RIGHT-SIDED AORTIC ARCH WITH ABERRANT LEFT SUBCLAVIAN ARTERY AND DUPLICATION OF SUPERIOR VENA CAVA

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PRESENTATION OF CASE

Right-sided aortic arch is a rare anatomical variant present in about 0.1% of the adult population.[1,2] Half of the cases are associated with an aberrant left subclavian artery (0.05%-0.1%). Right-sided aortic arch with aberrant left subclavian artery is less common than left-sided aortic arch with aberrant right subclavian artery (0.5-2.0%).[3,4] A right-sided aortic arch is an anatomic variant resulting from persistence of the right fourth aortic arch and involution of the left. It can be associated with an aberrant left subclavian artery arises from Kommerell's diverticulum. It is usually asymptomatic and diagnosed incidentally during adult age.

A 40-year-old male presented with cough and a hump in the back. The patient was evaluated for scoliosis and plain CT thorax was done.

Contrast-enhanced computed tomography (CECT) of the thorax was done to evaluate the thoracic aorta. The right-sided aortic arch was confirmed by the thoracic aorta descending on the right side of the spine. There was an aberrant left subclavian artery arising from an aortic arch diverticulum (Kommerell's diverticulum). The branches of the right-sided aortic arch from right to left were right subclavian, right common carotid, left common carotid and left subclavian arteries. The Kommerell's diverticulum was posterior to the trachea and oesophagus. However, there was no compression of the oesophagus and trachea. The anteroposterior diameter of the diverticulum was 2.5 cm. The patient also had duplication of superior vena cava, right-sided hemivertebra of D2 with dextroscoliosis and a genetic left thyroid lobe. Since, he was asymptomatic, there was no interventional procedure planned for him.

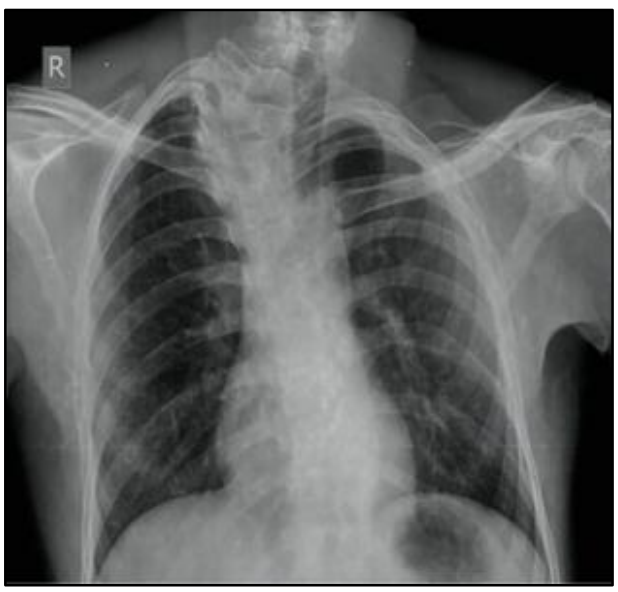

Figure 1. Chest X-Ray PA View Shows Absence of Left-Sided Aortic Knuckle, Dorsal Spine Dextroscoliosis

