

Congenital Anomalies of Aortic Arch: A Pictorial Essay

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Abstract

Keywords

- ▶ aortic anomalies
- ▶ interrupted aortic arch
- ▶ coarctation of aorta
- ▶ right aortic arch
- ▶ aberrant subclavian artery
- ▶ double aortic arch
- ▶ cervical aortic arch

Aortic abnormalities account for 15 to 20% of all congenital cardiovascular diseases. The purpose of this pictorial essay is to illustrate various congenital anomalies of the aortic arch, which influence patient management and outcome.

Introduction

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Imaging Protocol

All patients underwent multidetector computed tomography (MDCT) angiography examinations, performed with a 128 slice MDCT (Discovery CT750 HD; GE Healthcare, Milwaukee, WI, USA). Images were acquired during a single breath-hold with patients in the supine position and extending from the base of the neck to the diaphragm after administration of intravenous nonionic contrast agent. In adult patients, automatic tube current dose modulation was used. In pediatric patients, a body weight-based low-dose protocol was used depending on patients age and surface area. The examination was initiated 6 seconds after the

attenuation of a region of interest positioned in the ascending aorta reached 100 Hounsfield units. For three-dimensional image reconstruction, the raw MDCT data were processed on a separate workstation (Advanced Workstation 2.0; GE Healthcare).

Embryologic Development of the Aorta

To understand the various aortic arch anomalies, a quick look at embryology is essential. The two most popular theories proposed for the embryological development of the aorta are the Rathke's diagram and the Edwards hypothetical double arch.

According to Rathke's diagram, the branchial apparatus consisting of six paired branchial arches connect paired dorsal and the ventral aortae (▶ **Fig. 1**). The third arch along with the parts of the ventral and dorsal aortae forms the arteries of the head and neck, the fourth arch forms the aortic arch, and the sixth arch forms the pulmonary arteries. The dorsal component of the sixth arch on the left side forms

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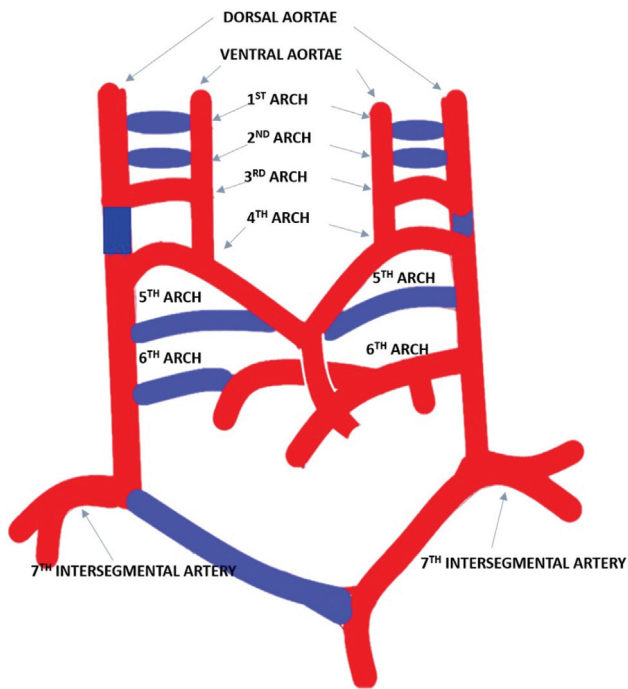


Fig. 1 Schematic representation of Rathke's diagram. Red areas represent segments that persist, while blue areas represent segments that normally involute.

the ductus arteriosus and the intersegmental arteries form the subclavian arteries.^{1,2}

According to the Edwards hypothetical double arch theory, a double aortic arch (DAA) with a ductus arteriosus exists on each side (→ **Fig. 2**). The two aortic arches give rise to bilateral common carotid arteries and subclavian arteries as individual arteries.¹

Aortic Arch Variants and Anomalies

Interrupted Aortic Arch

In interrupted aortic arch (IAA), there is anatomic discontinuity between the ascending and the descending aorta. The interruption may be complete, or an atretic fibrous band may be seen connecting two segments. The descending aorta is supplied by patent ductus arteriosus (PDA) (→ **Figs. 3 and 4**).

IAA is classified into three types:

Type A IAA: Interruption occurs distal to the origin of the left subclavian artery.

Type B IAA: Interruption occurs between the origins of the left common carotid and left subclavian arteries. It is the most common type.

