cava, right pulmonary artery, and left atrium. From this position sweep and angle superiorly towards the head while following the course of the first branch. The first branch, the brachiocephalic, will course leftward and will branch into the left common carotid and left subclavian arteries, unless an aberrant vessel is present. The second branch is the right common carotid artery and finally the right subclavian artery (Fig. 7C).

Vascular rings and aortic arch anomalies that lead to clinical symptoms

Vascular rings represent 1% to 3% of congenital cardiac anomalies (15). A vascular ring is defined as an abnormality of the aortic arch, its branches, or remnants that results in encircling of the trachea and esophagus with variable degrees of compression to the trachea and/or esophagus. Vascular rings are classified as complete or incomplete. Complete vascular ring refers to conditions in which the abnormal vascular structures or their remnants form a

complete circle around the trachea and esophagus. Double aortic arch and right aortic arch with left ligamentum arteriosum are examples of complete vascular ring. Incomplete vascular ring refers to vascular anomalies that do not form a complete circle around the trachea and esophagus but do compress the trachea or esophagus. These include anomalous innominate artery, left aortic arch with aberrant right subclavian artery, and anomalous left pulmonary artery ("vascular sling" or "pulmonary sling") (3,8,16). Tracheal compression from both the right and left side lead to respiratory symptoms such as: respiratory distress, stridor, seal-bark cough, apnea, cyanosis, or recurrent respiratory infections, typically in the first year of life. Respiratory symptoms have been reported in 70–97% of patients with vascular rings at presentation (17). Feeding difficulties such as dysphagia, slow feeding, and hyperextension of head while eating may present later in life since liquid diets are tolerated earlier and symptoms typically manifest when solid foods are initiated (18). Vascular rings that are "less tight" may not manifest until childhood (or rarely adulthood) when patients are noted to have respiratory symptoms with activity. Symptoms such as dyspnea, wheezing, and cough are often misdiagnosed as asthma, particularly if they occur in older children. Vascular rings can also present as failure to wean from mechanical ventilation following unrelated surgery (19).

Double aortic arch

A double aortic arch is the result of both embryonic right and left arches persisting, passing on both sides of the trachea and esophagus and joining posteriorly to form the descending aorta (Fig.8). This results in a complete encircling of the trachea and esophagus (10). A double aortic arch is the most common symptomatic vascular ring, as the trachea and esophagus are completely encircled and may be compressed by the two arches (16). Clinical manifestations of a double aortic arch in infants and children may be present from birth and

include wheezing and stridor. Double aortic arch is rarely associated with congenital heart disease but when present, tetralogy of Fallot is the most common disorder, followed by transposition of the great arteries (20). In the setting of a double aortic arch, the right arch is usually larger and higher than the left arch. The descending aorta is typically opposite the dominant arch. The most common configuration being a larger right arch, left-sided descending thoracic aorta, and left-sided ligamentum arteriosum (11). Occasionally a smaller arch can be partially atretic and may not show color flow Doppler by echocardiogram. Determining the dominant aortic arch is important for surgical planning. Division of the non-dominant arch is performed through a lateral thoracotomy. Typically the thoracotomy is performed on the side of the non-dominant arch. Advanced cardiac imaging may be helpful to determine the smaller arch and the relationship of the airway to the vascular anatomy (8, 21).

In patients with a double aortic arch, both left and right arches must be evaluated for obstruction. Imaging of a double aortic arch is best visualized in the suprasternal short axis plane. With the transducer indicator at ~3 o'clock, sweeping superiorly will demonstrate the ascending aorta giving off symmetrical head and neck vessels, two on each side (Fig.9A). The double aortic arch can be confirmed in the orthogonal long axis plane by sweeping back and forth between the two arches separated by the trachea. With the transducer indicator at 1 o'clock, the transducer is angled right and left to display each individual arch discretely separate from the other (Fig 9B). Magnetic resonance imaging nicely shows the bilateral arches encircling the space occupied by the airway and esophagus (Fig 9C).

Right aortic arch with aberrant left subclavian artery with diverticulum

A right aortic arch with an aberrant left subclavian artery results from regression of the left 4th aortic arch segment between the left common carotid and subclavian segments. Referring

to Figure 1 with a line break at 3 leads to right arch with aberrant subclavian artery. As a result, the left subclavian originates as the last branch from the aortic arch, at a posterior location, coursing behind the esophagus to the left arm (Fig.10). A left ductal ligament originates from a bulbous dilation at the base of the left subclavian artery (termed diverticulum of Kommerell) and attaches to the proximal left pulmonary artery. This effectively pulls the aorta and diverticulum anteriorly, forming a vascular ring and creating varying degrees of compressing the esophagus and trachea (10). Those with significant narrowing of the trachea and esophagus will present in infancy, while some with loose rings will be asymptomatic even as adults. For those with clinical symptoms, surgical ligation, and division of the ductal ligament is performed via a lateral thoracotomy.

Begin imaging in the suprasternal short axis views. Start with an inferior to superior sweep towards the head. The first branch off the aortic arch will be the left common carotid artery which courses leftward and superiorly. Follow the common carotid artery leftward and superiorly. The aberrant left subclavian artery will become evident, running parallel to the common carotid artery, with pulsatile, red flow. The second and third branches are the right common carotid and right subclavian artery. Imaging from the sagittal long axis focus on the proximal descending aorta, the aberrant left subclavian crosses the midline posterior to the esophagus to achieve its leftward course (Fig 11A). Doppler interrogation of the vessel should be performed to confirm the aberrant vessel. Figure 11B demonstrates a computed tomography image showing the right arch with a prominent posterior diverticulum of Kommerell which results in posterior compression by barium swallow (Figure 11C).

Circumflex aorta

A circumflex aorta is a rare aortic arch anomaly caused by retroesophageal crossing of the aorta to the opposite side. This can present as a right or left circumflex arch, with multiple variations. Embryologically, a right circumflex aorta develops when there is regression of the left fourth primitive arch and persistence of the left ductus arteriosus and left dorsal aorta. Left circumflex aorta develops from regression of the right fourth branchial arch with persistence of the right ductus arteriosus and right dorsal aorta. A right circumflex aorta is more common than a left circumflex aorta. The left aortic arch with right thoracic descending aorta (circumflex aortic arch) crosses immediately midline in the superior thorax, coursing posterior to the trachea and esophagus, then descends to the right of midline (Fig.12). The branching pattern may have a left innominate artery or an aberrant left subclavian artery. In either case, there is a vascular ring when there is a left-sided ligamentum. There is 50% to 60% prevalence for additional cardiac anomalies (21). As with other vascular rings, patients will present with wheezing, stridor, dyspnea, frequent respiratory infections or dysphagia. A chest x-ray will demonstrate a widened mediastinum and a barium swallow or endoscopy will demonstrate a posterior indentation in the esophagus where the aortic arch crosses over the midline (5, 8).

Echocardiographic imaging of a circumflex aortic arch is challenging (Fig 13A). Branching can be difficult to demonstrate. Begin with suprasternal short axis imaging in the suprasternal notch with the indicator at ~3 o'clock. Sweep superiorly and follow the course of the first arch vessel to evaluate for branching. The first branch will typically be the innominate artery coursing leftward, followed by the right common carotid, and right subclavian artery. Often, the long axis view will be unable to show the classic candy cane view as the course of the circumflex arch is too tortuous to display in one view. Challenging suprasternal views coupled with a leftward descending aorta may be the only clues to suggest a circumflex aortic arch.

Cross-sectional imaging is often necessary to confirm this diagnosis. Figure 13B shows a magnetic resonance angiography of a right circumflex arch that descends leftward resulting in a vascular ring.

Innominate artery compression syndrome

This syndrome occurs when the innominate artery originates more distal along the course of the transverse arch, resulting in takeoff to the left of the trachea. The innominate artery then has to course rightward and anteriorly over the trachea as it then divides into the right subclavian artery and right common carotid artery. Anterior tracheal compression with associated tracheomalacia results when the innominate artery courses rightward (22). Infants with this anomaly, typically present with recurrent stridor and respiratory issues. Chest x-ray is non-determinate. In order to diagnose this, patient present with respiratory symptoms and then have anterior tracheal narrowing seen on bronchoscopy or dynamic computed tomography. Surgical repair has been used in severe cases with an aortopexy (moving aorta anteriorly and attaching to sternum) or innominate artery re-implantation; however, many cases will improve as the infant grows and airway increases in size (8, 16).

When imaging by echocardiography, innominate artery compression will present as a normal left branching arch. The only exception being the bifurcation will be more distal from the aortic arch. The role of echocardiography in diagnosing this syndrome is limited and typically is necessary for excluding associated congenital heart disease. It is rarely associated with congenital heart disease. In Figure 14, compression of the trachea can be seen on the computed tomography image which demonstrates the innominate artery coursing anterior and causing mass effect.

Left pulmonary artery sling

Pulmonary artery sling occurs when there is failure of development of the left sixth aortic arch during gestation. A left pulmonary artery sling is also known as anomalous left pulmonary from the right pulmonary artery. The left pulmonary arises distally from the proximal right pulmonary and courses over the right mainstem bronchus, posterior to the trachea and anterior to the esophagus to reach the left hilum (Fig.15A, B). This is the only vascular anomaly to course between the trachea and esophagus, causing anterior esophageal compression and displacement of the trachea anteriorly. A left pulmonary artery sling is frequently associated with complete tracheal cartilaginous rings. About 10% to 20% of patients with this anomaly have associated cardiac defects, such as patent ductus arteriosus, ventricular septal defect, atrial septal defect, atrioventricular canal, single ventricle, or aortic arch (7). Advanced cardiac imaging is useful to delineate the relationship of the intrathoracic structures to the vascular anomaly. This can be repaired by dividing the left pulmonary artery and implanting it into the main pulmonary artery anterior to the trachea (5, 22).

Focused assessment of the main pulmonary artery and the branch pulmonary arteries should demonstrate the unusual course of the left pulmonary artery. Imaging is optimal in the high left parasternal window. It may be useful to lay the patient in the left lateral decubitus position, or in the supine position with the neck hyperextended. The transducer indicator should be at ~3 o'clock. From this view, the main pulmonary artery and branching is well visualized. The left pulmonary artery can be visualized originating from the proximal posterior portion of the right pulmonary artery. The left pulmonary artery courses posterior and slightly leftward to course between the trachea and esophagus to reach the left lung. The echobright cartilaginous rings of the trachea may sometimes be visible as the left pulmonary artery courses posterior to

the structure. Care should be taken not to confuse a large patent ductus arteriosus with the left pulmonary artery in newborns. A high parasternal ductal view with the indicator at ~12 o'clock can be employed to visualize the main pulmonary artery separate from the patent ductus arteriosus as it connects to the descending aorta.

Conclusion

Echocardiography plays an essential role in the noninvasive evaluation of the aortic arch. The cardiac sonographer's systematic approach to determine arch sidedness, branching pattern and the patency of the aortic arch is important to fully assess the aorta. Computed tomography angiography and magnetic resonance angiography are utilized to further define the two and three-dimensional vascular anatomy within the thorax. Knowledge of the common anatomical variants, typical clinical symptoms and echocardiographic findings is critical in order to diagnose these relatively rare forms of congenital heart disease.

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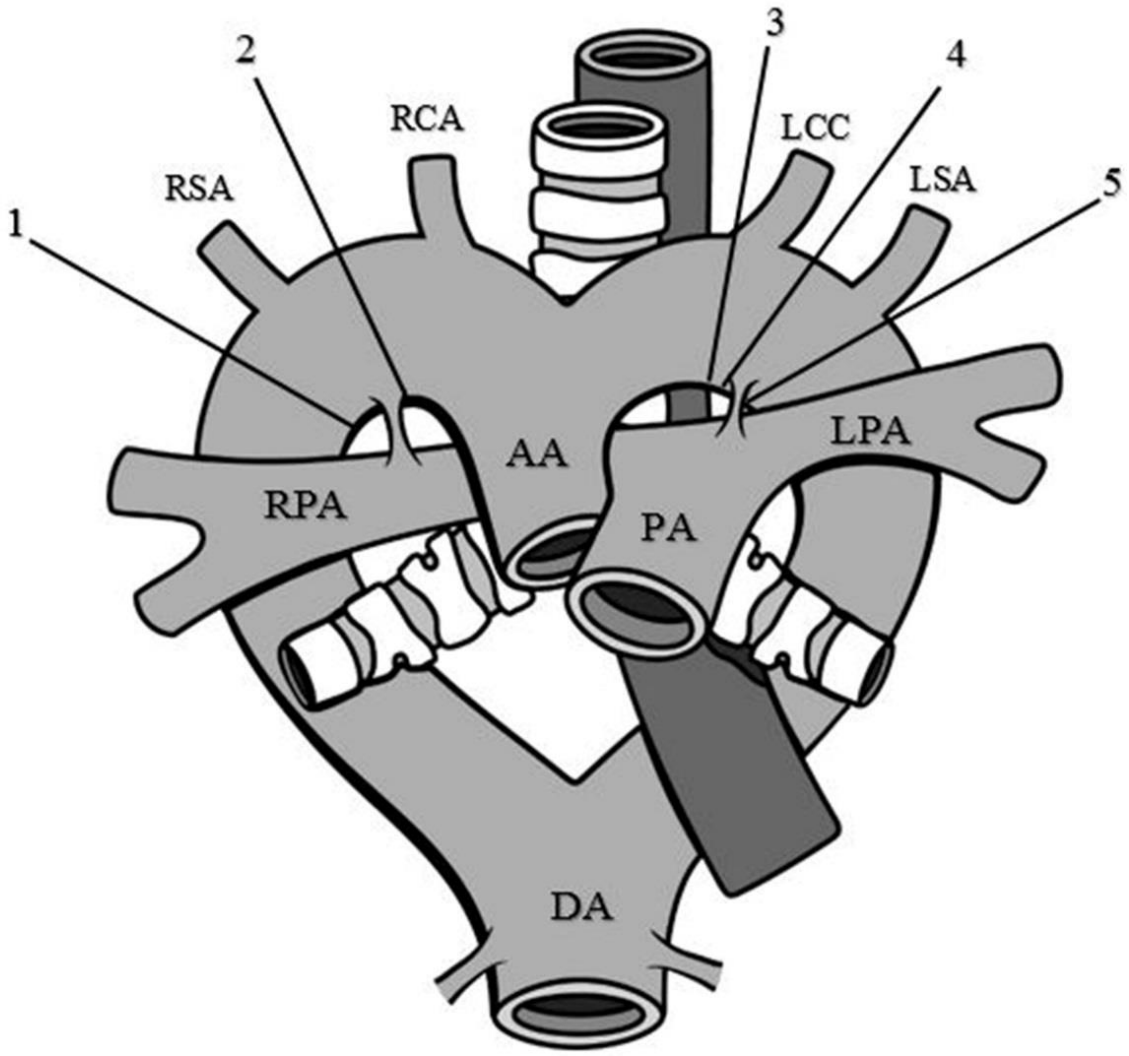
Aortic Arch Variants	First branch relative to trachea	1st Branch	Bifurcation of 1st branch	2nd Branch	3rd Branch	Other findings
LAA	Rightward	IA/ BT	Yes	LCCA	LSA	
Bovine	Rightward	IA/ LCCA	Yes	LSA		The IA and LCCA have a common origin
LAA with vertebral artery	Rightward	IA	Yes	LCCA	Vertebral artery	4th vessel LSA
LAA ARSA	Rightward	RCCA	No	LCCA	LSA	ARSA coursing parallel and postero-inferior to RCCA
RAA	Leftward	IA/ BT	Yes	LCCA	RSA	
RAA ALSA	Leftward	LCCA	No	RCCA	RSA	ALSA coursing parallel and postero-inferior to LCCA
DAA	Bilateral	N/A	N/A	N/A	N/A	4 symmetric branches.
Right circumflex	Leftward	IA/BT	Yes	RCCA	RSA	LAA with right thoracic Dao

