

## Research Article

# Retrospective analytical five months study of aortic arch abnormalities

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### ABSTRACT

**Background:** Aortic arch abnormalities include congenital anomalies and acquired pathologies. Many of aortic arch congenital anomalies are asymptomatic lesions. But some of these anomalies are symptomatic and need to be treated. Radiological appearances should be familiar for these anomalies to the radiologist for correct interpretation. Aortic arch abnormalities reported in our institute are analysed in this article.

**Methods:** This is a retrospective analytical study of five months from July 2015 to October 2015. The study was carried in the radiodiagnosis department of tirunelveli medical college hospital. In our institute, 494 cases of Computerised tomography (CT) Chest scans have been taken in above period for various conditions. In which Patients with vascular abnormalities of aortic arch in the scan were selected for the study i.e. sample selection. No of patients selected for analysis are 20. They have undergone CT scan, Magnetic resonance angiography (MRI) scan. Those findings were analysed and hereby are presented, in which age varied as low as 1 year and as high as 78 years.

**Results:** Most of aortic arch abnormality cases belong to age group 60-80. About 75% of aortic abnormalities are acquired. The aortic arch abnormalities are commoner in males than females especially acquired diseases of aorta. Rare cases of Double aortic arch, aberrant right subclavian artery with dissection of aorta are diagnosed in these patients.

**Conclusions:** It is noted that acquired conditions are the commonest abnormalities. CT scan and MRI complement each other. MR angiogram gives definitive diagnosis in aortic arch abnormalities. Double aortic arch is most common symptomatic vascular ring. MRI is the best single imaging study for the diagnosis and characterization of vascular rings. Aortic dissection needs urgent imaging. Multislice CT, MRI has replaced conventional angiogram.

**Keywords:** Computerised tomography, Magnetic resonance angiography, Aortic arch, Dissection, Angiogram

### INTRODUCTION

Aortic arch abnormalities include congenital anomalies and acquired pathologies. Variant anatomy of the aortic arch occurs when there is failure of normal aortic development. It results in a number of heterogenous anomalies of the aorta and its branch vessels. Radiological investigations are important for diagnosis of these conditions. Many of aortic arch anomalies are asymptomatic lesions. But some of these anomalies are symptomatic and need to be treated. Radiological

appearances should be familiar for these anomalies to the radiologist for correct interpretation. Congenital anomalies of the aortic arch are rare disorders and they may be associated with congenital cardiovascular diseases.<sup>1</sup> Knowledge of aortic abnormalities and variant branching sequence is very important in diagnostic and interventional radiology.<sup>2</sup> Aneurysm of the thoracic aorta is a serious condition as it may be extensive or may be associated with a more distant aneurysm.<sup>3</sup> Aortic arch abnormalities reported in our institute are analysed in this article.

**METHODS**

This is a retrospective analytical study of five months from July 2015 to October 2015. The study was carried in the radiodiagnosis department of tirunelveli medical college hospital. In our institute, 494 cases of CT Chest scans have been taken in above period for various conditions. In which Patients with vascular abnormalities of aortic arch in the scan were selected for the study i.e. sample selection. No of patients selected for analysis are 20.

**Inclusion criteria**

Patients with vascular abnormalities in CT study up to the age of 80.

**Exclusion criteria**

Patients who have not taken MRI scan for confirmation, post-operative patients on follow up, patients with history of chest injury. Contrast study was also done. Those findings were analysed and hereby are presented, in which age varied as low as 1 year and as high as 78 years. CT scan was done with Toshiba single slice helical CT scanner. MRI was done with 1.5 tesla SIEMENS MRI scanner.

**RESULTS**

Radiological findings are important before intervention of aortic arch abnormalities. Radiology plays important role in diagnosing aortic arch abnormalities. In this study all the 20 patients underwent MRI also. The results are analysed as follows.

Most of vascular abnormality cases belong to age group 60-80 i.e. (50%) out of all the cases.

**Table 1: Age wise distribution.**

Age group	No of cases	Percentage
0-10	3	15
11-20	1	5
21-30	2	10
31-40	1	5
41-50	1	5
51-60	2	10
61-70	5	25
71-80	5	25
total	20	100

It is seen in above table that about 75% of aortic abnormalities are acquired.

From table 3, it is seen that aortic arch abnormalities are commoner in males than females especially acquired diseases of aorta.