Type C IAA: Interruption occurs between the origins of the brachiocephalic and left common carotid arteries.³

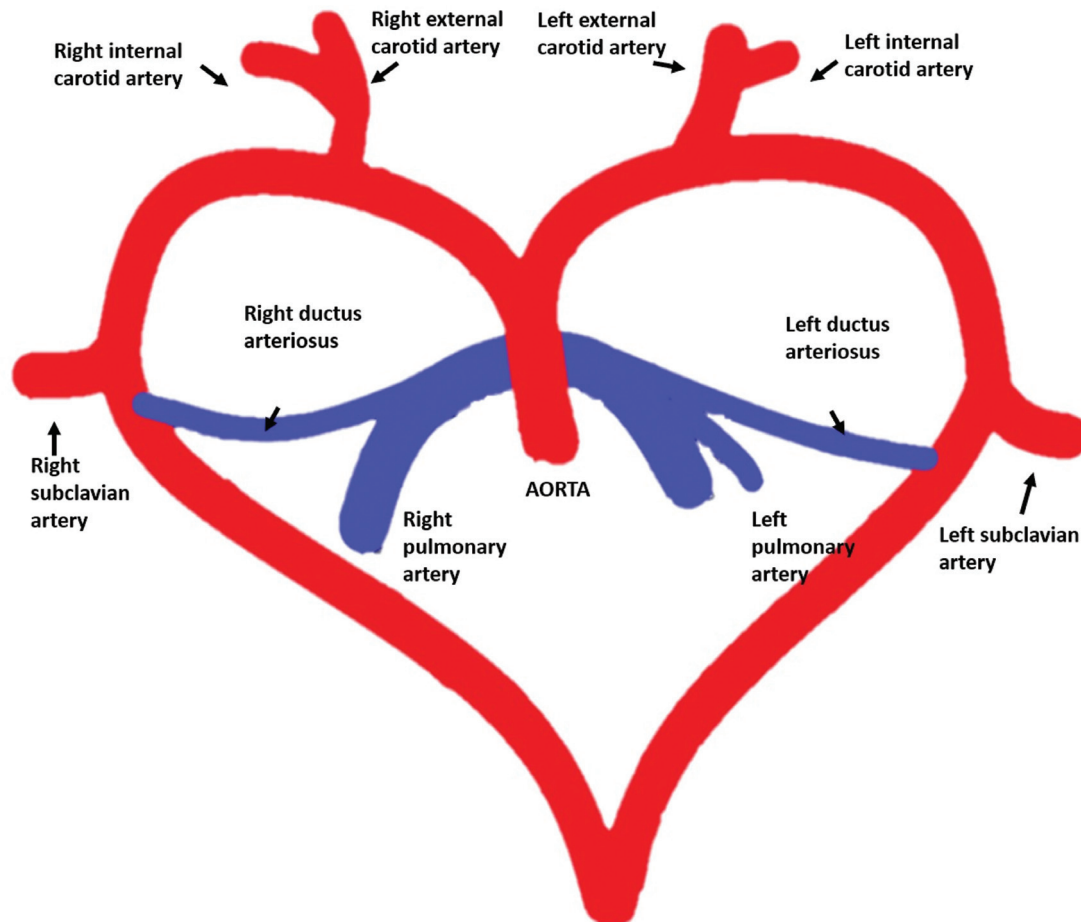


Fig. 2 Schematic diagram of the Edwards hypothetical double arch.

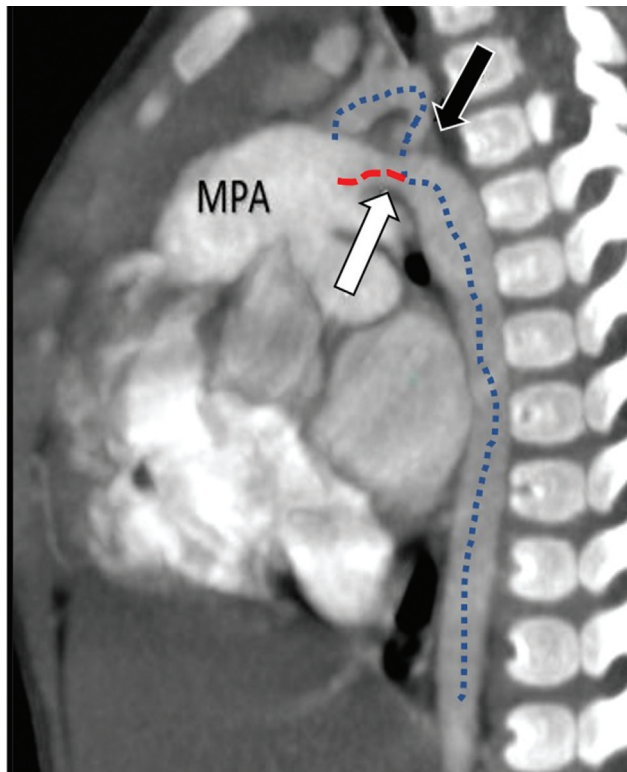


Fig. 3 A 14-month-old female child with failure to thrive. Computed tomography sagittal image shows the interruption (*black arrow*) of aorta (*blue dotted line*). Patent ductus arteriosus (*white arrow*) supplying the descending thoracic aorta. Flattened vascular (ductal) arch is shown in *red dashed line*. MPA, main pulmonary artery.

