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LETTER TO EDITOR CODEN: AAJMBG

Double Trouble - Radiological Abnormality

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Dear Editor:

Double aortic arch (DAA) is a rare congenital anomaly of the aortic arch system, representing the most common vascular ring malformation. Axial and Coronal CT aortogram images of our patient demonstrate a prominent right arch and a smaller left arch forming a complete vascular ring, causing trachea-esophageal compression. This condition can lead to significant respiratory and digestive symptoms due to the encirclement of the trachea and esophagus by connected segments of the aortic arch.

DAA originates from the failure of the right-sided aortic arch to regress during embryogenesis, leaving both right and left arches intact. This developmental anomaly is linked to chromosomal abnormalities, including 22q11 micro deletion (DiGeorge syndrome) and trisomy 21, as well as associated cardiac malformations such as Tetralogy of Fallot, ventricular septal defects, and coarctation of the aorta.

The clinical presentation of DAA often involves nonspecific respiratory symptoms, such as stridor, chronic cough, recurrent respiratory infections, or even life-threatening airway obstruction, making diagnosis challenging. Computed tomography angiography with three-dimensional reconstruction remains the diagnostic modality of choice, providing detailed visualization of the aortic arches and their impact on surrounding structures. Bronchoscopy can further assess the severity of tracheal compression and rule out alternative causes of airway obstruction.

Prompt surgical intervention is essential in cases of severe trachea-esophageal compression, with excellent long-term outcomes reported in patients undergoing repair. Early diagnosis and treatment are critical to prevent complications such as recurrent infections, failure to thrive, or airway compromise.

This case underscores the importance of recognizing the clinical and imaging features of DAA, enabling timely diagnosis and intervention to optimize patient outcomes.

Question: This is the CT aortogram scan of a patient who was admitted with history of recurrent lower respiratory tract infection. What is the radiological abnormality?

Fig-1 (A & B): Axial CT images showing (R) sided dominant aortic arch (Red arrow) and (L) sided small arch (blue arrow) forming a complete vascular ring around trachea and esophagus.

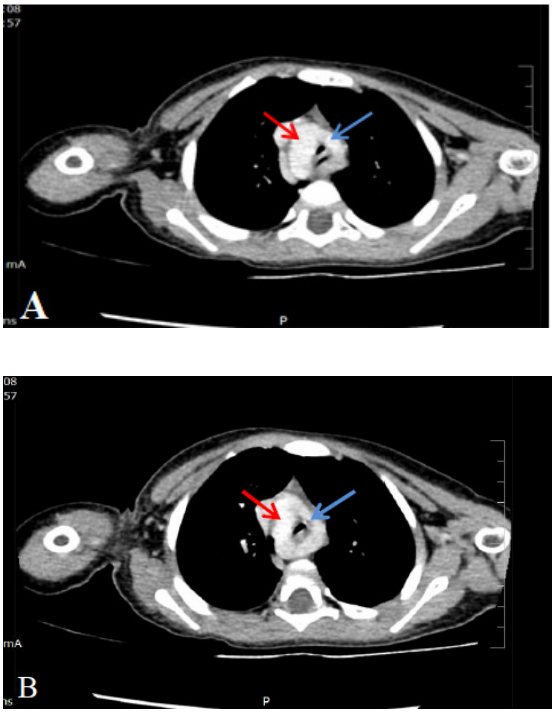

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Fig-2: Coronal CT images showing (R) sided dominant aortic arch (Red arrow) and (L) sided small arch (blue arrow)

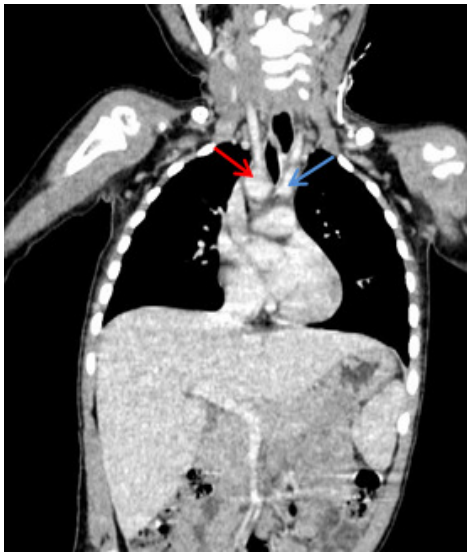
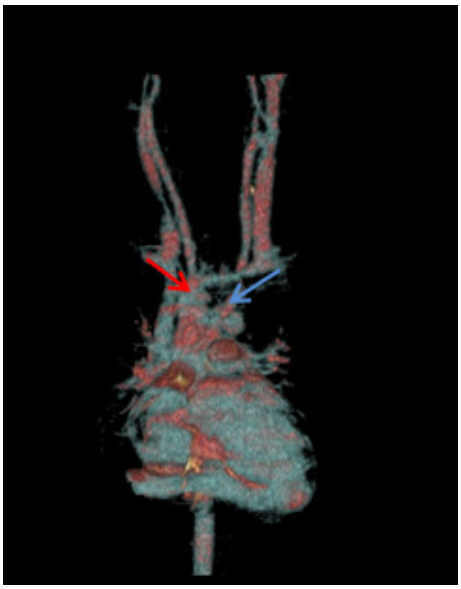