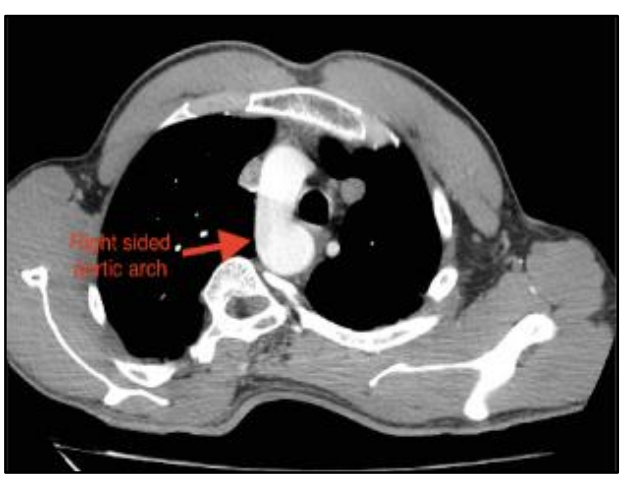

Figure 2. CECT Arterial Phase Axial Section Shows Right-Sided Aortic Arch

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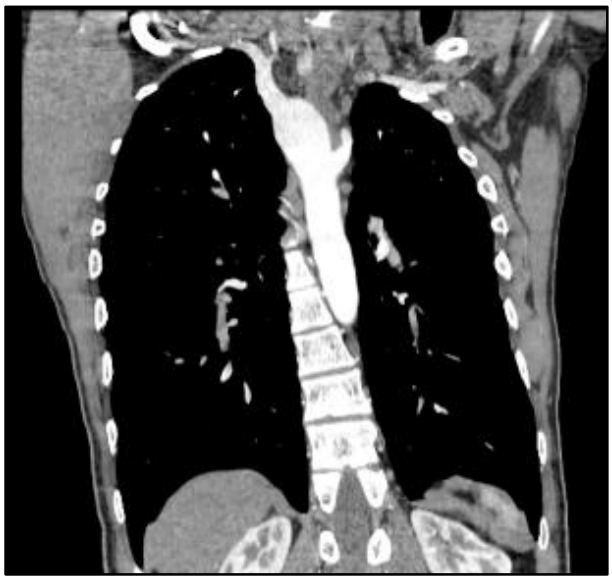


Figure 3. CECT Arterial Phase, Coronal Section Showing Kommerell's Diverticula and Origin of Left Aberrant Subclavian Artery

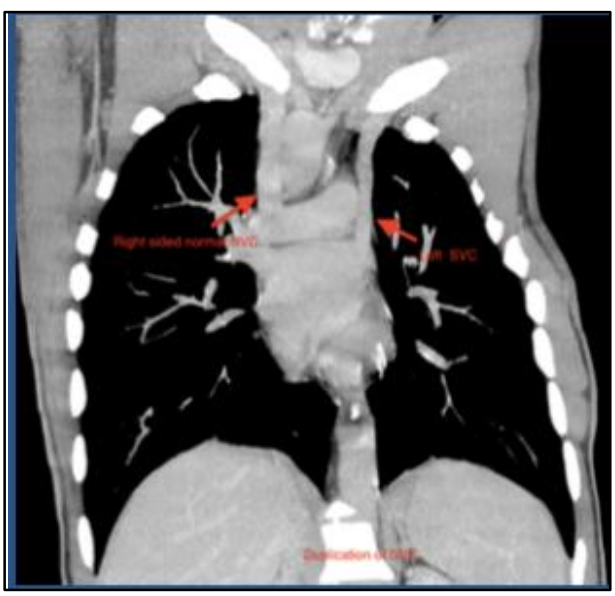


Figure 4. CECT Venous Phase Coronal MIP Image Showing Double SVC- Both are Draining to Right Atrium