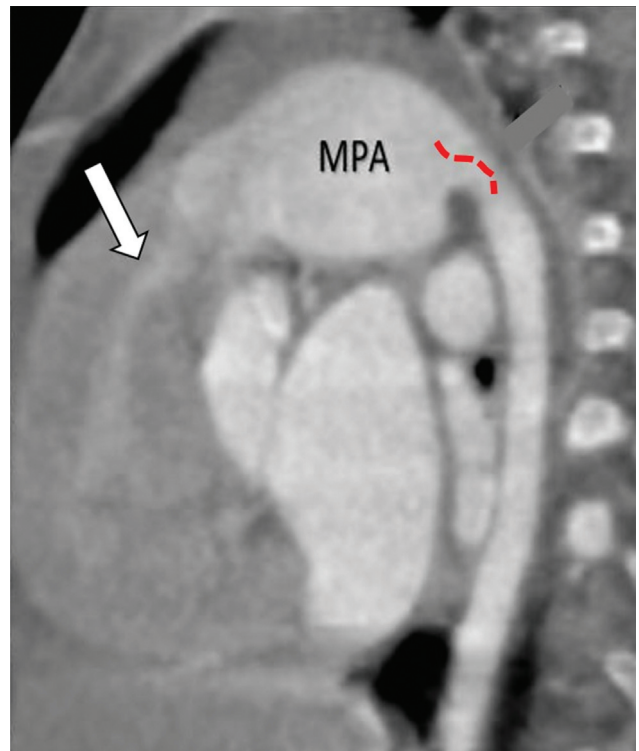


Fig. 4 A 14-month-old female child with failure to thrive. Computed tomography sagittal image shows infundibular pulmonary stenosis (*white arrow*). Flattened vascular (ductal) arch is shown in *red dashed line*, supplying the descending thoracic aorta.

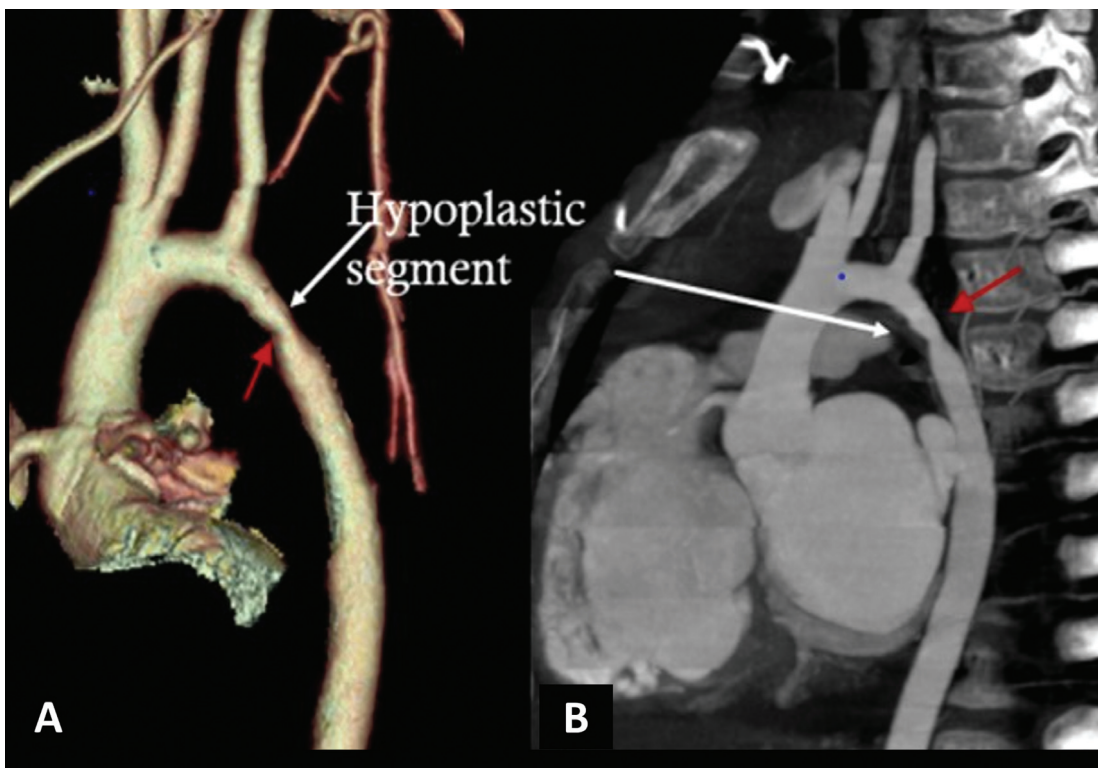


Fig. 5 A 16-year-old female child with fatigue and atypical chest pain. Computed tomographic images show mild hypoplasia of distal aortic arch and isthmus (*white arrows—A and B*) and coarctation of aorta (*red arrows—A and B*) in the isthmus.