Fig 3: 3D reconstruction image showing (R) sided dominant aortic arch (Red arrow) and (L) sided small arch (blue arrow)



Answer: **Double Aortic Arch (DAA)**;

Axial (Figure 1- A & B) and Coronal (Figure 2) CT aortogram images shows Double aortic arch with prominent right arch and smaller left arch forming a complete vascular ring causing trachea-esophageal compression. (Figure 3) is the 3D Reconstruction image of Double Aortic Arch. Aortic arch malformations are anomalies which originate from disordered embryogenesis of branchial arches [1]. Double aortic arch (DAA) is the most common type of vascular ring malformation, in which the trachea and esophagus are completely encircled by connected segments of the aortic arch and its branches that can result in life-threatening airway obstruction [1-2].

First post-mortem case was described in 1737 by Hommel [2] Clinical syndrome of vascular compression produced by a DAA was first reported by Wolman in 1939 [1]. And in 1947, Robert Gross carried out first documented surgical repair of DAA [2]. The incidence of double aortic arch is generally unknown but published data estimates the prevalence of vascular rings to be 1% [2]. Also there is an association of DAA with chromosome 22q11 microdeletion (DiGeorge syndrome), trisomy 21 (down syndrome) and other cardiac malformations such as Fallots tetralogy, ventricular septal defect, coarctation of the aorta [3].

Aortic arch development is a complex process that occurs between 2nd to 7th weeks of gestational life [2]. Six pairs of arches appears and regress in a sequential fashion leaving remnants that form important blood vessels [2]. Table-1 shows the derivatives of the Six Branchial arches.

Table-1: Branchial arches and its derivatives	
First and Second pair	Disappears leaving the maxillary, hyoid, and stapedial arteries.
Third pair	Common carotid arteries and a small portion of the internal carotid arteries.
Fourth pair	*Bilateral aortic arches* which later forms (R) side – Subclavian artery & (L) side – Aortic arch.
Sixth pair	Pulmonary arteries on each side. Ductus arteriosus on left side.

In 5th week of gestation, the right-sided aortic arch regresses and the left-sided arch remains, leaving a normal left-sided aortic arch [2]. The formation of a DAA originates from a failed regression of the right-sided arch with the persistence of the left-sided arch [2]. Depending on its anatomical characteristics,

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DAA has been divided into three types. Right dominant aortic arch - Large right posterior arch and small left anterior arch (75%) as seen in our patient. Left dominant aortic arch - small right rear arch and large left front arch (20%). Balanced type aortic arch - equal diameter DAA (5%) [1].

DAA can be difficult to diagnose because the symptoms are not typical of a cardiac disorder. The clinical picture is dominated by respiratory symptoms like stridor, dyspnea, chronic cough, recurrent lower respiratory tract infections (i.e non resolving pneumonias) and digestive symptoms like dysphagia, vomiting, feeding intolerance [4-5]. Sometimes the diagnosis can be confused with laryngomalacia, tracheomalacia, asthma, subglottic stenosis, viral induced wheeze, bronchiolitis or recurrent pneumonia.

Computed tomography (CT) angiography with three-dimensional reconstruction is an excellent non-invasive imaging technique that is diagnostic procedure of choice, permitting clear delineation of the aortic arches, its location and branching pattern, arch dominance and extend of trache-oesophageal compression. Echocardiography to look for co-existent intra cardiac abnormalities. Bronchoscopy plays a pivotal role to rule out other airway causes for respiratory symptoms and to evaluate airway anatomy pre-operatively. It may also help to confirm and assess the severity of airway compression [6].

Surgical repair remains the mainstay of treatment that is indicated if the trachea-esophageal compression syndrome is severe with repeated lower respiratory tract infections and life threatening airway compression which can be complicated by cardiopulmonary arrest [3]. It was noted that among the ones who underwent double aortic arch repair, had a very good outcome. Postoperative complications like aorto-esophageal fistula and esophageal erosion are very rare [7].

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