Table 1: Aortic arch branching variants

Abbreviation key: LAA- left aortic arch, IA- innominate artery, BT- brachiocephalic trunk, LCCA- left common artery, LSA- left subclavian artery, ARSA- aberrant right subclavian artery, RAA- right aortic arch, ALSA- aberrant left subclavian artery, DAA- double aortic arch, Dao- descending aorta

Figure 1



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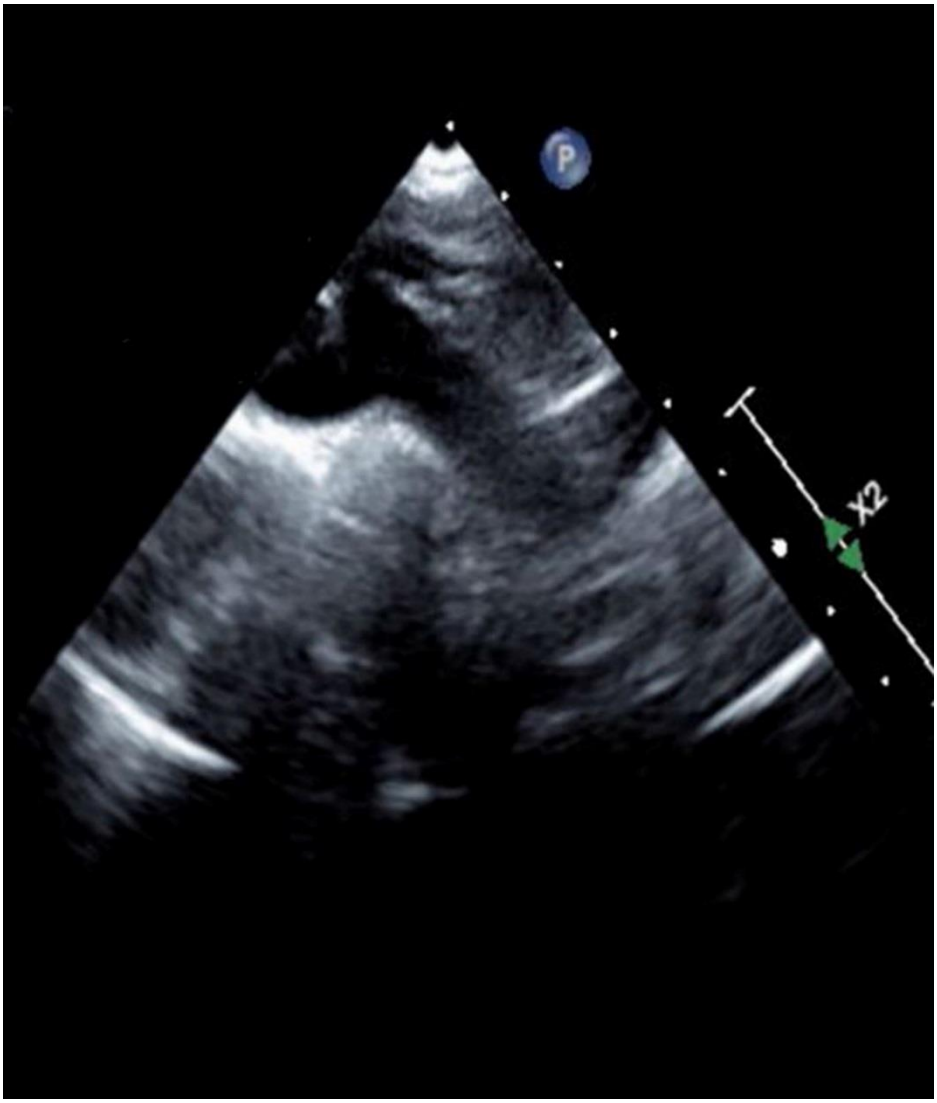

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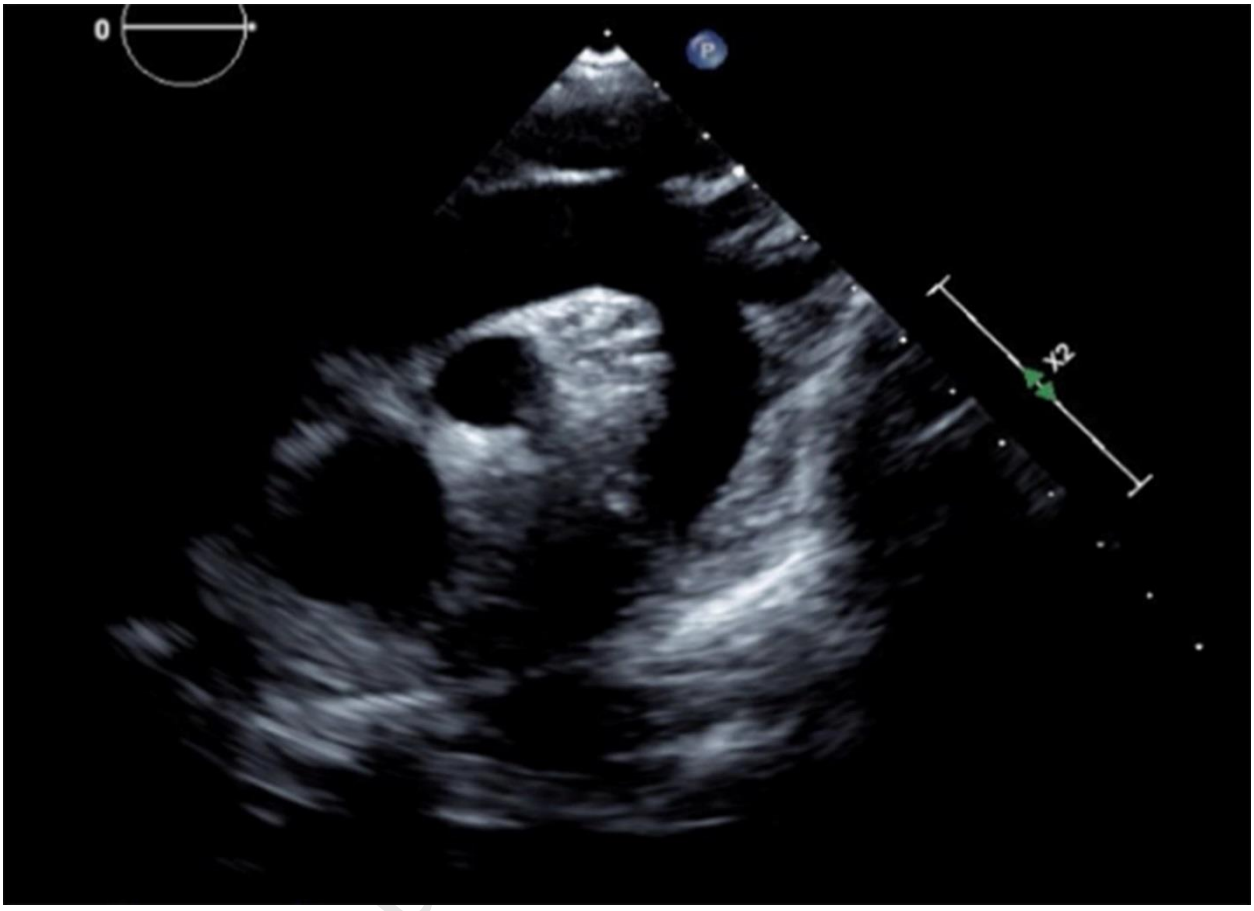

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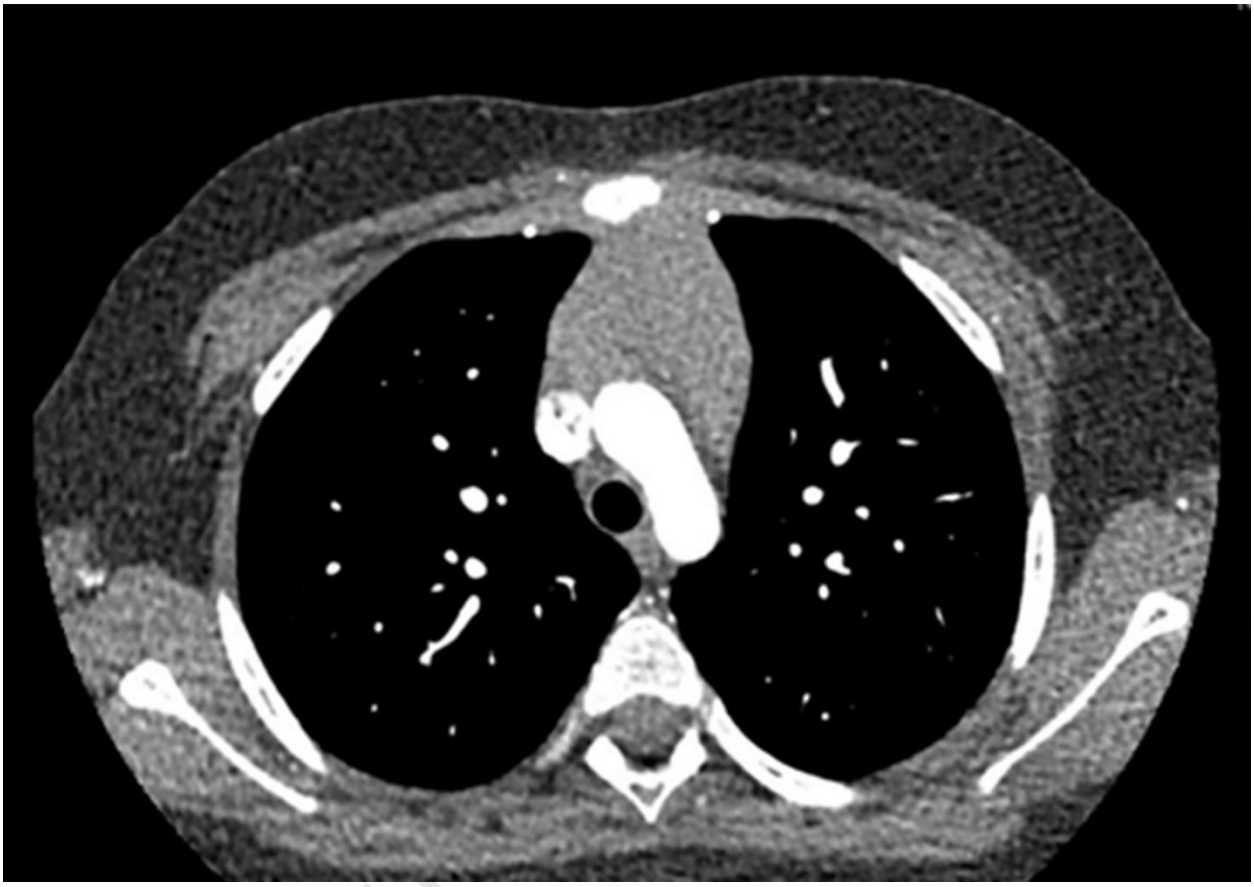