IAA is associated with other congenital heart diseases in 98% of cases with the most common being PDA (97%) followed by ventricular septal defect (VSD).³

Hypoplasia

It refers to the external diameter of the proximal or distal aortic arch being less than 60 or 50% of the ascending aortic diameter, respectively. It is commonly associated with coarctation of aorta (COA; ►Fig. 5). Hypoplasia of isthmus is defined as external diameter less than 40% than that of the ascending aorta.³

Coarctation of Aorta

It refers to discrete, focal constriction of the aortic segment, which is usually juxta ductal in location and more commonly affects male infants.³ It is of two types: preductal (constriction is proximal to ductus arteriosus) (►Figs. 6 and 7) and postductal COA (constriction is distal to ductus arteriosus).⁴

Clinical manifestations depend on the degree and location of luminal narrowing, the extent of the collateral circulation, and associated congenital heart diseases. Adults with postductal COA have systemic arterial hypertension in the arms with a diminished femoral arterial pulse. Treatment is surgical repair.⁴

Left Aortic Arch with an Aberrant Right Subclavian Artery

Aberrant right subclavian artery (ARSA) is the last branch of the aortic arch. It has retroesophageal course and supplies the right upper extremity (►Fig. 8).¹ Patients are usually asymptomatic. Retroesophageal course of the ARSA may result in esophageal compression causing dysphagia lusoria.⁵ ARSA along with persistent right ligamentum arterio-



Fig. 7 A 23-year-old female with limb claudication and dyspnea on exertion. Maximum intensity projection sagittal image shows fusiform aneurysm of the ascending aorta (black arrow) and coarctation of aorta (white arrowhead).

sum may result in true vascular ring causing airway compression.⁶

Treatment is required in a symptomatic patient in the presence of a true vascular ring or the aneurysm of the Kommerell's diverticulum.¹

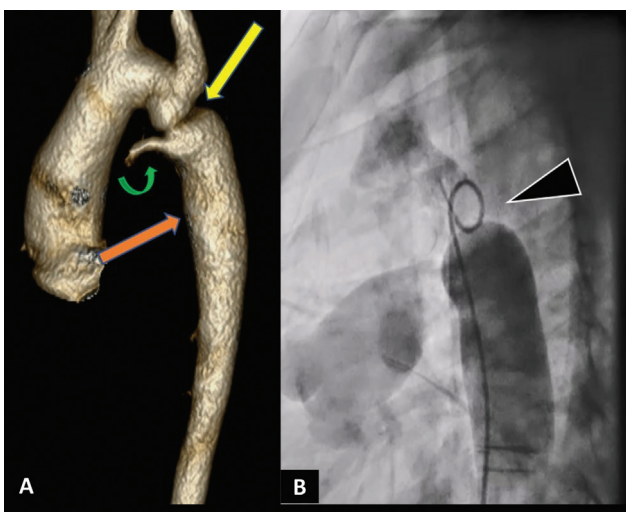


Fig. 6 A 13-year-old male child with lower limb claudication. Computed tomographic image of aorta (A) shows preductal coarctation of aorta (COA) (yellow arrow) with poststenotic dilatation (orange arrow) and patent ductus arteriosus (curved green arrow) supplying descending aorta. Catheter angiography image (B) shows tight COA (black arrowhead).

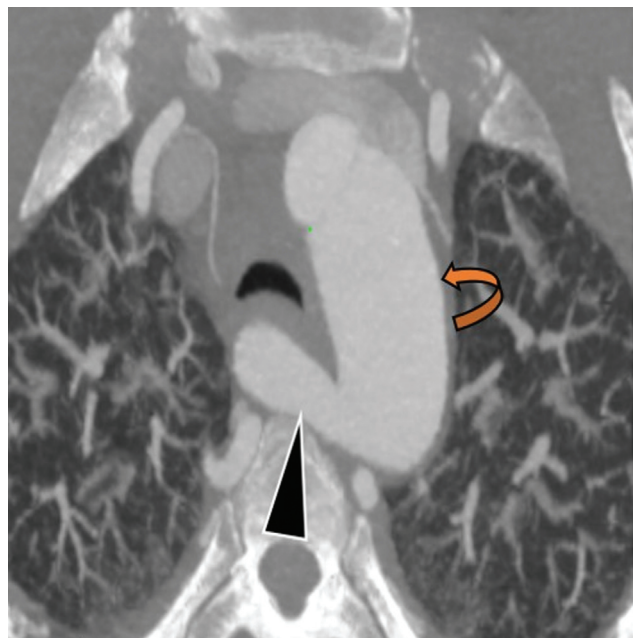


Fig. 8 Computed tomography maximum intensity projection axial image shows left aortic arch (curved orange arrow) with the aberrant right subclavian artery (black arrowhead).