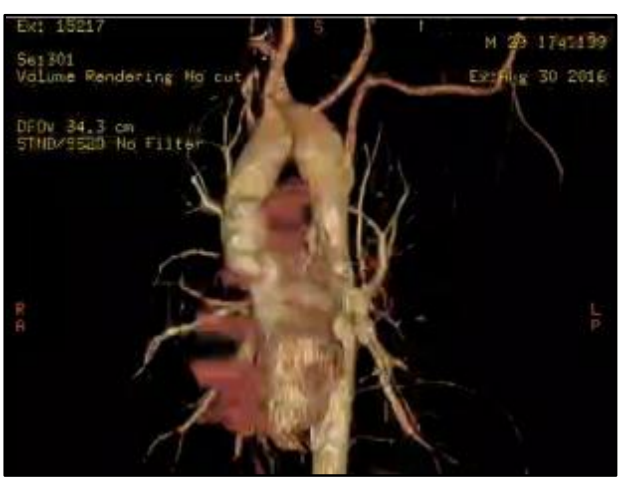


Figure 5. VR Image Showing Right-Sided Aortic Arch and its Branches

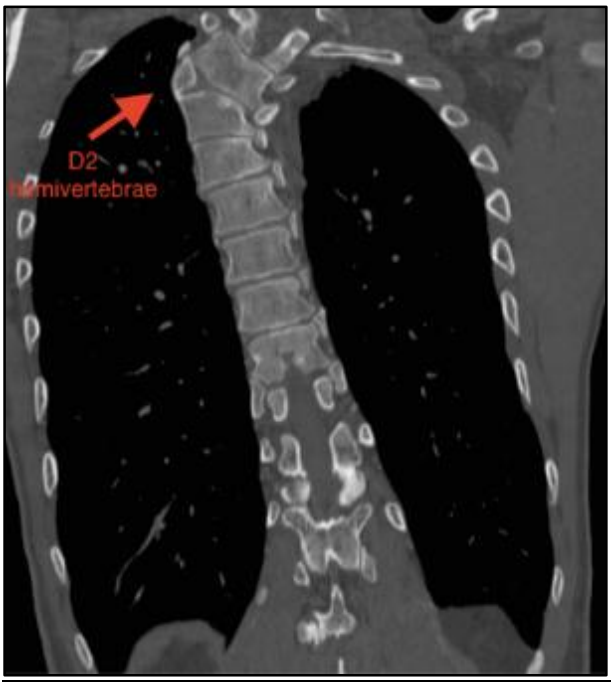


Figure 6. NCCT Bone Window Coronal Image Showing D2 Segmental Hemivertebra

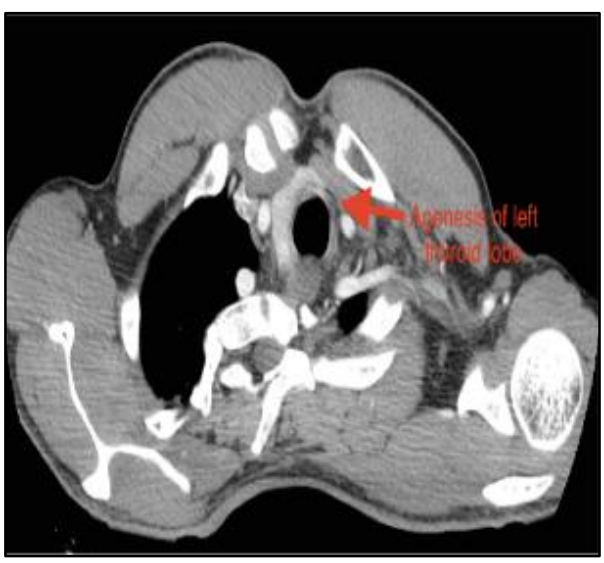
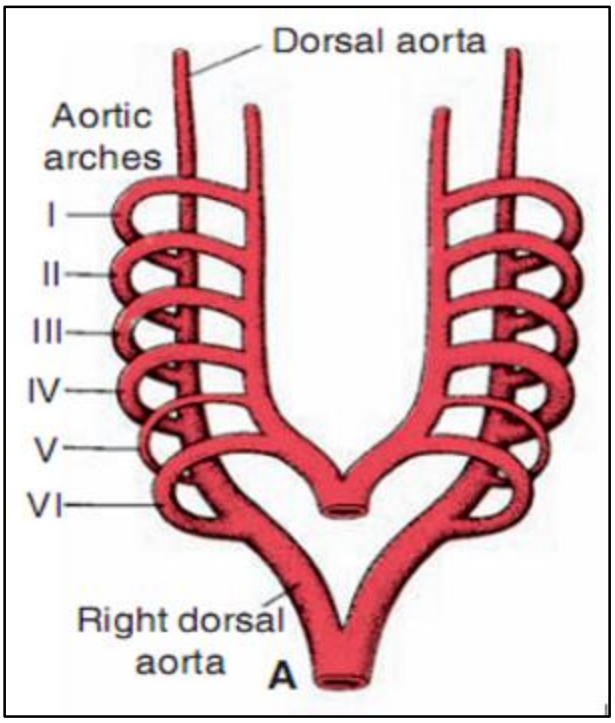


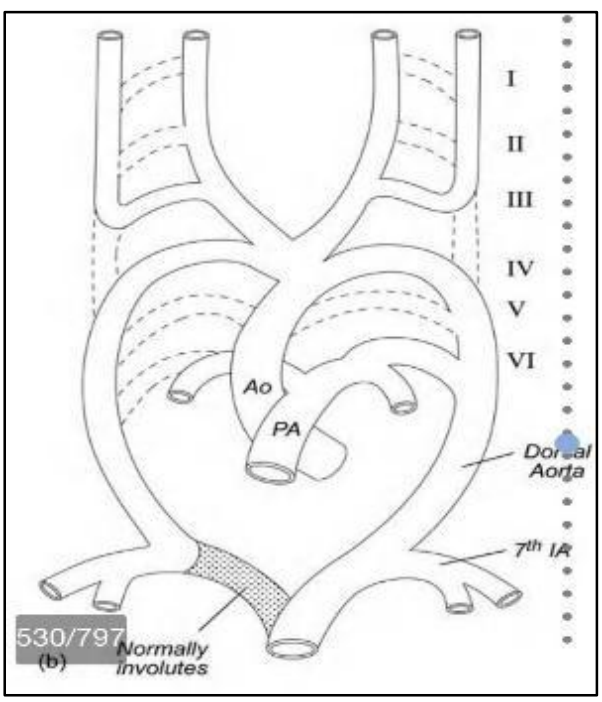
Figure 7. CECT Arterial Phase, Axial Section Showing Right Hemithyroid