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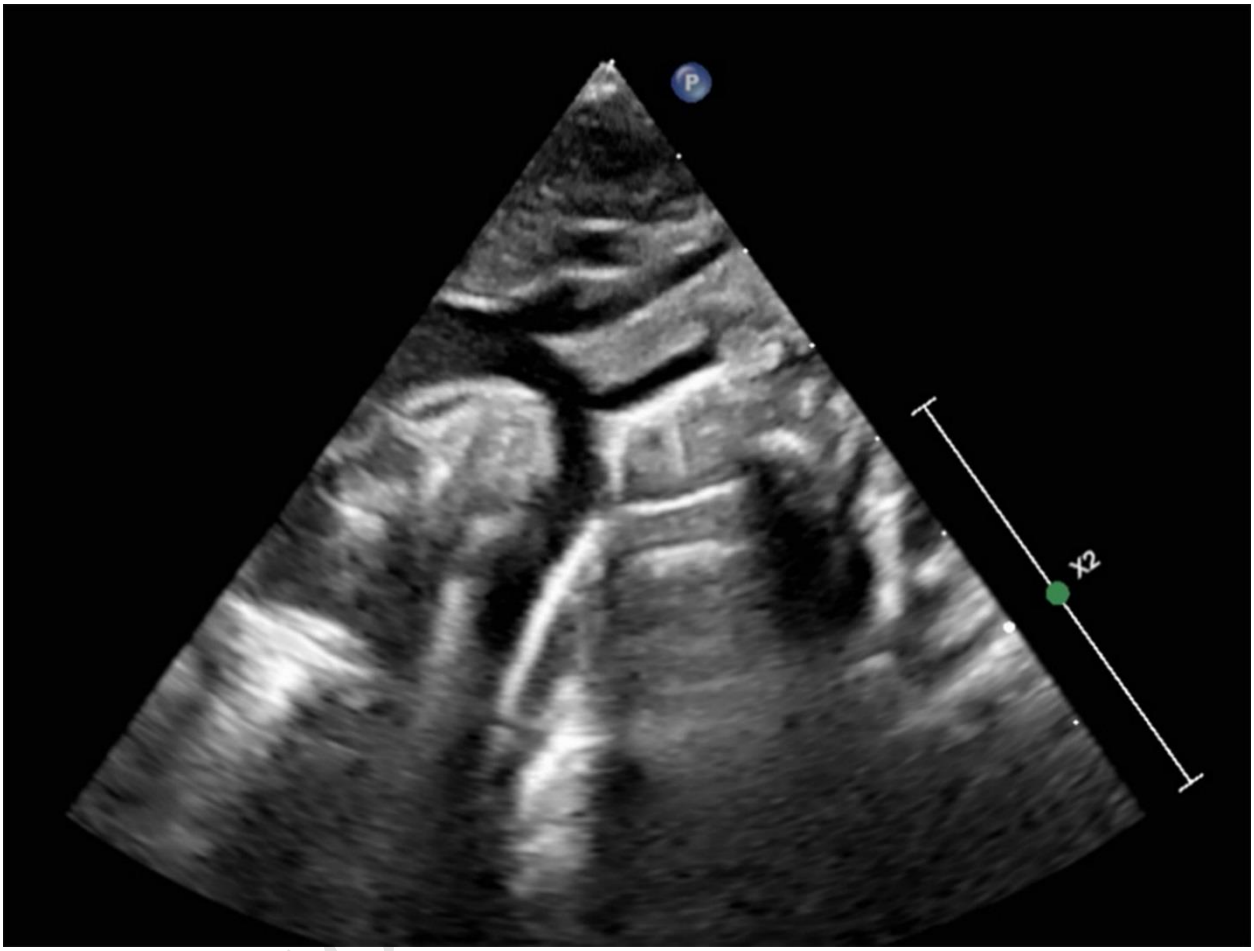

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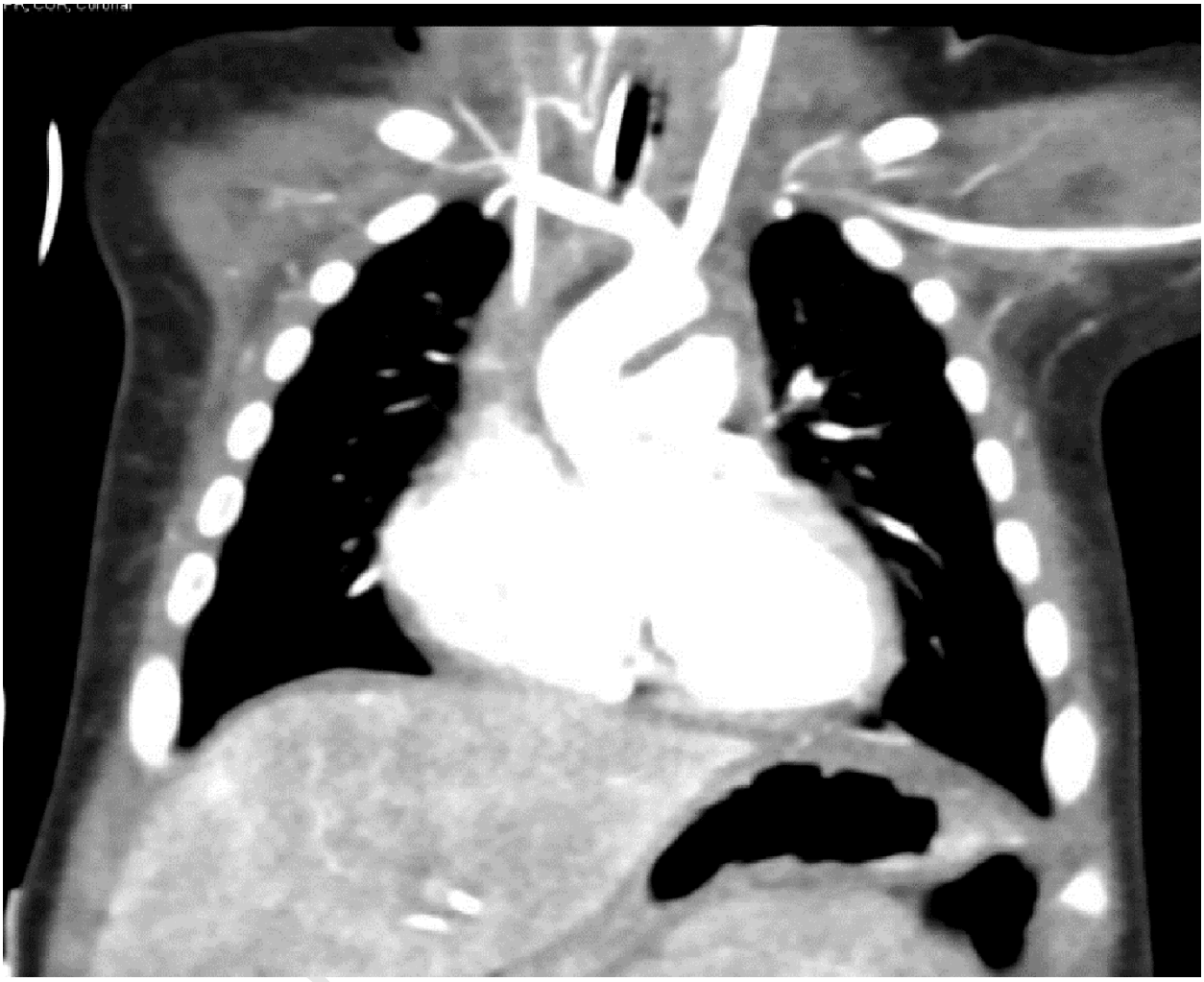

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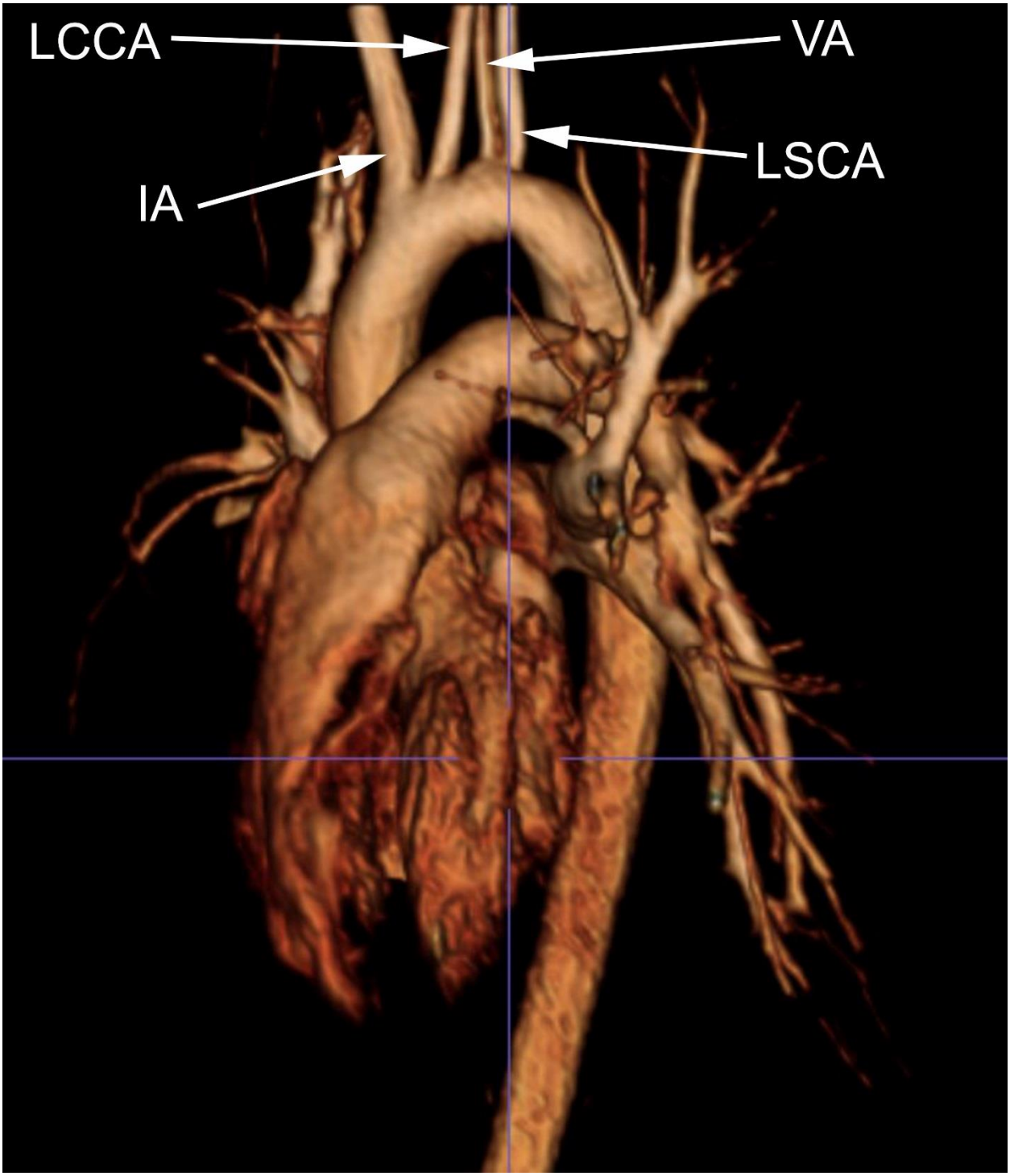

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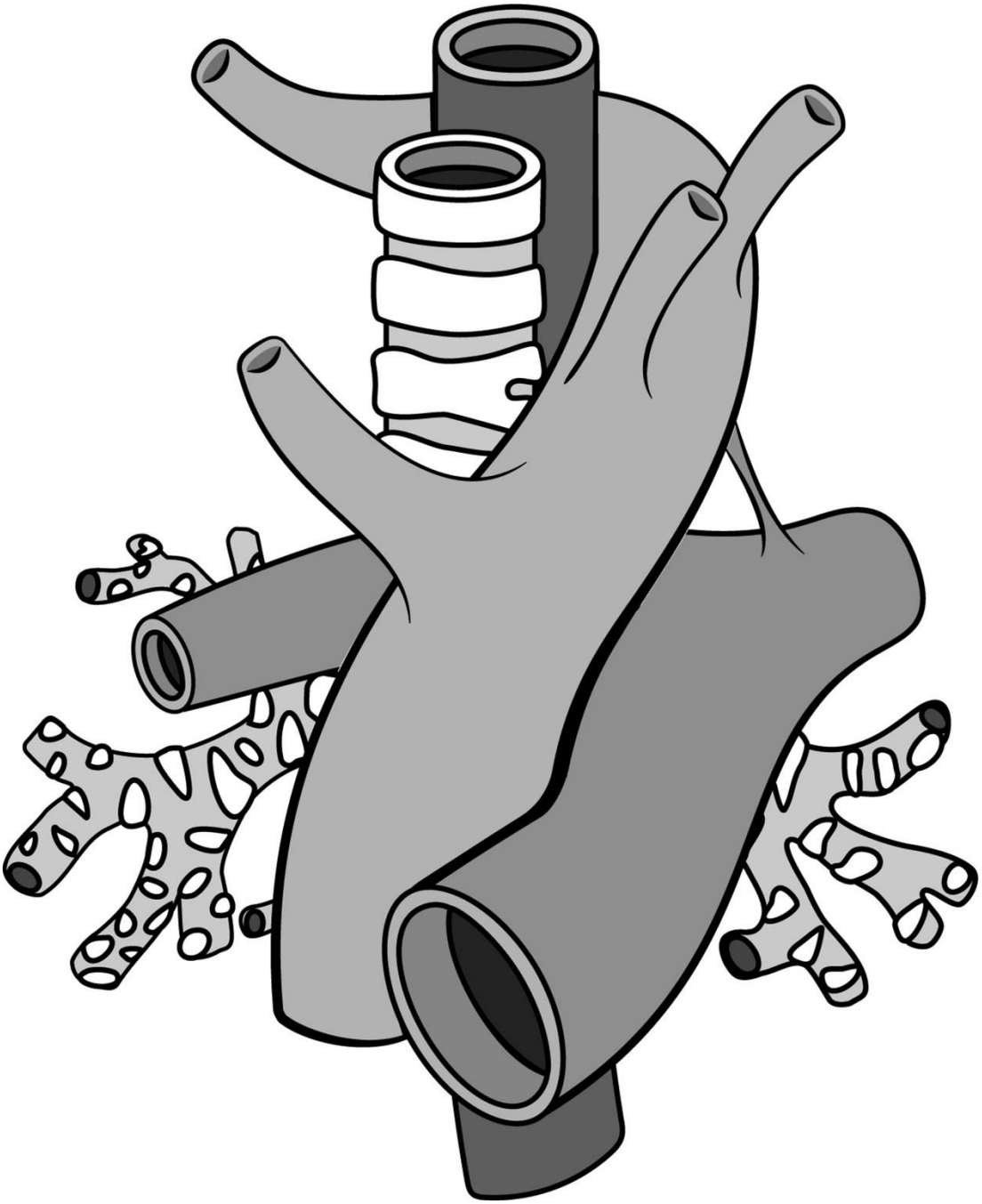