**Table 2: Imaging findings wise cases distribution.**

Diagnosis	No of cases
Double aortic arch	1
Right arch with mirror image branching	2
Aberrant right subclavian artery	2
Aortic atherosclerosis	12
Aortic aneurysm	1
Aortic dissection	1
Takayasu disease	1
Total	20

**Table 3: Gender wise distribution.**

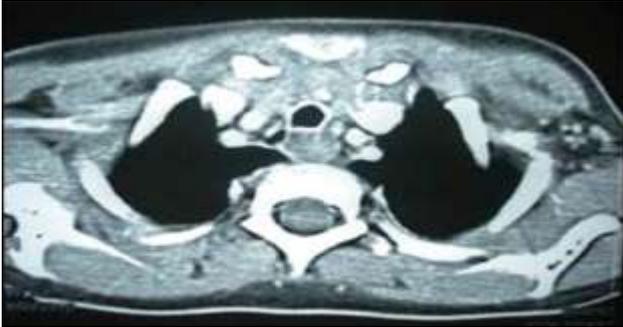
Diagnosis	Male	Female	Total no of cases
Double aortic arch	1	-	1
Right arch with mirror image branching	1	1	2
Aberrant right subclavian artery	1	1	2
Aortic atherosclerosis	8	4	12
Aortic aneurysm	1	-	1
Aortic dissection	-	1	1
Takayasu disease	-	1	1
Total	12	8	20



**Figure 1: Barium swallow AP -indentation of esophagus bilaterally at aortic arch level.**

Rare cases like Double aortic arch, aberrant right subclavian artery with dissection of aorta are diagnosed in this study. Radiologically one child of four years old was diagnosed as double aortic arch when he was imaged for stridor. Four years old male boy had biphasic stridor and recurrent respiratory tract infections evaluated with plain chest radiograph, bronchoscopy and barium studies. Chest x-ray shows prominent right paratracheal soft tissue opacity. Barium swallow AP view shows indentation of esophagus bilaterally at aortic arch level more on right side (Figure 1). Barium swallow lateral view shows posterior indentation of esophagus at aorta level. CT was taken. CT contrast study shows four vessel

sign in superior mediastinum (Figure 2). It also shows double aortic arch (Figure 3). The right arch is larger than left. Both arches encircle the trachea and esophagus causing compression. MRI was taken. T1W sequences confirmed the four vessel sign. MRI contrast angio shows the double aortic arch and its classical branches (Figure 4). CT and MRI helped for arriving the definitive diagnosis of double aortic arch which is the reason for the patient complaints.



**Figure 2: CT contrast -four vessel sign in superior mediastinum.**



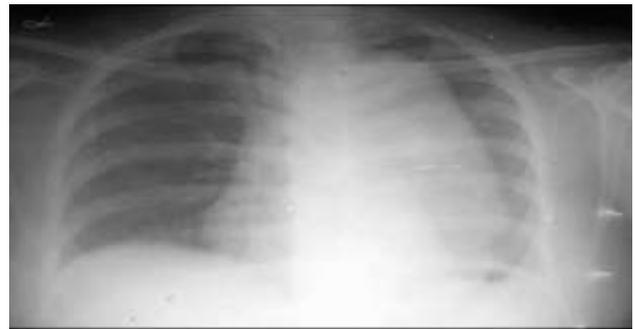
**Figure 3: Both arches encircle the trachea and esophagus causing compression.**



**Figure 4: MRI contrast angio the double aortic arch and its classical branches.**

Another 35 years female patient admitted with chest pain radiating to back, deep abdominal pain and bilateral lower limb pain. Chest x-ray was taken. It shows aneurysmal dilatation of arch and descending thoracic aorta (Figure 5). Chest left lateral view also shows gross dilatation of arch of aorta and descending thoracic aorta. CT scan was taken. Plain CT shows aneurysm of descending thoracic

aorta. Contrast CT shows aberrant right subclavian artery, aneurysm with thrombus and dissection flap (Figure 6). It also shows the true and false lumens of aorta due to dissection. MRI was taken. T1W Sequence shows aneurysm of descending thoracic aorta with thrombus. MR angio with contrast confirms the aberrant right subclavian artery (Figure 7). It also shows that there is dissection of aorta distal to aberrant right subclavian artery and the dissection involves the abdominal aorta extending to left common iliac artery (Figure 8). CT and MRI contrast studies helped for arriving the definitive diagnosis. This case is presented because of rare association of dissection with aberrant right subclavian artery.



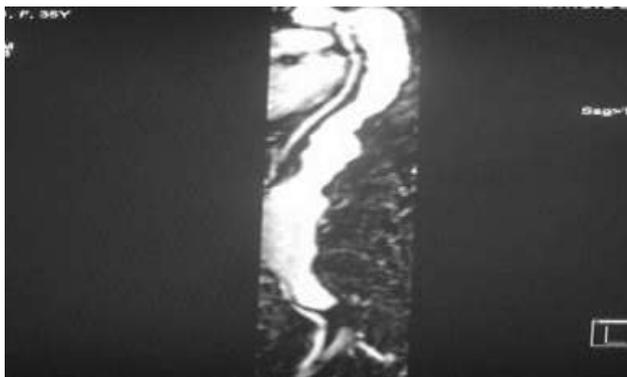
**Figure 5: Chest x-ray-aneurysmal dilatation of arch and descending thoracic aorta.**



**Figure 6: Contrast CT-aberrant right subclavian artery, aneurysm with thrombus and dissection flap.**



**Figure 7: MR angio with contrast confirms the aberrant right subclavian artery.**



**Figure 8: MR angio-dissection of aorta distal to aberrant right subclavian artery, the dissection involves the abdominal aorta extending to left common iliac artery.**

## DISCUSSION

Aortic arch abnormalities include congenital anomalies and acquired pathologies. Radiology plays major role in diagnosis and is important before intervention. Anomalies of the aortic arch can present as a double arch or as a single arch with variations in respect to branching pattern and its course.<sup>4</sup> The most common congenital aortic arch anomaly is left aortic arch with an aberrant (retroesophageal) right subclavian artery.<sup>5</sup> Commonly, failure of normal regression of the 4th arch vessels results in a double aortic arch or right sided aortic arch. Other arch anomalies include hypoplastic ascending aorta, patent ductus arteriosus, ductus diverticulum, and coarctation of the aorta, interrupted aortic arch and cervical aortic arch.