EMBRYOLOGICAL DISCUSSION

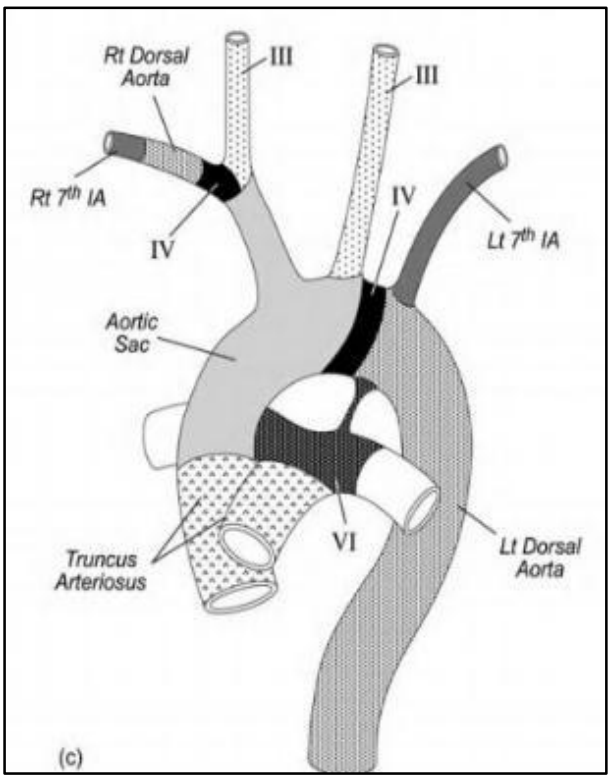
- Six pairs of arterial arches appear connecting the dorsal and ventral aorta.
- All are not present at the same time.
- Selective regression and persistence of these arch vessels forms the major arteries of head, neck and thorax.



The 1st, 2nd, 5th arches disappear.
1st arch- disappeared, small portion → maxillary artery.
2nd arch- disappeared, remain portion → hyoid + stapedial arteries.
5th arch- never forms/forms incompletely → regresses.

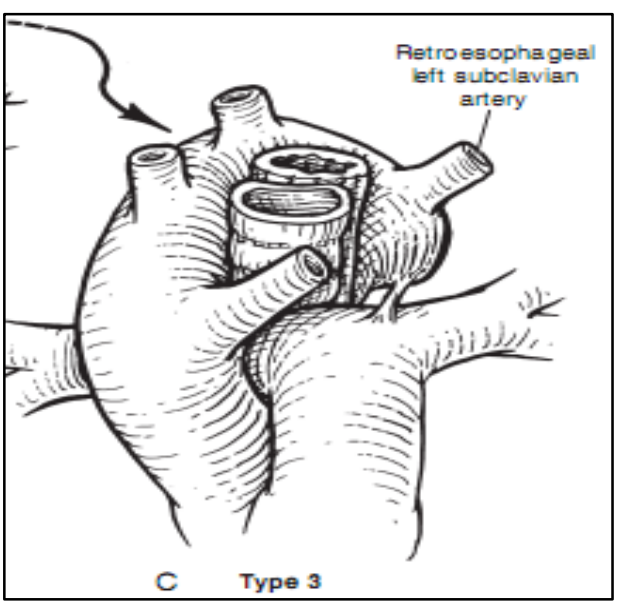


- 3rd arch → Common carotid artery.
- 4th arch-
- Right- Proximal part of right subclavian artery.
- Left- Part of aortic arch between left carotid artery and left subclavian artery.
- 6th arch → Pulmonary arteries and ductus arteriosus.
- Right side-
- Proximal part → Proximal segment of right pulmonary artery.
- Distal portion → Disappears.
- Left side-
- Proximal part → Proximal segment of left pulmonary artery.
- Distal part- Forms ductus arteriosus.



Aortic Arch

+ Proximal segment from aortic sac.
+ Middle segment from the left 4th aortic arch.
+ Distal segment from the left dorsal aortic arch.