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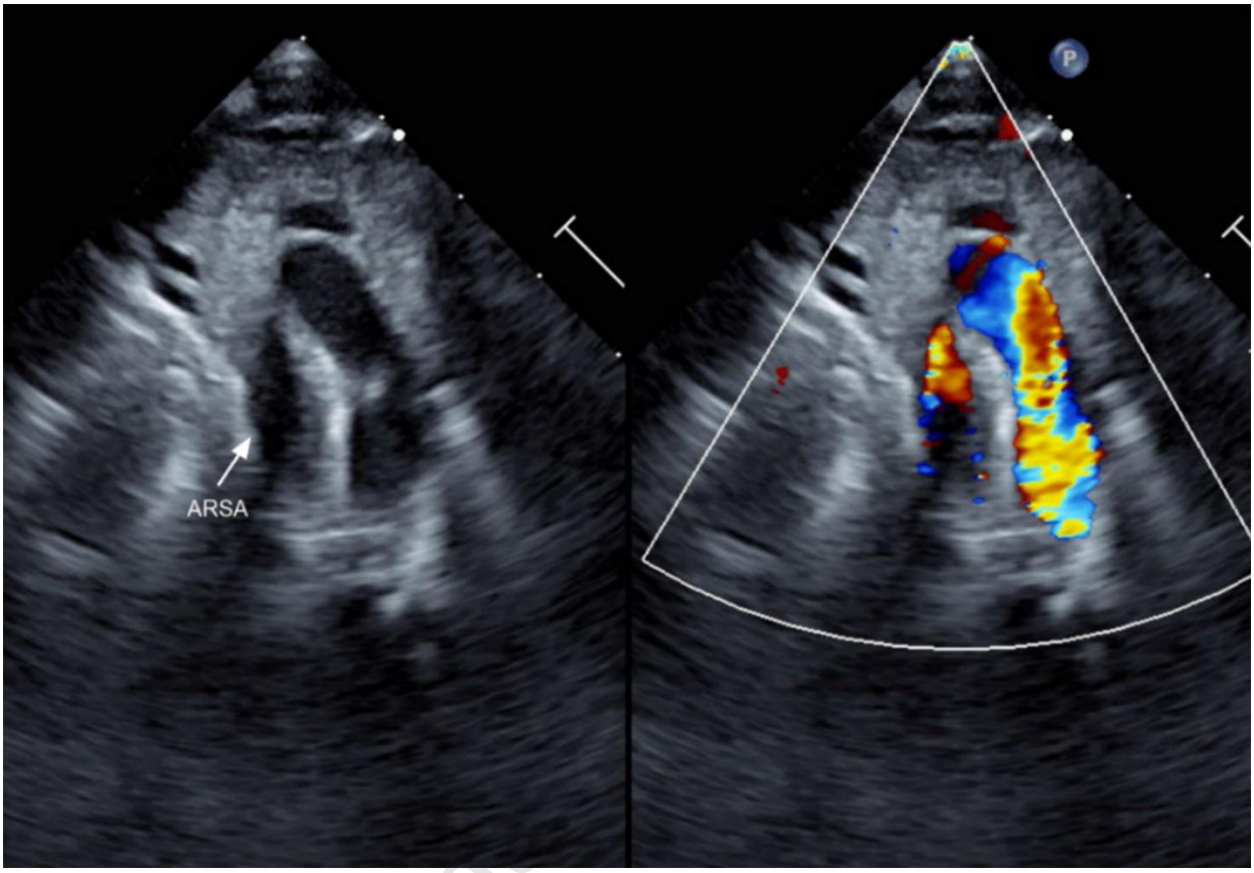

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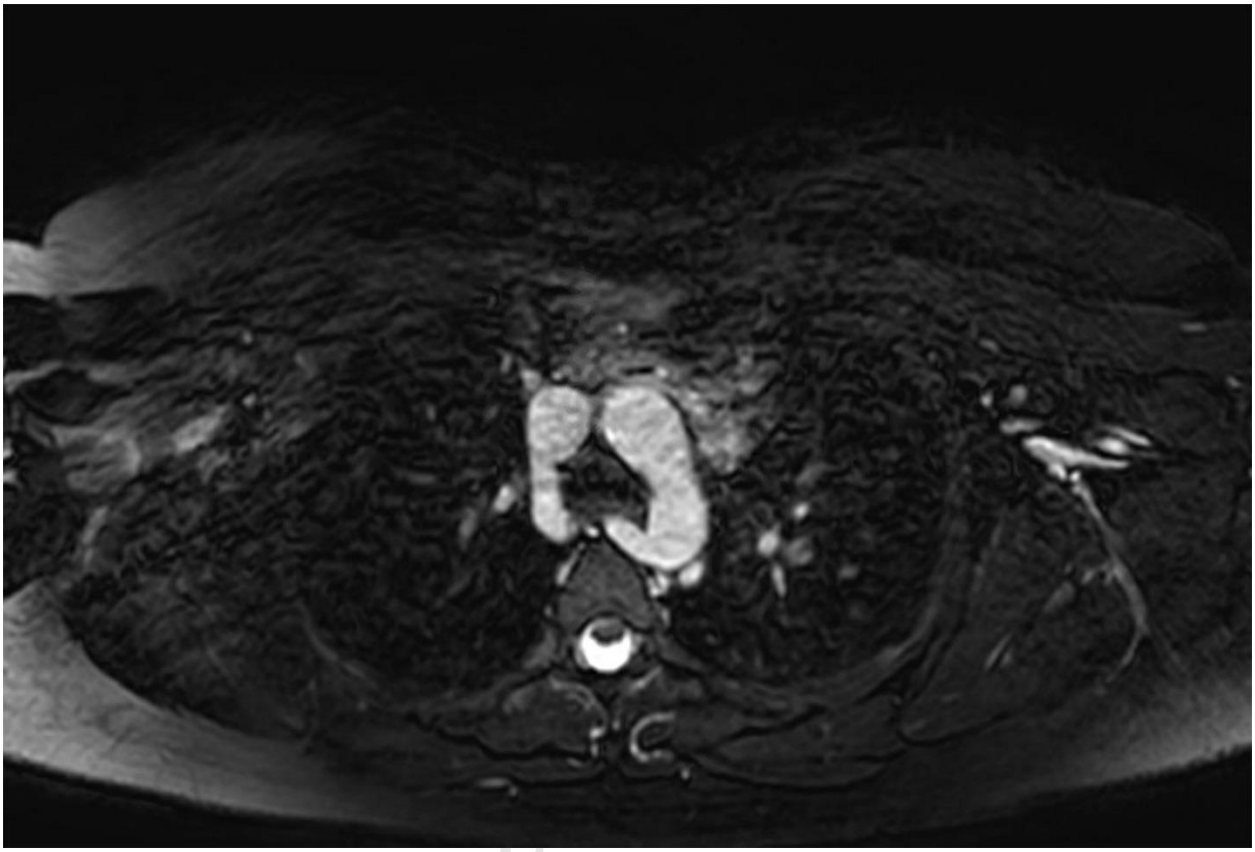

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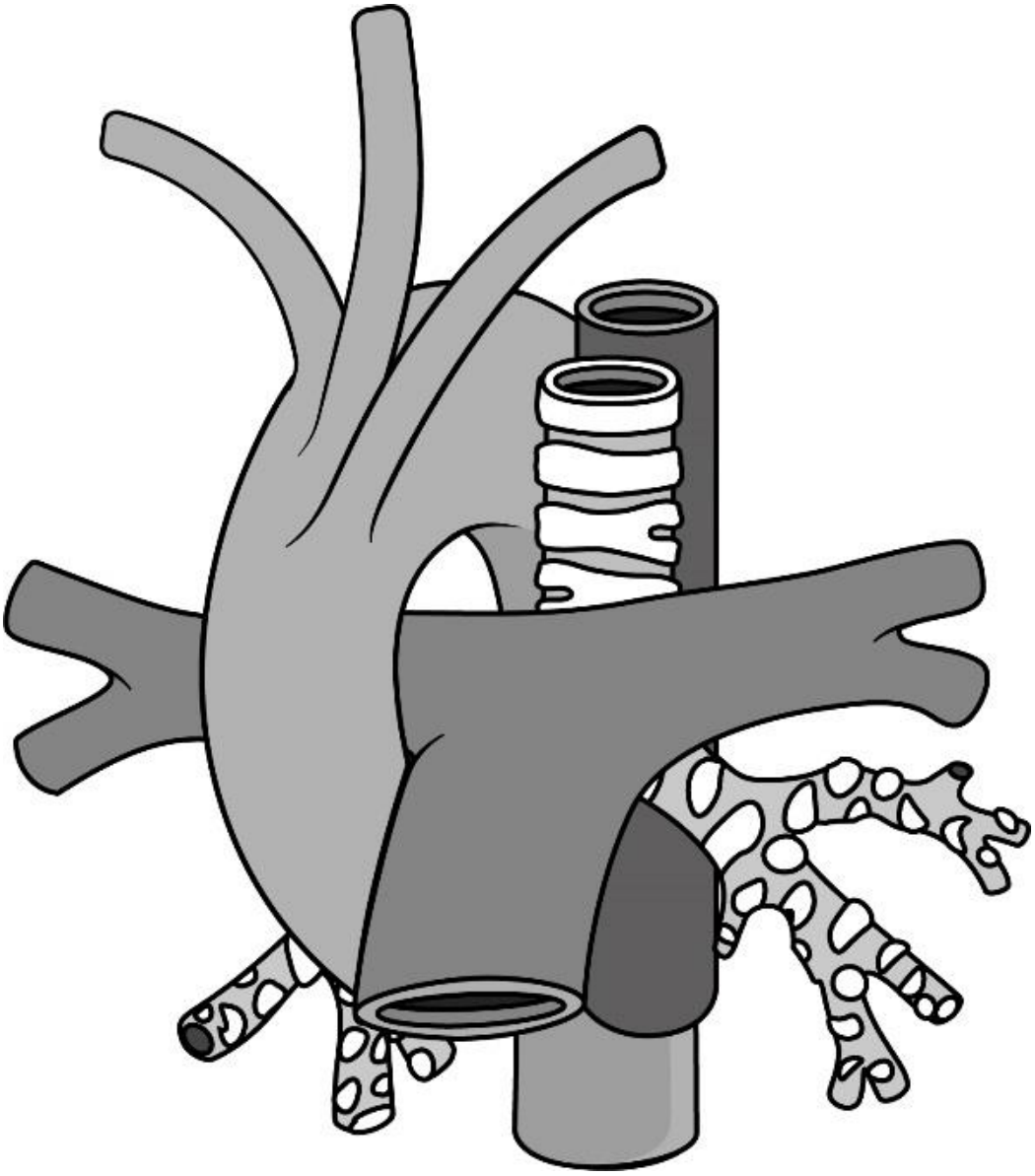

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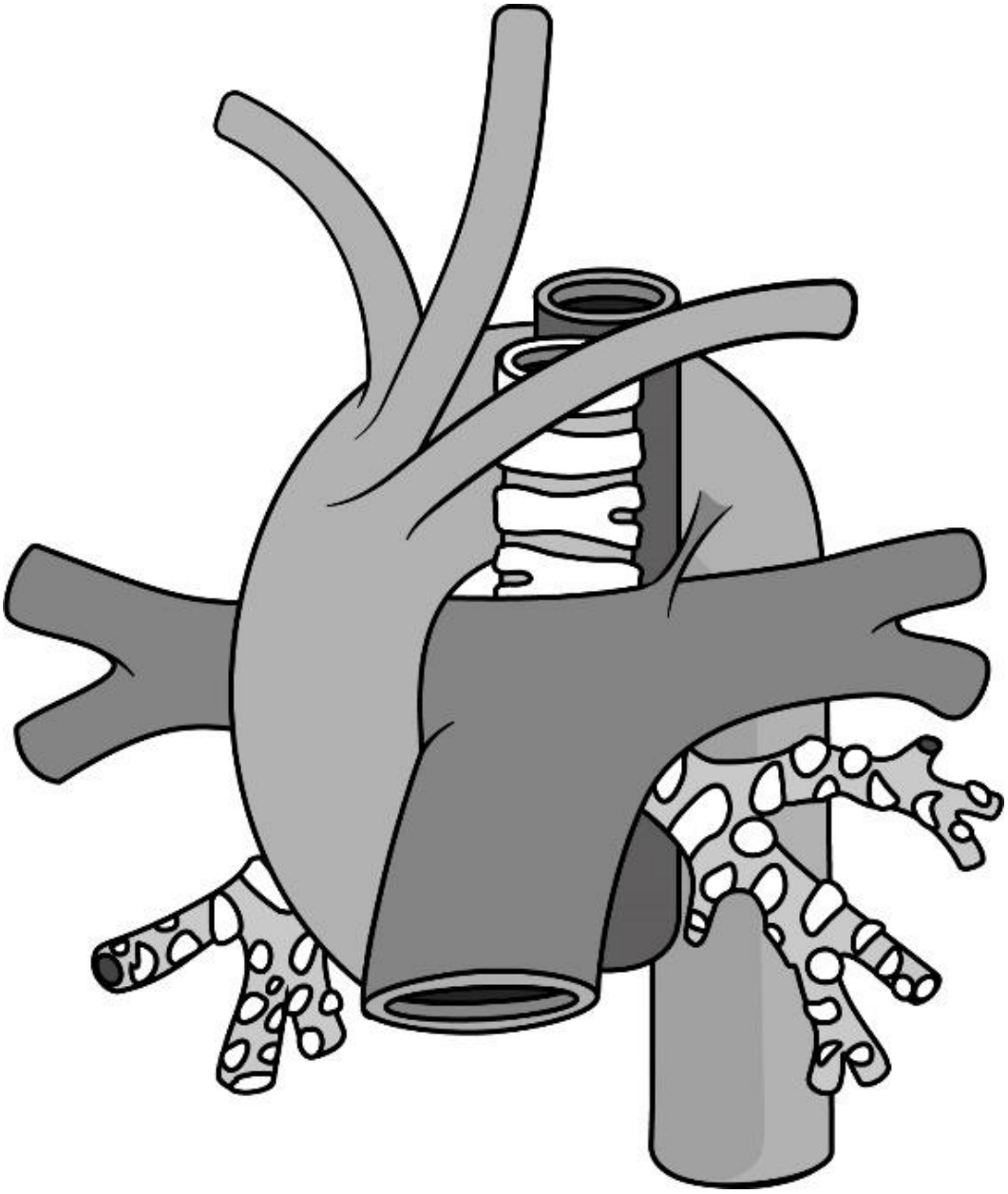