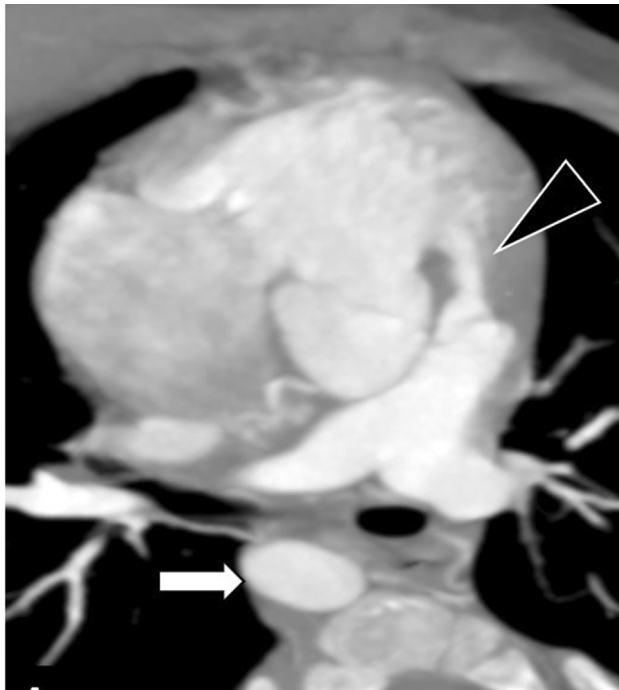


Fig. 9 A 16-month-old female child with tetralogy of Fallot. Computed tomography maximum intensity projection image shows infundibular pulmonary stenosis (black arrowhead) and right-sided aortic arch (white arrow).



Fig. 10 A 16-month-old female child with tetralogy of Fallot. Maximum intensity projection coronal image shows the right aortic arch (white arrow) with mirror imaging of branch vessels.

Left Aortic Arch along with Isolation of the Subclavian Artery

It is most commonly associated with right-sided aortic arch. There is loss of continuity between the aortic arch and the subclavian artery. The isolated subclavian artery is supplied by the vertebral artery collaterals or the ductus arteriosus.^{1,7} It may be associated with tetralogy of Fallot (TOF), IAA, or PDA. Treatment surgical correction by reconnecting the isolated subclavian artery to the aorta.^{1,7}

Right Aortic Arch

It results from the regression of the left dorsal aortic root instead of the right dorsal root.^{1,8} Right aortic arch (RAA) has three subtypes¹:

1. Type I: RAA with mirror image branching (→Figs. 9 and 10). It is associated with congenital heart diseases such as TOF, pulmonary atresia with VSD, tricuspid atresia, and truncus arteriosus in ~75% of cases.¹
2. Type II: RAA with an aberrant left subclavian artery (ALSA). It is the most common type of RAA where ALSA arises as the last branch from the arch (→Fig. 11) or



Fig. 11 A 9-year-old female child with cyanotic heart disease. Computed tomographic images show pulmonary atresia (black arrowhead—A) with multiple major aortopulmonary collaterals (white arrows—A), type II right aortic arch (white arrowhead—B) and aberrant left subclavian artery (black arrow—B). Sagittal image C shows cervical aortic arch extending up to the clavicle (curved black arrow).