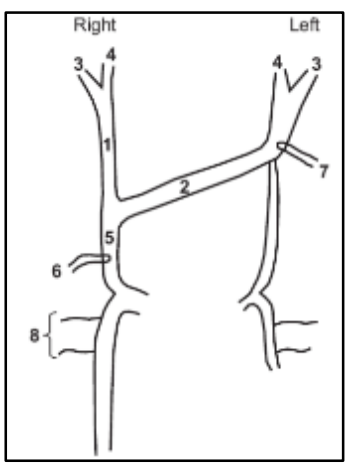


RAA + Aberrant LEFT Subclavian Artery

Right-Sided Aortic Arch with Aberrant Left Subclavian Artery Embryological Aetiology-

- Regression of left aortic arch segment between left common carotid artery and left subclavian artery (4th arch) → Left subclavian artery originates as last branch from the aortic arch → Left subclavian artery pass posterior, left of oesophagus.
- Left ductus ligament originates from bulbous dilation at the base of left subclavian artery (diverticulum of Kommerell) and attaches to proximal left pulmonary artery → the ring compresses oesophagus + trachea.
- Ductus arteriosus can be left/right side.
- Usually, an isolated anomaly.

Duplication of Superior Vena Cava- Results from failure of the embryonic left anterior cardiac vein to regress.



Diagrammatic Representation

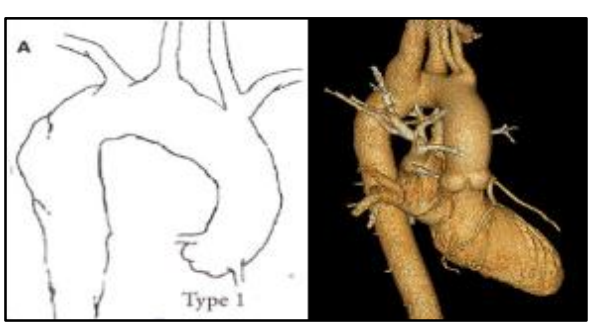
Diagrammatic representation of development of major veins and right internal mammary vein opening into superior vena cava- 1) Right brachiocephalic vein; 2) Left brachiocephalic vein; 3) Subclavian vein; 4) Internal jugular vein; 5) Superior vena cava; 6) Right internal mammary vein; 7) Left internal mammary vein; 8) Intercostal veins.

DISCUSSION

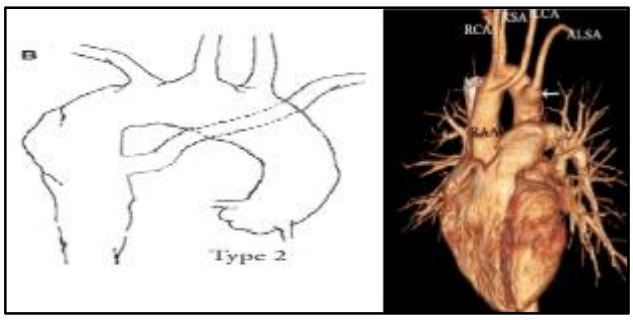
Right-sided aortic arch was first documented by Fioratti and Aglietti in 1763.[5] This has been classified by Edward in 1948, Felson and Palayew in 1963 and Steward et al in 1964.[6]

Edward's Classification[7]

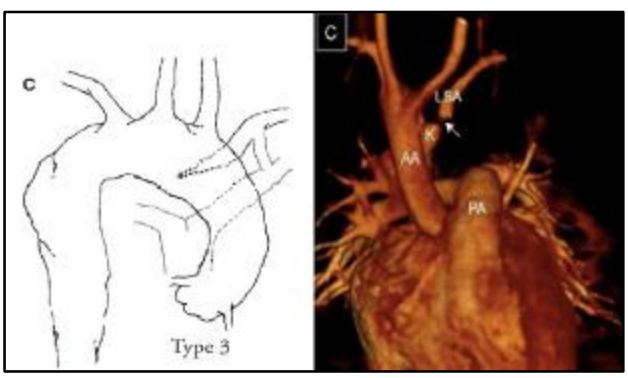
- **Type 1**- The major arteries branching out from the arch are the left innominate artery, the right common carotid and right subclavian artery (mirror image branches).



- **Type 2**- The right-sided arch with the major arteries arising independently (i.e., right subclavian, right common carotid, left common carotid, left aberrant subclavian arteries).



- **Type 3**- The left subclavian artery is isolated and does not attach to the aorta (the left subclavian artery is connected to the pulmonary artery through the ductus arteriosus).