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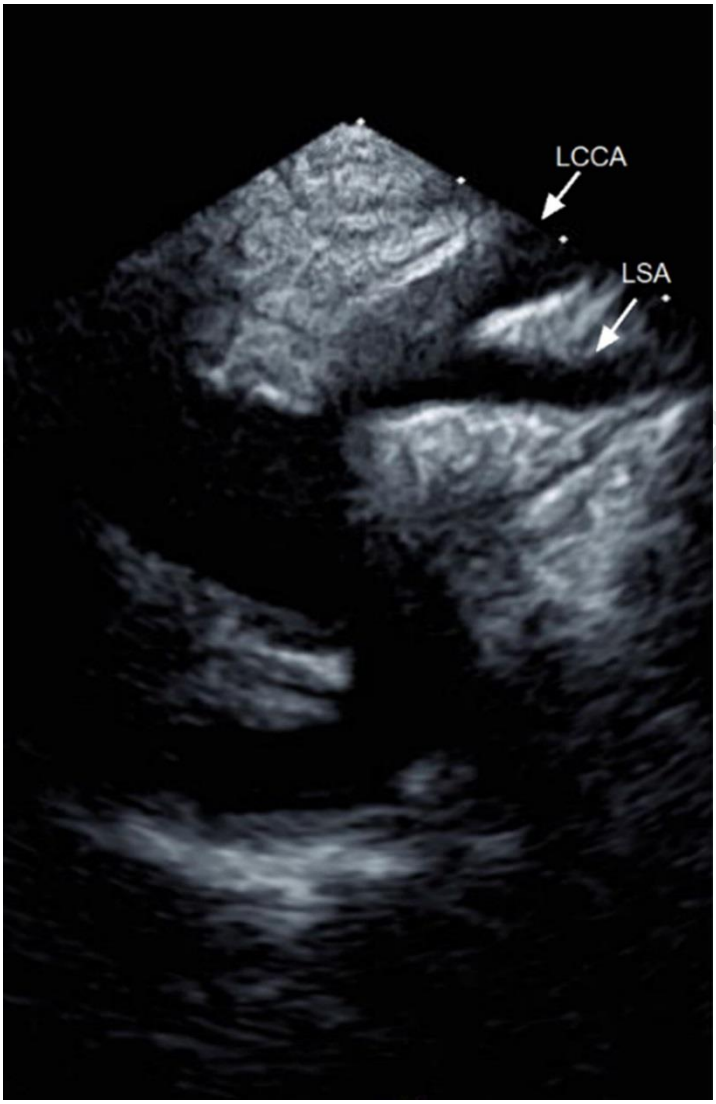

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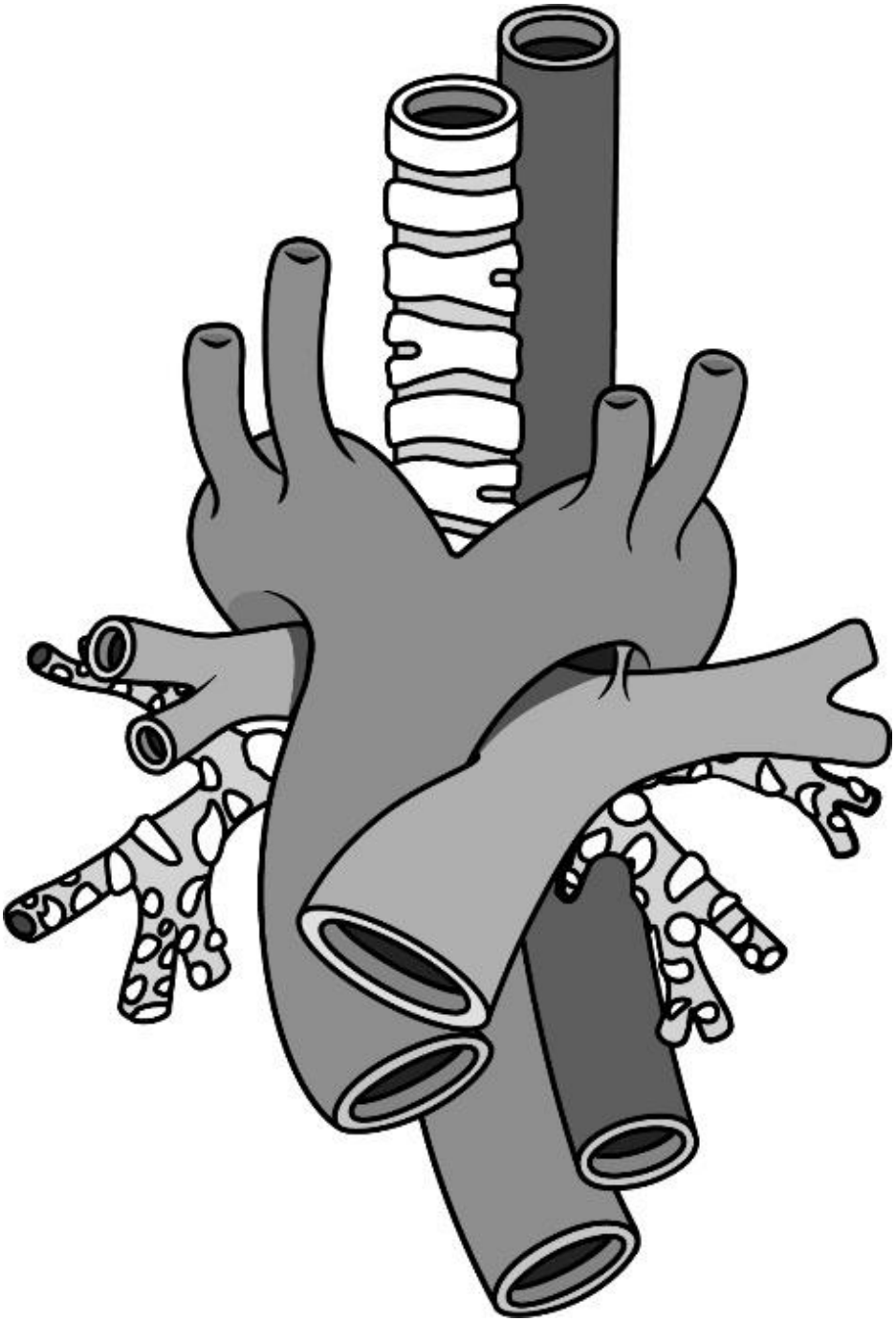

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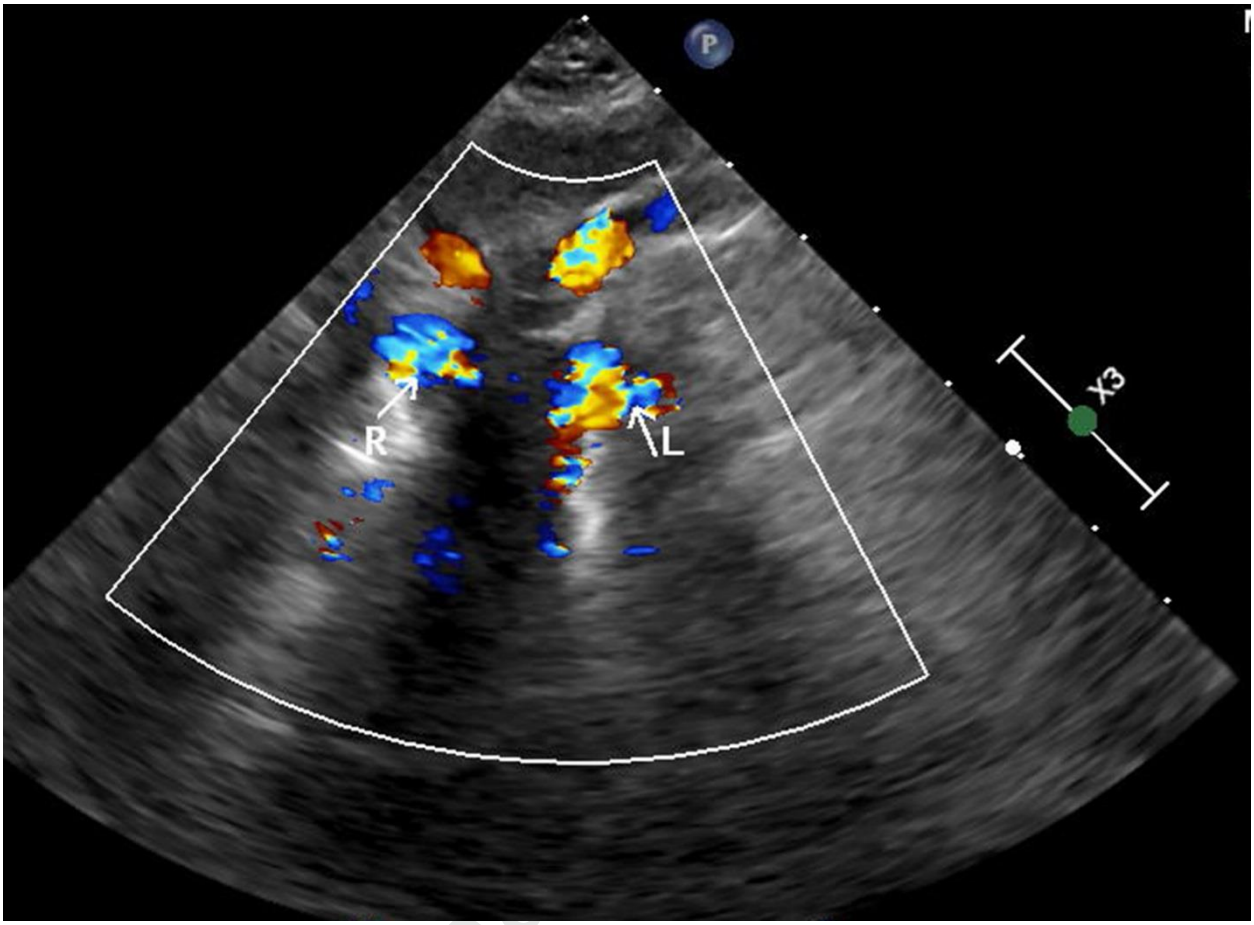

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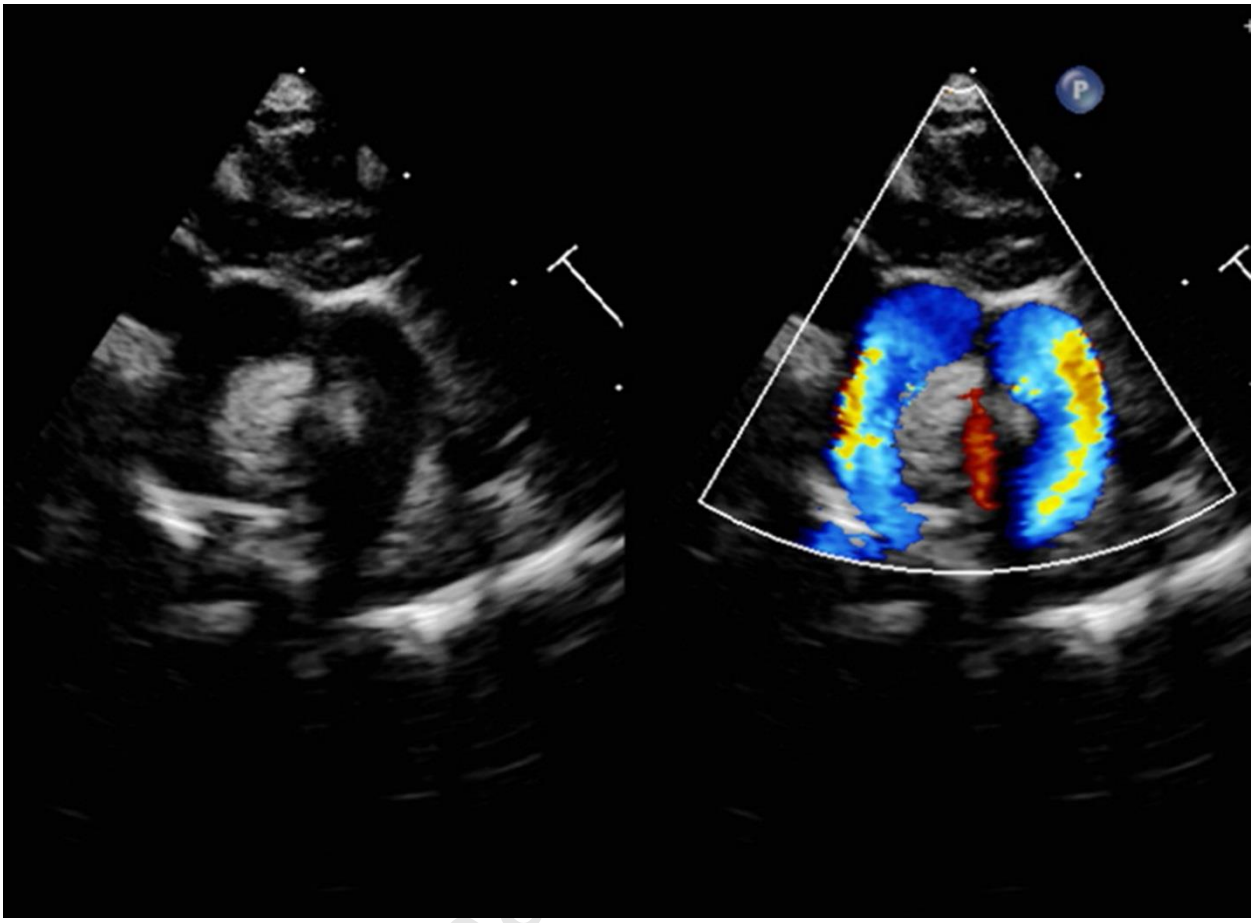