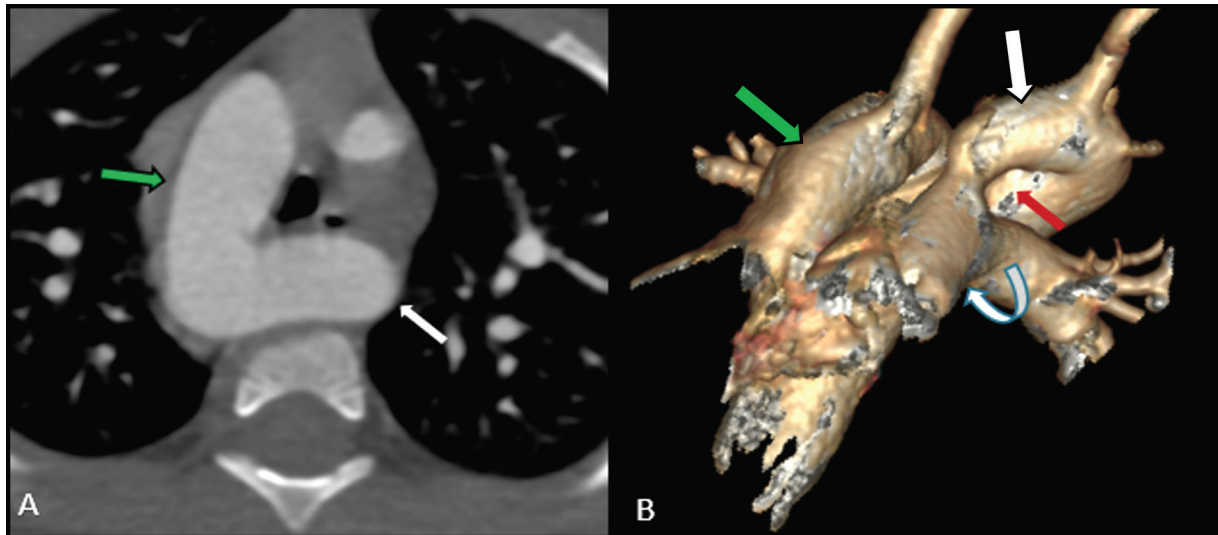


Fig. 12 A 25-month-old female child with failure to thrive. Type II right aortic arch with Kommerell's diverticulum and left persistent ductus arteriosus (vascular ring). Computed tomographic images show right aortic arch (green arrow—A) with Kommerell's diverticulum showing retroesophageal course (white arrow—A), left patent ductus arteriosus (red arrow—B) from the left proximal pulmonary artery (curved white arrow—B) joining the Kommerell's diverticulum (white arrow—B).

Kommerell's diverticulum (►Figs. 12–14). When it is associated with left ligamentum arteriosum, a vascular ring forms that can compress the trachea and esophagus (►Figs. 12–1314),^{1,3,9} Surgical treatment involves resection of the Kommerell's diverticulum (if present) and reimplantation of the ALSA to the aortic arch.¹

3. Type III: RAA, along with isolation of the left subclavian artery. This anomaly occurs due to regression of the segments of aortic arch proximal and distal to the left subclavian artery.^{3,10} Congenital subclavian steal phenomenon may occur if retrograde filling of LSCA occurs from the vertebral or craniocervical arteries.^{1,3}

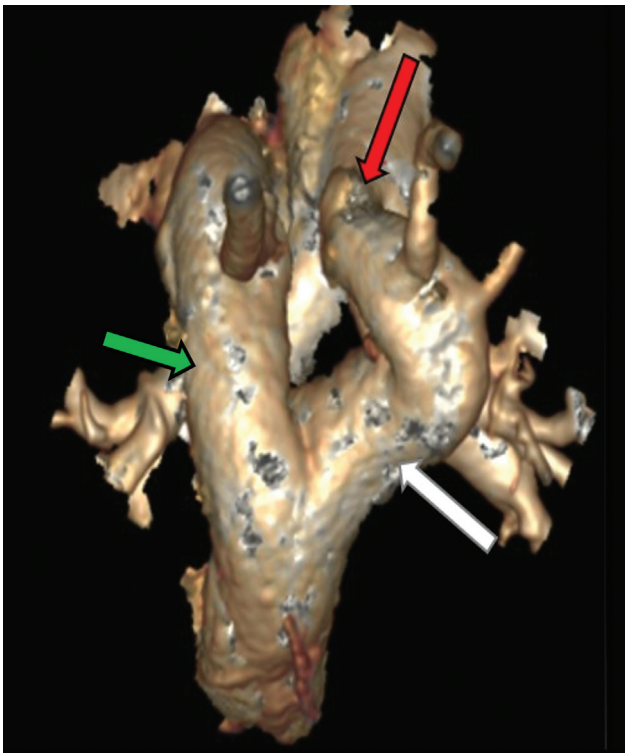


Fig. 13 Computed tomographic volumetric rendering technique image shows right aortic arch (green arrow) with Kommerell's diverticulum (white arrow), left patent ductus arteriosus (red arrow) joining the Kommerell's diverticulum and forming a vascular ring.

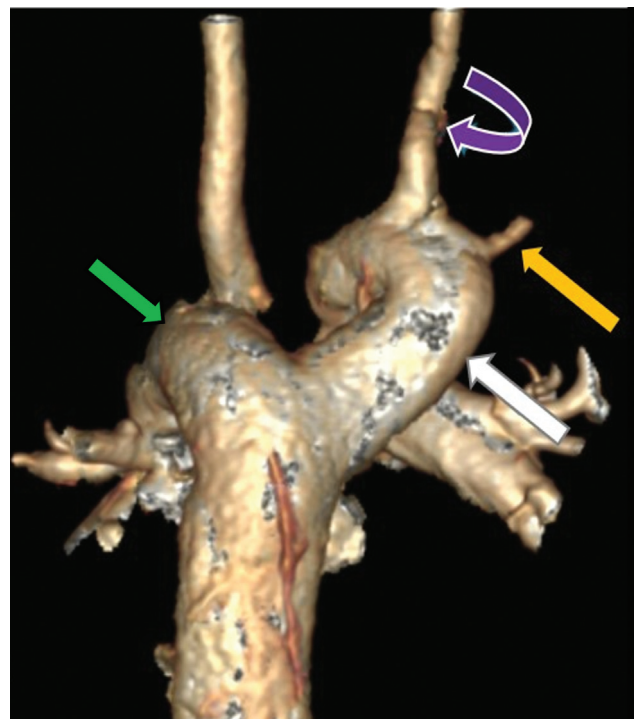


Fig. 14 Computed tomographic image shows right aortic arch (green arrow). Left subclavian artery (yellow arrow) and left common carotid artery (curved purple arrow) are arising from the Kommerell's diverticulum (white arrow).