Type 1 and 2 form 98% of the right-sided aortic arch cases and type 3 is very rare.[8] There are approximately 50 cases of right-sided aortic arch with aberrant left subclavian artery in the literature.[3] The anomaly is related to the persistence of the right fourth aortic arch and regression of embryonic left fourth arch between the left common carotid artery and left subclavian artery.[9,10] In this anomaly, as also noted in our case, the first trunk branching from the arch was the right subclavian artery, then right common carotid artery, then left common carotid artery and lastly subclavian artery.[7,11] The branching pattern and radiological features were similarly reported by other authors.[3,4,12,13]

The patient with right-sided aortic arch with aberrant subclavian artery is generally asymptomatic and there is no particular association with cardiac anomalies.[13,14] There might be symptoms related to the presence of the vascular ring. However, more symptoms are due to atherosclerotic changes of the anomalous vessels, dissection and aneurysm with compression of adjacent structures causing dysphagia (dysphagia lusoria) and dyspnoea.[4,9,15,16] In this reported case, there were no symptoms related to the vascular variations and therefore no surgery was indicated.

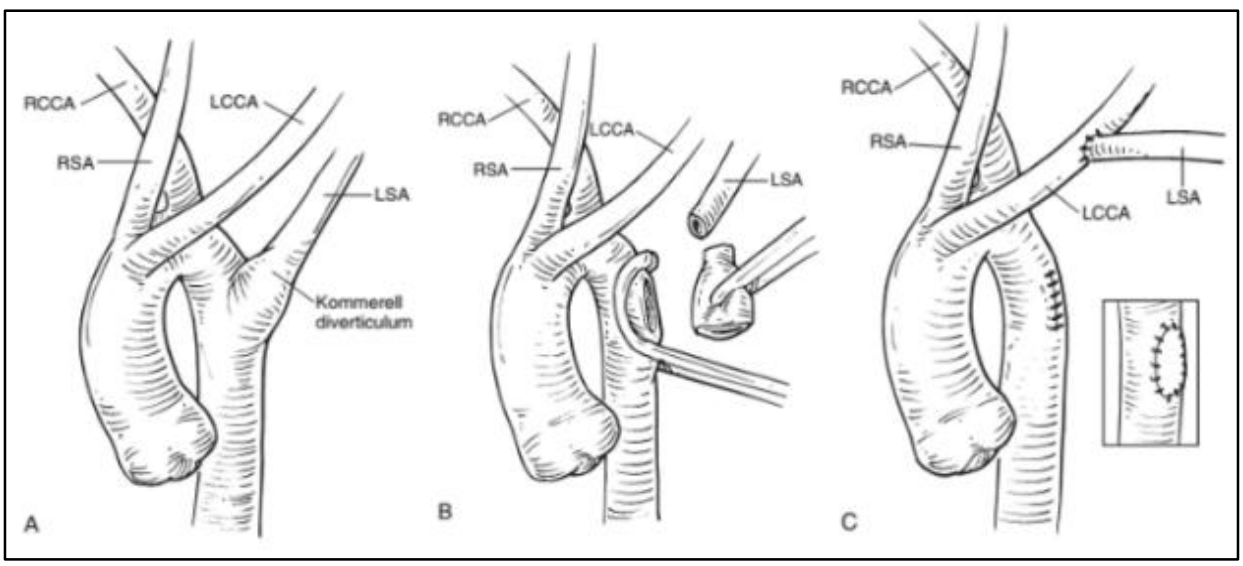
The aberrant left subclavian artery usually originates from a Kommerell's diverticulum. The diverticulum is defined as a conical dilatation of the proximal portion of an aberrant subclavian artery near its origin from the aorta.[4,11] It is also

known as "lusoria diverticulum", "remnant diverticulum" or "lusoria root."[15]

It is the remnant of the left fourth aortic arch in the aberrant left subclavian artery. It was originally described by Burckhard Friedrich Kommerell in 1936[17] in a case of aberrant right subclavian artery (remnant of the right fourth aortic arch) associated with the left aortic arch.[4,12] The location of Kommerell's diverticulum could be behind the oesophagus in 80%, between the trachea and oesophagus in 15% and behind the trachea in 5%.[15]

SURGICAL TREATMENT

The treatment for right-sided aortic arch with aberrant left subclavian artery and Kommerell's diverticulum is excision of the Kommerell's diverticulum and anastomosis of the left subclavian artery to the left common carotid artery.



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