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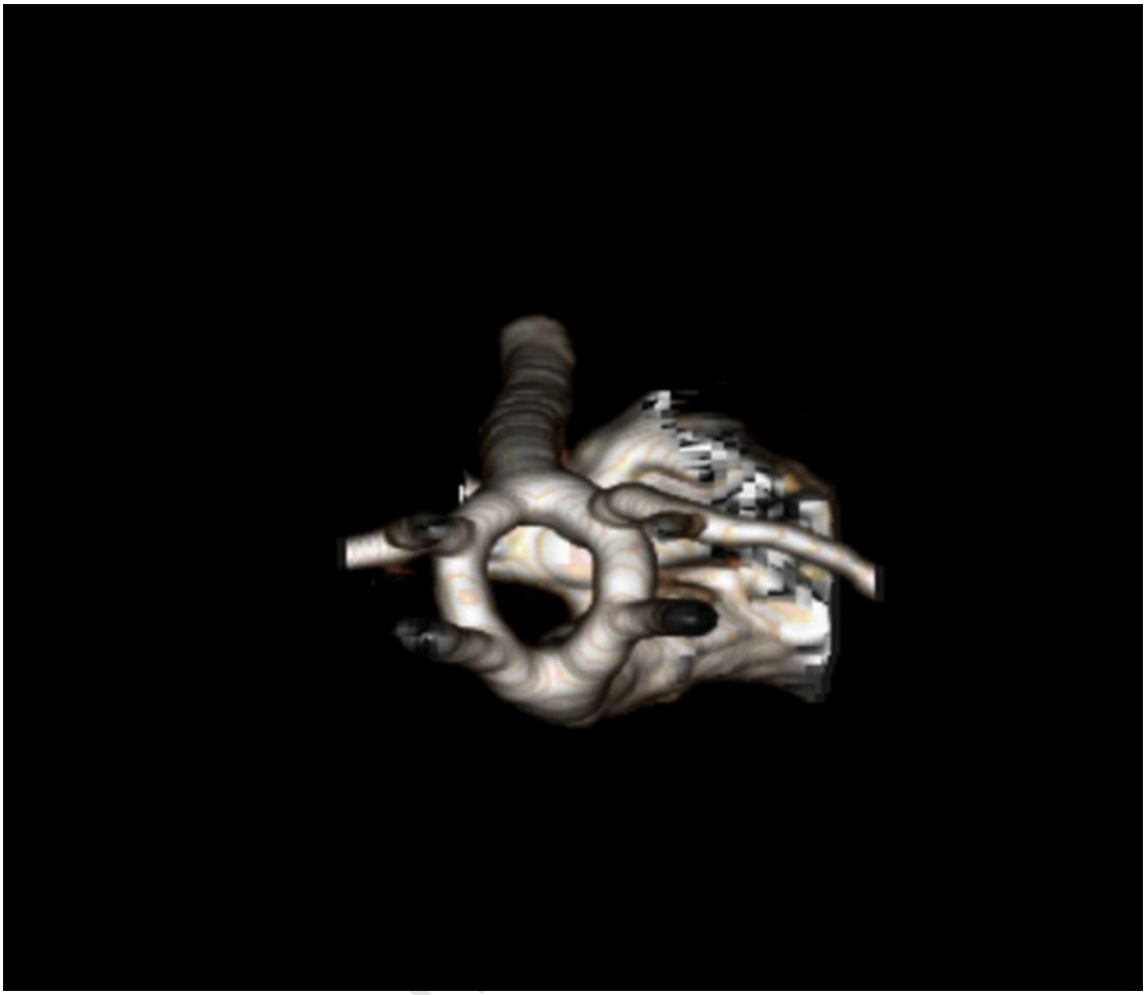

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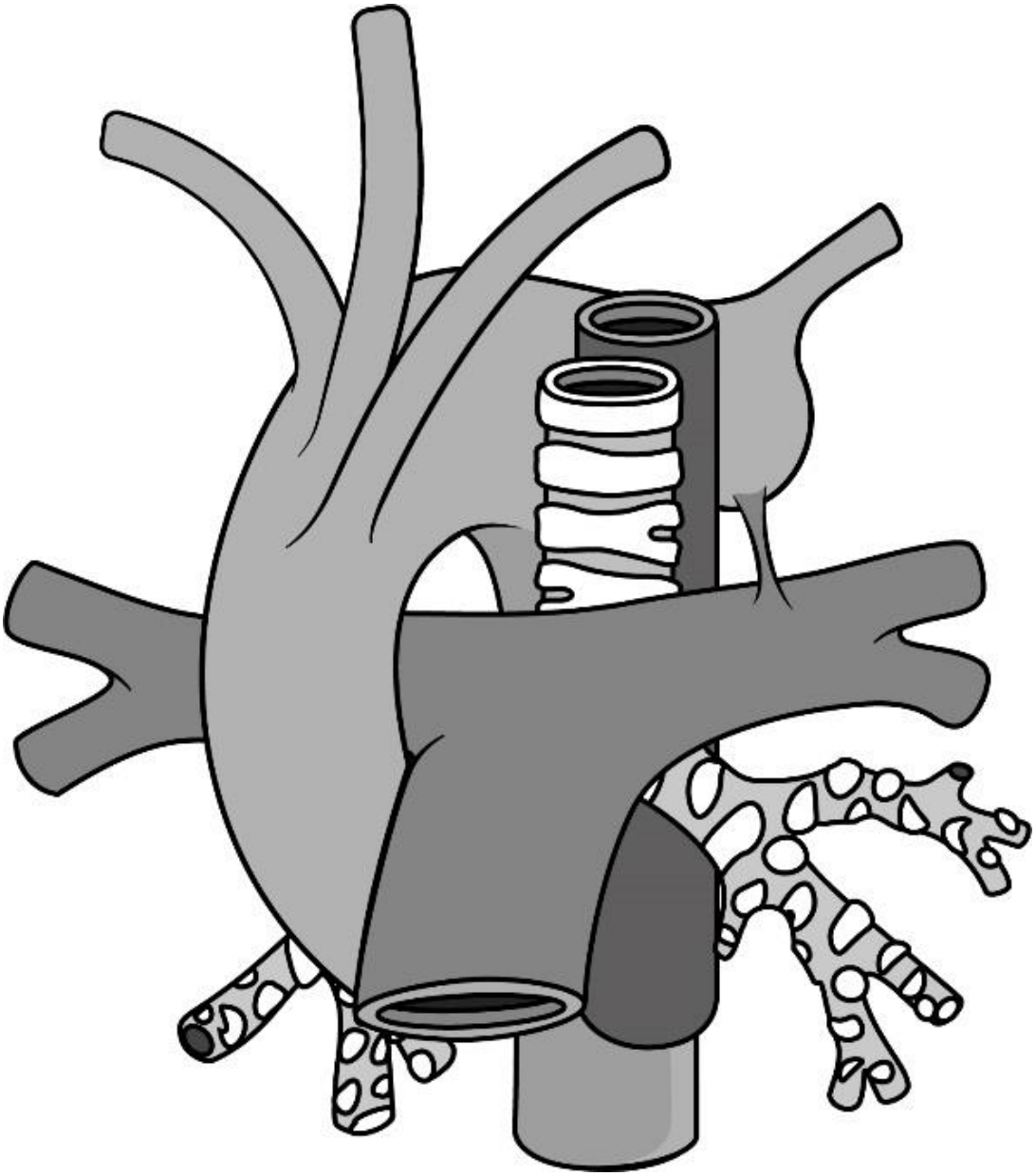

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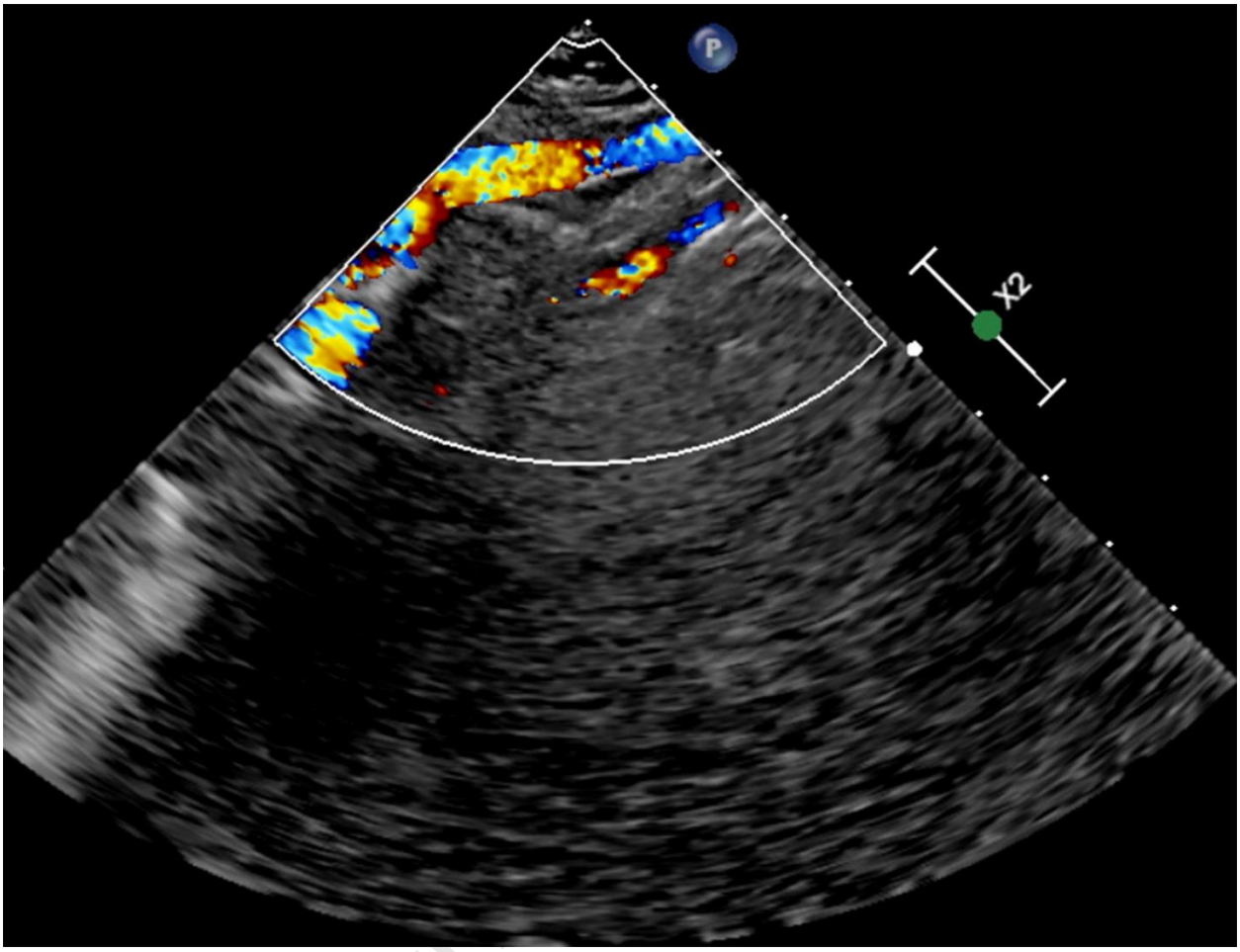

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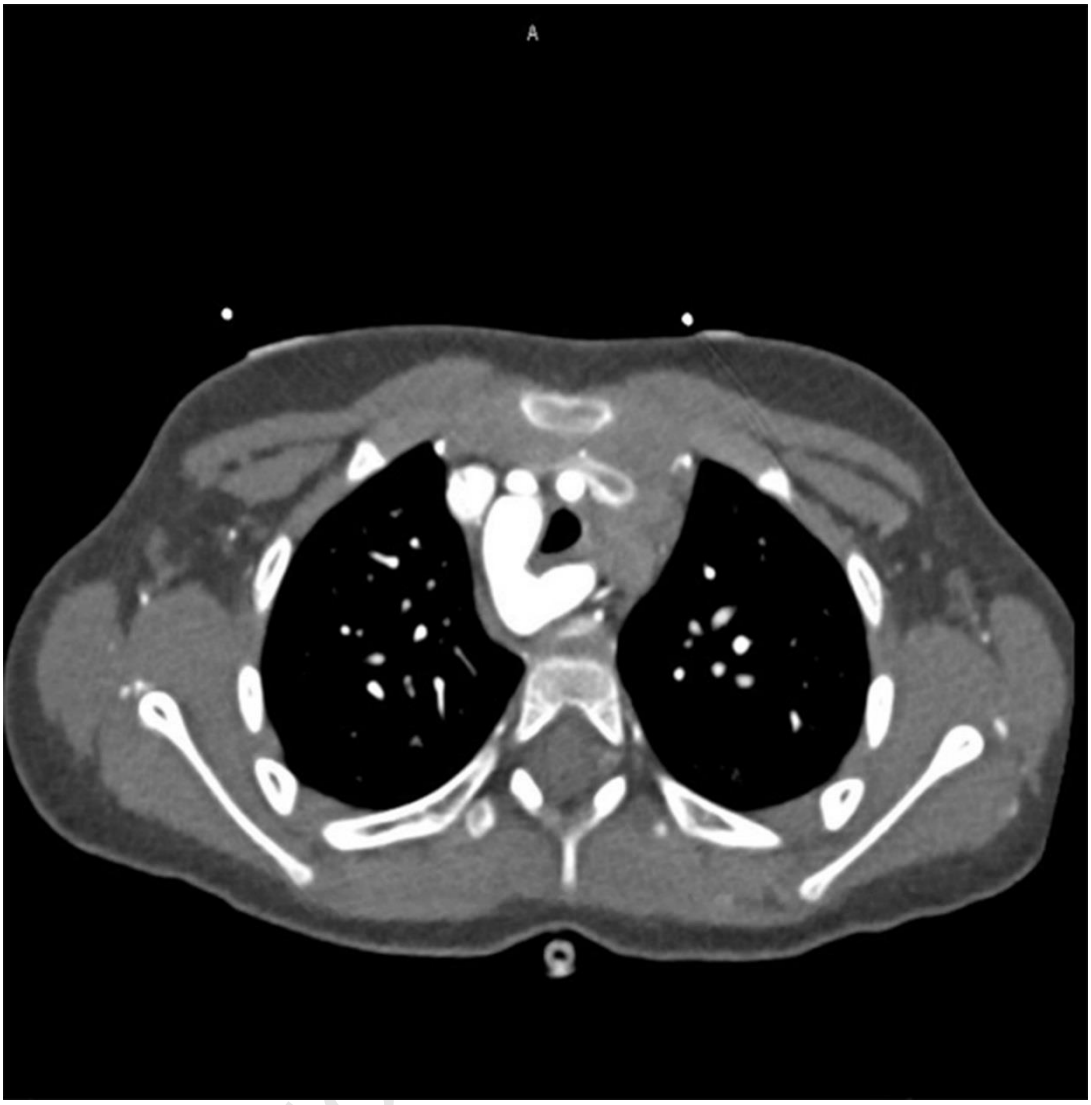