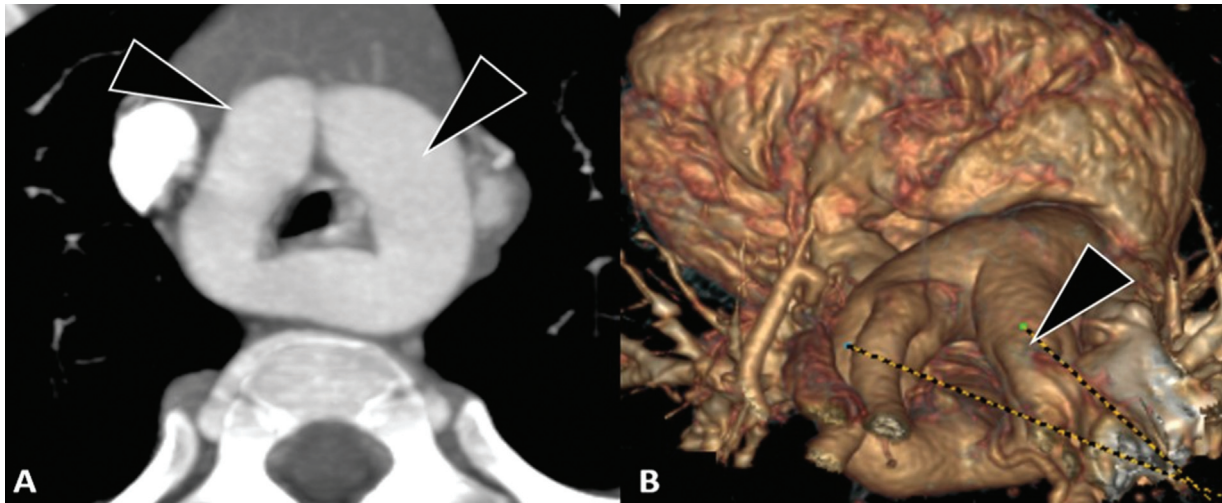


Fig. 15 A 7-year-old male child with cyanosis and wheezing. Computed tomographic images (A and B) show complete double aortic (black arrowheads) forming a vascular ring.

Double Aortic Arch

This anomaly results from the persistence of both the right and left arches (Edward hypothetical double arch theory) (→Fig. 15). Each arch gives rise to separate ipsilateral carotid and subclavian arteries (four-vessel sign on axial CT image) (→Figs. 16 and 17). Ductus arteriosus is most commonly present on the left side. Ascending aorta bifurcates anterior to the trachea and reunites posterior to the esophagus forming a vascular ring and a single descending aorta.¹ Right arch is usually the dominant arch (75% cases) with the left-sided descending thoracic aorta.^{11,12} The smaller (usually left) arch may be atretic in which case it is called incomplete DAA (→Fig. 18).⁶

DAA is the most common cause of symptomatic vascular ring with patients presenting with wheezing and stridor exacerbated on crying, cyanosis, dysphagia, etc.¹ Rarely, it is associated with other congenital heart diseases such as TOF (most common) or transposition of great arteries.² The treatment comprises of surgical division of the vascular ring to relieve compression of the trachea (→Fig. 19) and esophagus.¹

Cervical Aortic Arch

It refers to the aortic arch that is abnormally placed higher upper in the neck than its usual mediastinal position at the level of the fourth thoracic vertebra, extending above the clavicles (→Fig. 11). It is more commonly present on the right side. Usually, it is an asymptomatic anomaly; however, it can

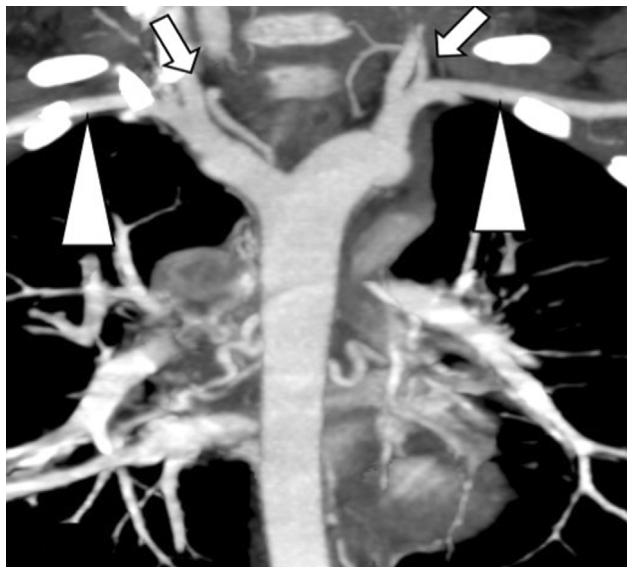


Fig. 16 Computed tomographic maximum intensity projection image shows double aortic arch, each arch giving rise to ipsilateral subclavian (white arrowhead) and carotid (white arrows) arteries.