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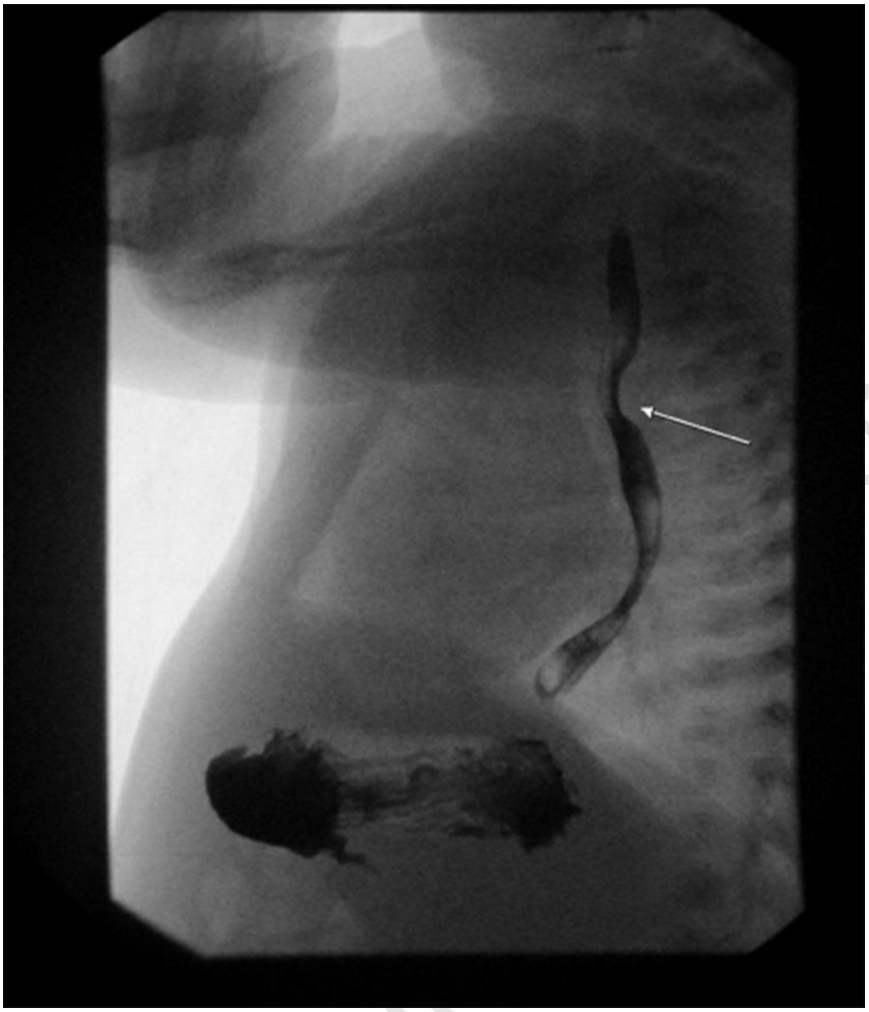

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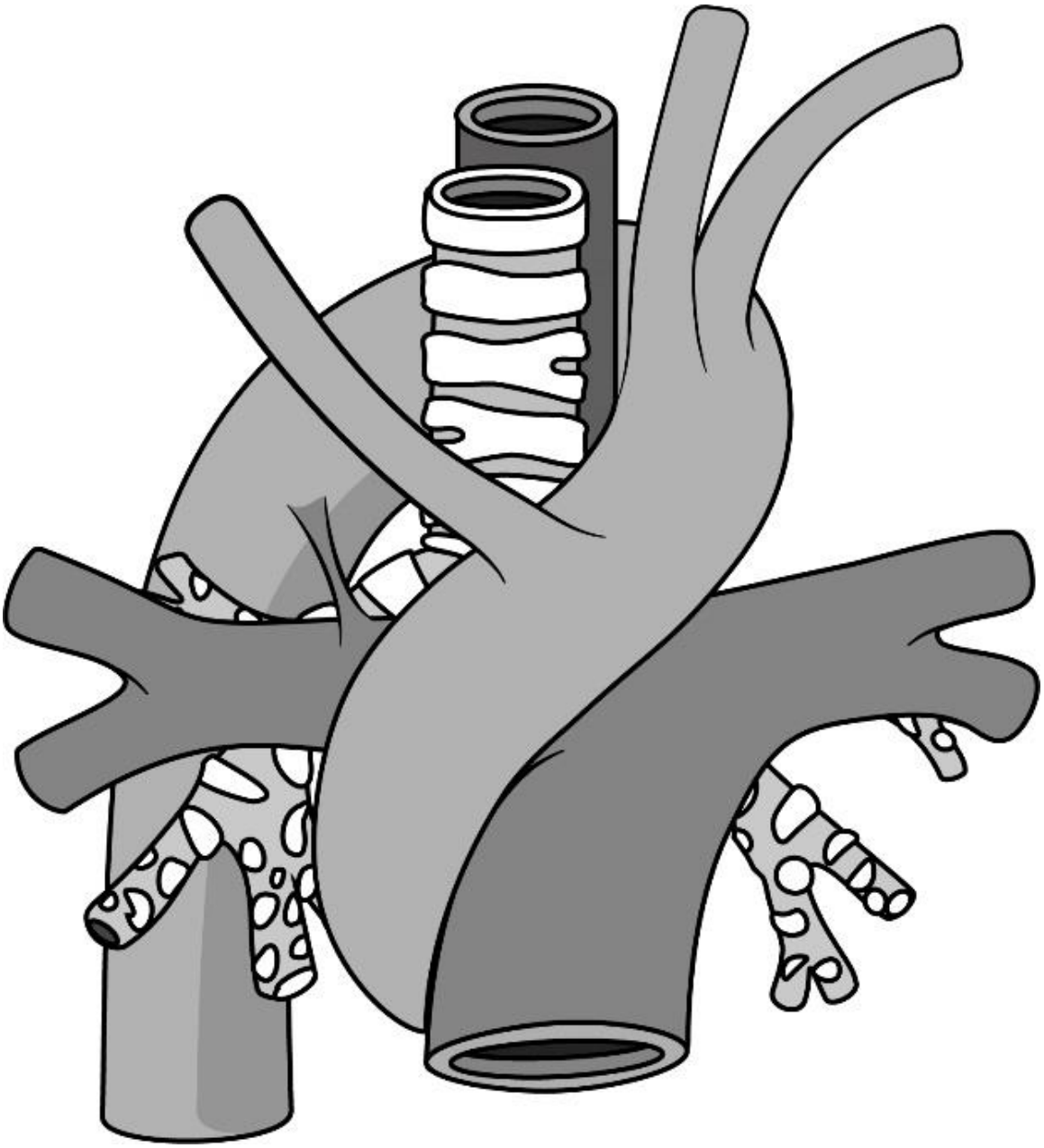

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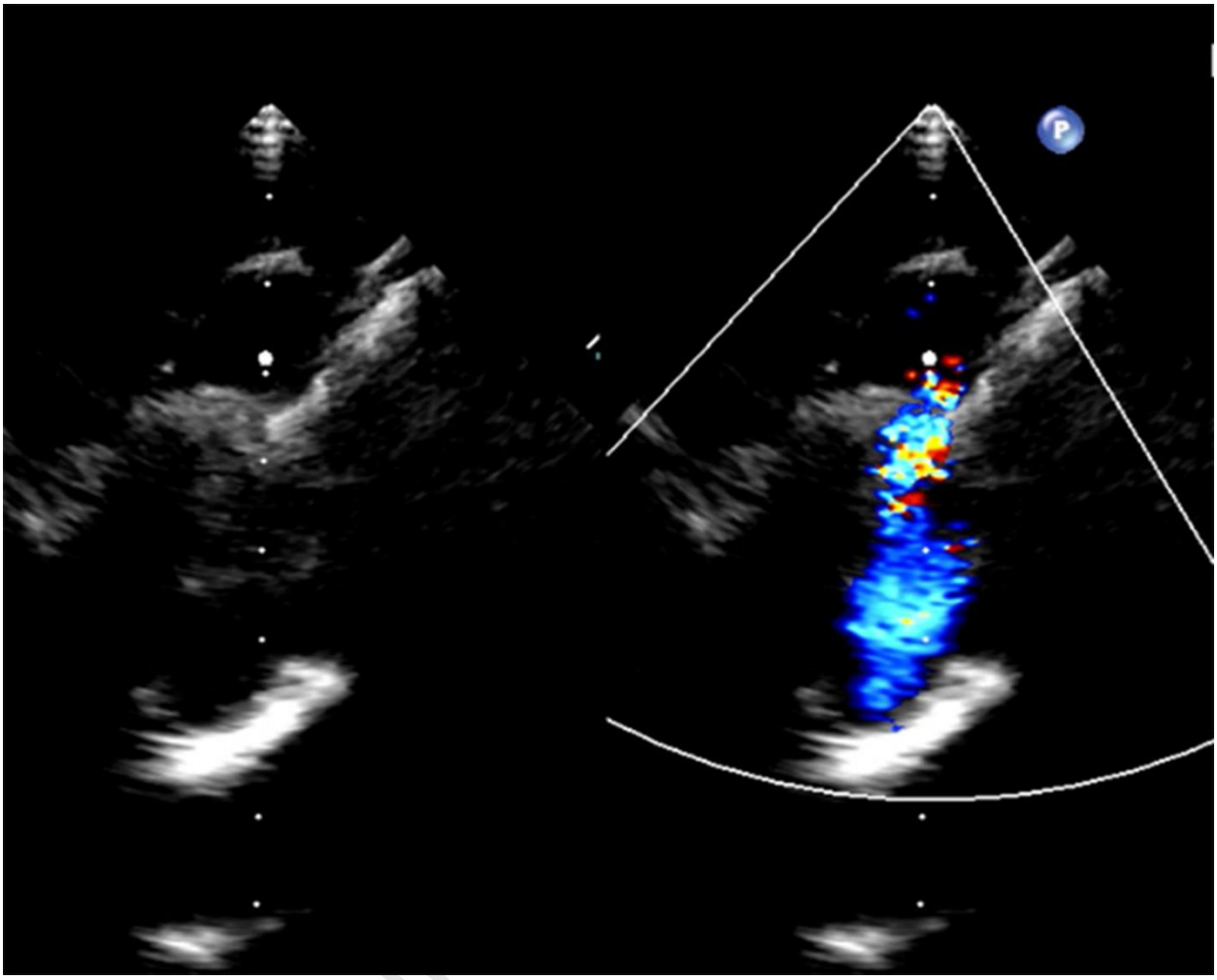

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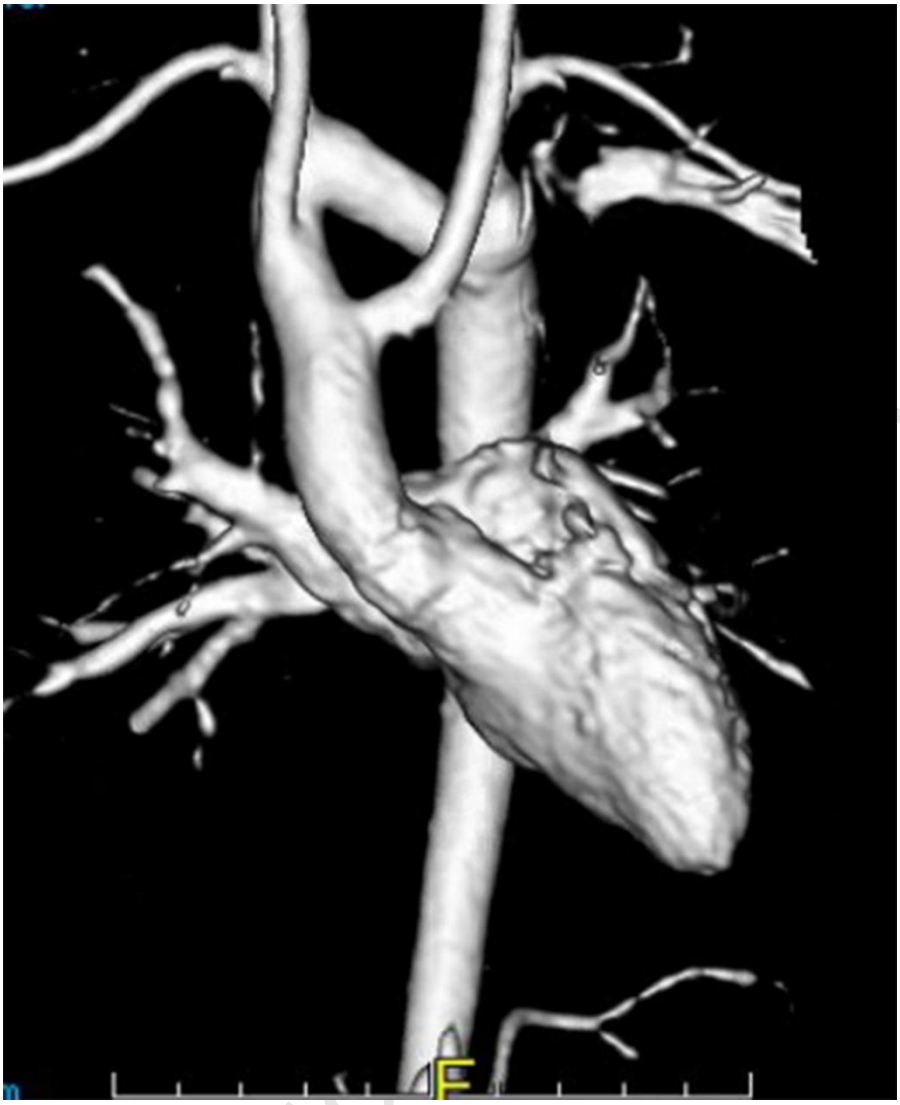

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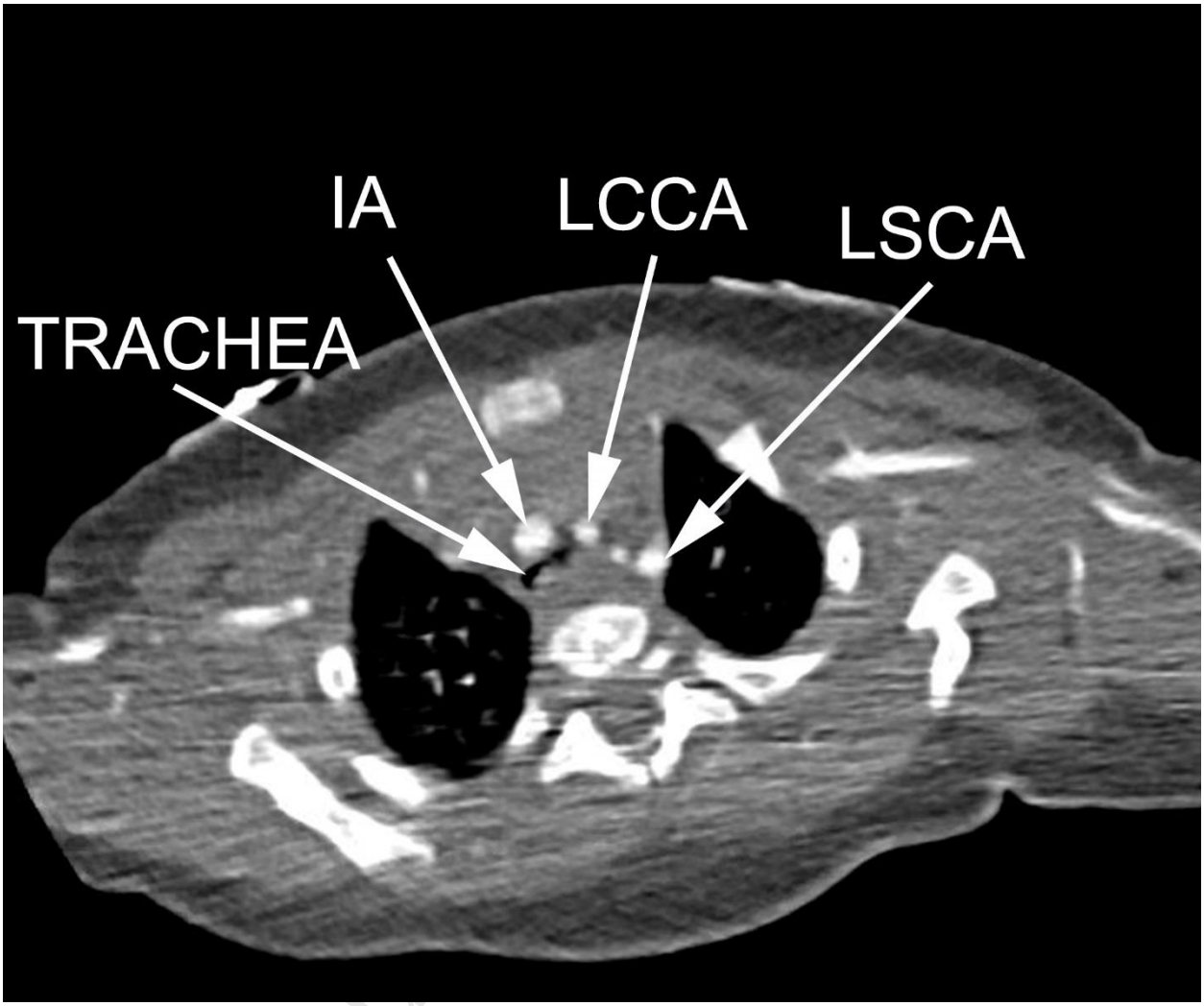

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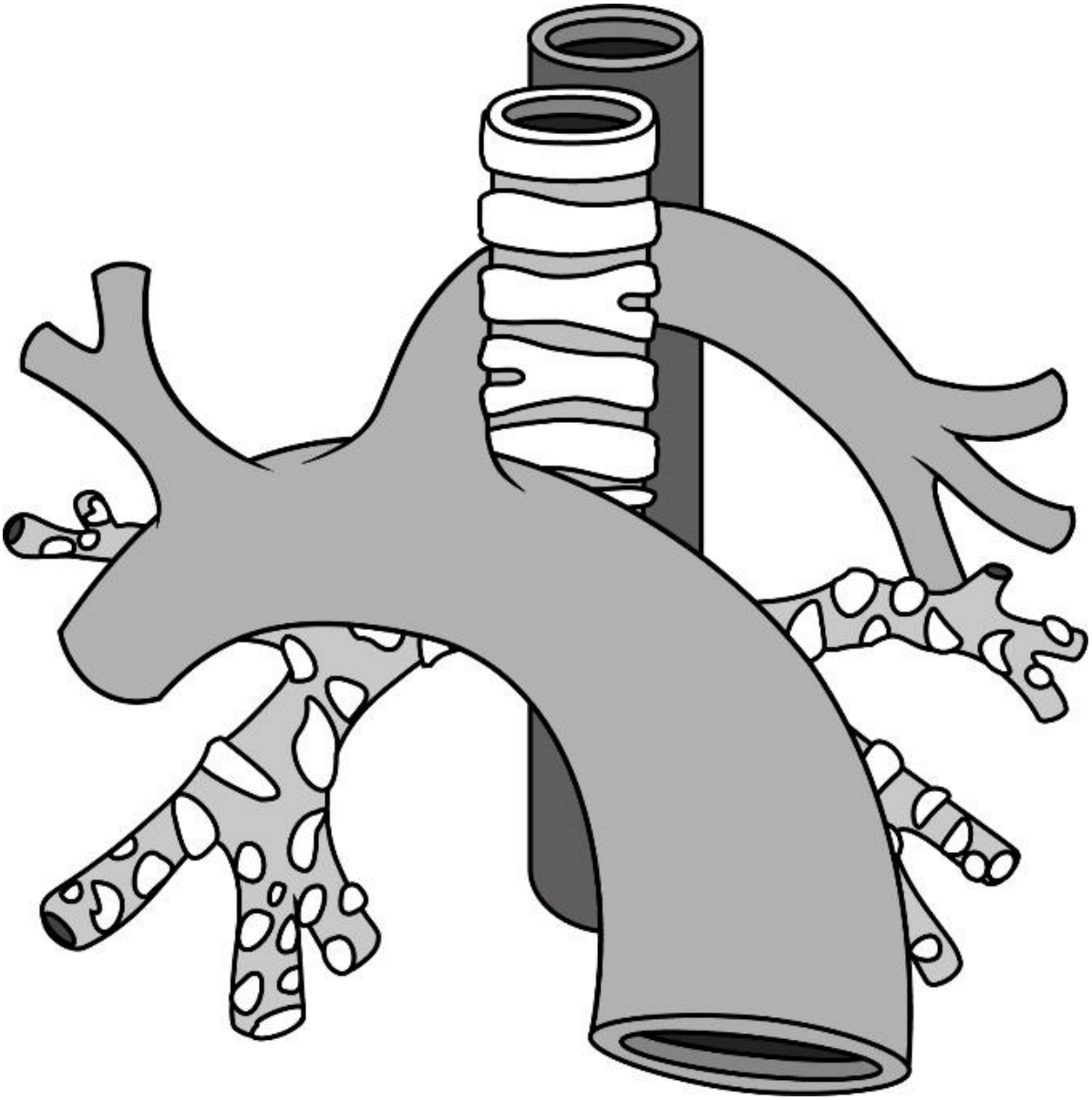

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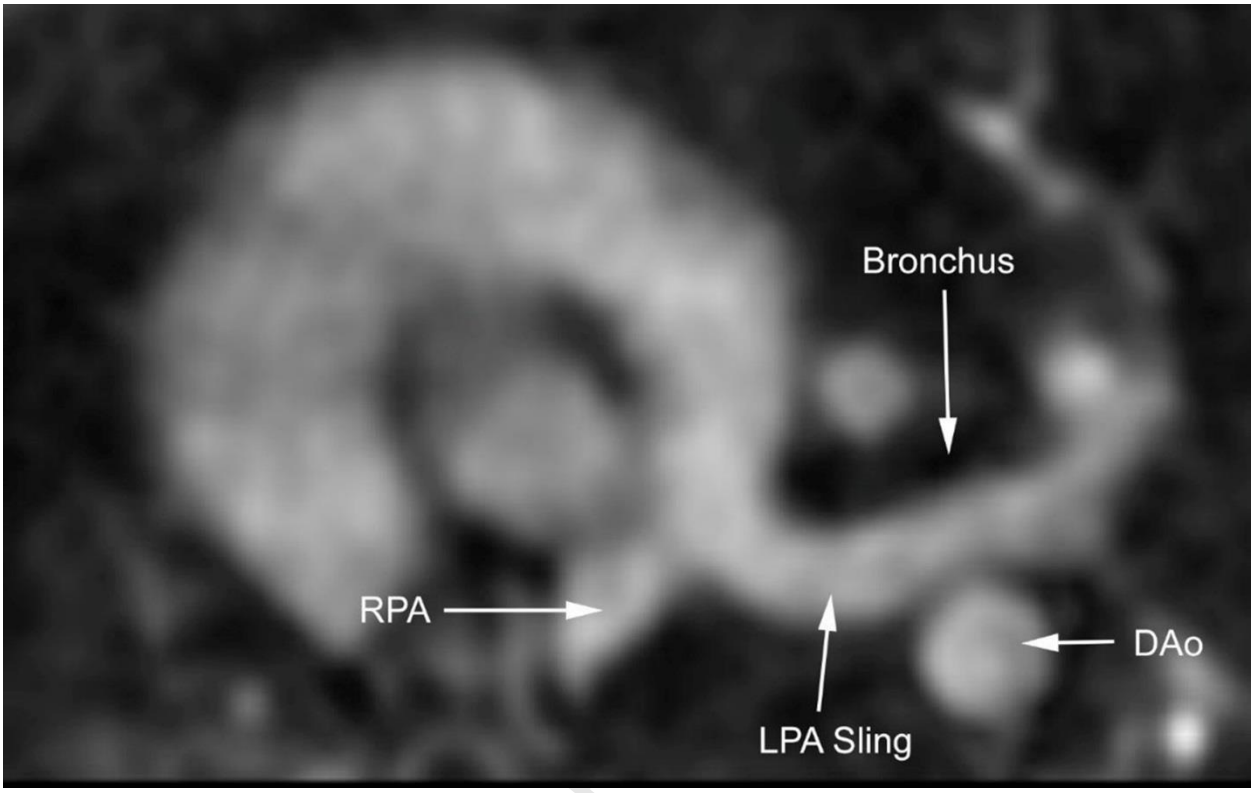

Figure Legends -

Figure.1: Illustration of Edwards hypothetical double arch with bilateral ductus arteriosum. Breaks at 1 and 2 lead to left arch; breaks at 3 and 5 lead to right arch. A break at 2 results in left arch with anomalous subclavian artery. A break at 4 results in right arch with mirror-image branching; the ductus courses from the innominate artery to the left pulmonary artery, not a vascular ring. A break at 3 leads to right arch with aberrant subclavian artery; typically, the left ductus persists, coursing from the left subclavian to the left pulmonary artery and forming a vascular ring. A break at 5 differs in that the anomalous vessel is the innominate artery. AA, ascending aorta; DA, descending aorta; E, esophagus; LCA, left carotid artery; LPA, left pulmonary artery; LSA, left subclavian artery; PA, main pulmonary artery; RCA, right carotid artery; RPA, right pulmonary artery; RSA, right subclavian artery. (4)

Figure.2: Left aortic arch. (A) Two dimensional echocardiogram image of suprasternal notch short axis, showing the bifurcation of the innominate artery into the right common carotid artery and right subclavian artery. Branching of the first arch vessel to the right, indicates a left aortic arch. (B) Standard suprasternal long axis window, demonstrating the "candy cane" view of the left aortic arch. (C) Cardiac tomography image showing the aortic arch coursing leftward of the trachea.

Figure.3: Bovine Arch. (A) Standard suprasternal sagittal view showing the left common carotid artery and innominate artery with a common origin from the aortic arch. (B) Cardiac tomography image showing a common origin of the innominate and left common carotid arteries.

Figure 4: Normal variant left aortic arch with a four vessel branching pattern. The innominate artery branches first (IA), followed by the left common carotid artery (LCCA), left vertebral artery (VA) and left subclavian artery (LSCA).

Figure 5: Left aortic arch aberrant right subclavian artery. Illustration showing the proximal segment of the aberrant right subclavian artery arising from the distal right dorsal aorta and following a retroesophageal course.

Figure.6: Left arch with aberrant right subclavian artery. (A) Echocardiogram color compare image from suprasternal notch showing the innominate artery coursing rightward without bifurcation. The color imaging shows the pulsatile flow in the aberrant right subclavian artery (ARSA). (B) Cardiac magnetic resonance image of a left arch with the posterior origin of the aberrant right subclavian artery.

Figure 7: Right aortic arch with mirror image branching (A) Illustration showing the first branch is the left brachiocephalic artery followed by the right common carotid and finally the right subclavian artery. (B) Right aortic arch with mirror image branching with left side patent ductus arteriosus from the descending aorta. (C) Echocardiogram from the suprasternal notch with superior angling towards the head. The first branch, the brachiocephalic, will course leftward and will branch into the left common carotid and left subclavian arteries.

Figure.8: Double aortic arch. Illustration showing a right and left aortic arch encircling the trachea and esophagus (with a left sided ligamentum).

Figure.9: Double aortic arch. (A) Echocardiogram image in the suprasternal short axis view, showing the "4-vessel sign" characteristic of double aortic arch. The two arches encircle the trachea and esophagus and each gives rise to two head and neck vessels. (B) Echocardiogram color compare image showing classic right and left arches of similar caliber. (C) Cardiac magnetic resonance imaging showing the double aortic arch from a birds eye view to visualize the complete ring.

Figure 10: Right aortic arch with an aberrant left subclavian artery with diverticulum illustration showing the complete vascular ring is formed by the segment of the ascending aorta anteriorly, Kommerell diverticulum posteriorly and the ligamentum arteriosum coursing on the left side.

Figure.11: Right aortic arch with an aberrant left subclavian artery. (A) Echocardiogram image from the suprasternal notch showing the innominate artery coursing leftward with the inferior aberrant vessel running parallel. (B) Cardiac tomography showing the aberrant subclavian artery from the bulbous diverticulum of Kommerell. (C) Posterior indentation noted on a barium swallow.

Figure 12: Circumflex aorta illustration showing a left aortic arch, a right descending aorta, and right ductal ligament. The vascular ring encircles the trachea and esophagus.

Figure.13: Circumflex Aorta. (A) Echocardiogram using color compare showing the tortuous course in the long axis view. Note the inability to demonstrate the entire arch in one plane. (B) Cardiac magnetic resonance imaging showing a right circumflex aortic arch that courses leftward. Note, the first branch is the left innominate artery.

Figure 14: Innominate artery compression syndrome. Computed tomography showing compression of the trachea by the innominate artery coursing anterior. Innominate artery (IA), left common carotid artery (LCCA) and left subclavian artery (LSCA).

Figure 15: Left pulmonary artery sling illustration (A) showing the left pulmonary arising distally from the proximal right pulmonary and coursing over the right mainstem bronchus, posterior to the trachea and anterior to the esophagus. (B) Cardiac magnetic resonance imaging showing an LPA sling coursing posterior to the bronchus and anterior to the descending aorta.

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Conflict of interest none