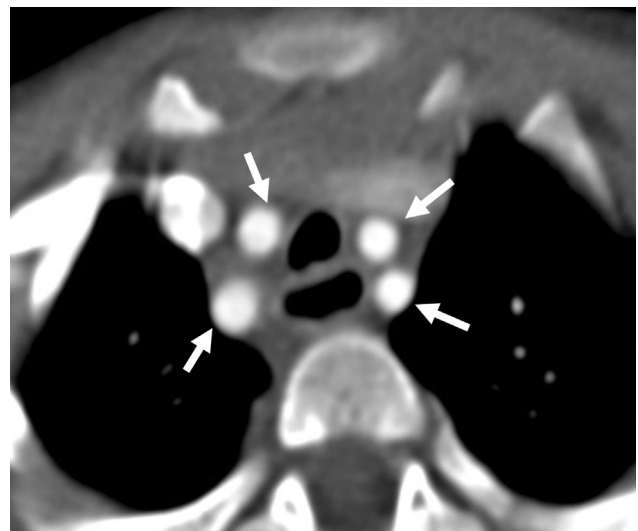


Fig. 17 Computed tomographic axial image of a patient with double aortic arch shows "four vessels sign" (white arrows).

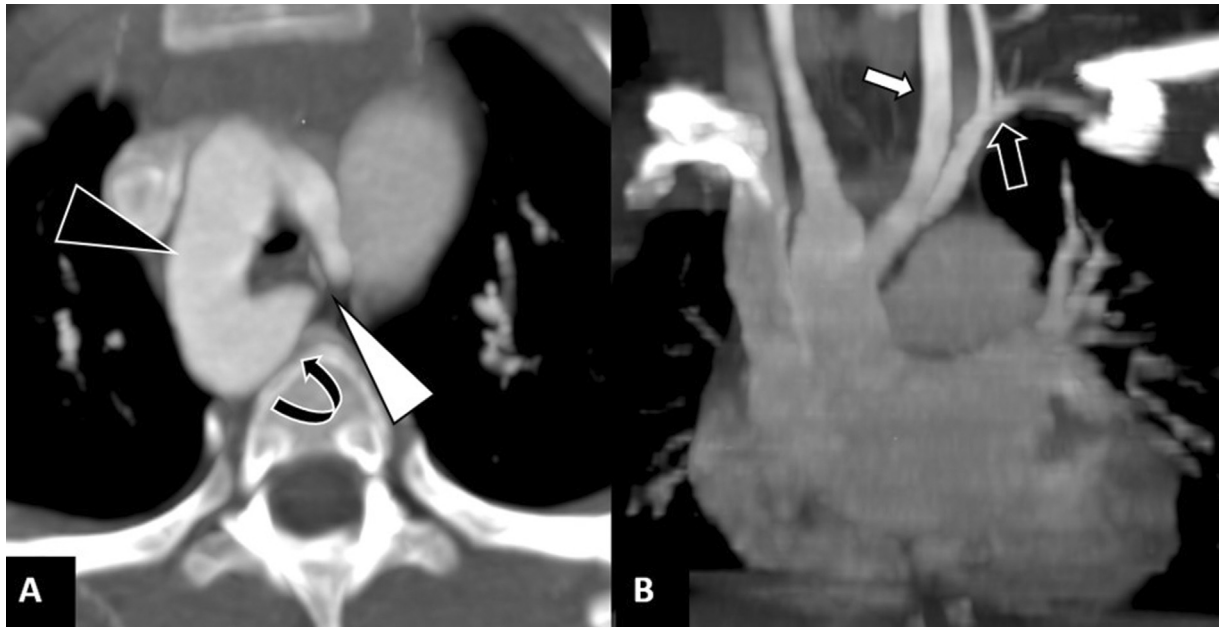


Fig. 18 A 3-year-old male child with tetralogy of Fallot. Computed tomographic images show incomplete double aortic arch, dominant right arch (black arrowhead—A), retroesophageal Kommerell's diverticulum (curved black arrow—A), and distal atresia of the nondominant left arch (white arrowhead—A). Left common carotid (white arrow—B) and left subclavian arteries (black arrow—B) arising from left arch proximal to atresia.



Fig. 19 A 7-year-old male child with complete double aortic arch. Tracheal compression is seen due to vascular ring (curved black arrow).

present with a pulsatile neck mass or with symptoms arising due to tracheal or esophageal compression.^{1,3}

Conclusion

Early and accurate identification of the aortic arch anomalies is of utmost importance for patient management, preoperative planning, and determining postoperative patient prognosis. This pictorial essay will not only allow the cardiac imagers to promptly identify these anomalies but will also

allow them to understand the pathophysiology, treatment options available for various conditions, and guide the clinicians suitably.

Presentation at a Meeting

Sent as e-poster educational exhibit to ECR 2020 and RSNA 2020.

Conflict of Interest

None.

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