Double aortic arch is the most common form of symptomatic vascular ring. The trachea and esophagus are completely encircled by connected segments of the aortic arch and its branches. Various forms of double aortic arch exist, the common defining feature- both the left and right aortic arches are present. Vascular rings - formed when the process of regression and persistence does not occur normally, the resulting vascular anatomy completely encircles the trachea and esophagus. A double aortic arch is formed when both fourth arches and both dorsal aortas remain patent. In double aortic arch, vascular ring is formed by the splitting of the ascending aorta into two limbs that pass to either side of the trachea and oesophagus.<sup>6</sup> classification based upon Edwards' hypothetical double aortic arch system is as follows; Types A, B and C double aortic arches with interruption of left arch respectively distal to P.D.A., proximal to P.D.A. and proximal to left subclavian artery. Type D double aortic arch-double aortic arch with interruption of its left component proximal to the site of origin of left common carotid artery.

>75% of patients, the right arch are dominant. In 20% of patients, the left arch is dominant. In these patients, the minor right arch typically is patent. In general, the apex

of the right-sided arch is more superior to the left arch, and on occasion, a cervical arch may be present on either side. Vascular rings encircle the trachea and esophagus, usually causing compression of both structures. Compression of the trachea upper airway obstruction. Esophageal compression swallowing difficulties-manifest as vomiting and feeding intolerance in infants and younger children and as dysphagia later in life. Right arch supplies Right common carotid and Right subclavian arteries. Left arch supplies left common carotid and left subclavian arteries. This branching pattern produces the four vessel sign. If the minor arch is markedly atretic or hypoplastic, the affected segment tends to be always distal to the left subclavian artery.<sup>7</sup> Right arch is higher, left arch is lower producing reverse on esophagram in AP. MRI is the best imaging study for the diagnosis and characterization of vascular rings.it gives complete information regarding the arterial branching pattern and

Clearly demonstrates the locations and extent of airway and esophageal obstruction, it can be used to delineate cardiac anatomy. It is useful tool for preoperative planning. Aortic dissection is defined as an event that results in the separation of the layers of the media by blood, producing a false channel with variable proximal and distal extension. There are two types of classification. They are DeBakey classification and Stanford classification.

DeBakey classification:

Type I: The entire aorta is involved.

Type II: Only the ascending aorta is involved.

Type III: Only the descending aorta is involved.

Type IIIA involves the descending aorta as far as the diaphragm.

Type IIIB involves the descending aorta below the diaphragm.

Stanford classification:

Type A: The ascending aorta.

Type B: The descending aorta is involved.

Approximately 60% of dissections involve the ascending aorta.<sup>8</sup> Chest radiography is abnormal in 90% of patients. Abnormal aortic contour and widened mediastinum are the most common findings. A normal Chest x-ray does not rule out aortic dissection. Ring sign (displacement of the aorta >5 mm past the calcified aortic intima) is a specific radiographic sign. CT scan gives imaging information, including the type of lesion, location of the pathologic lesion, extent of the disease, and evaluation of the true and false lumen assessed quickly and helps the surgeon plan the operation. It has the advantage of short acquisition time. CT findings are Separation of calcifications from the outer aortic contour. Diagnostic hallmark is the intimal flap. The false lumen is usually anterior and to the right in the ascending aorta, superior and slightly posterior along the arch, and posterior and to the left in the descending aorta. MRI scan is multiplanar imaging, shows the site of intimal tear, type and extent of

dissection, and presence of aortic insufficiency, the surrounding mediastinal structures. It is preferred tool for imaging chronic dissections and postsurgical follow-up. Contrast MRA is an accurate non-invasive imaging modality which also evaluates the aortic valve more effectively. In patients with impaired renal function, Rapid non-contrast imaging techniques (e.g. true FISP) may see MRI having a larger role to play in the acute diagnosis.<sup>9</sup>

Aberrant right subclavian artery is the most common aortic arch anomaly, occurring in 1 in 200 patients. It is mostly posterior to oesophagus. It is almost asymptomatic. Dysphagia, stridor, GI bleeding, aneurysm are rare complications. Dysphagia is termed as dysphagia lusoria in this condition.<sup>10</sup> Rarely aberrant right subclavian artery takes a course anterior to the trachea or esophagus.<sup>11</sup>

## CONCLUSION

From this study, it is noted that most of aortic arch abnormality cases belong to age group 60-80. About 75% of aortic abnormalities are acquired. The aortic arch abnormalities are commoner in males than females especially acquired diseases of aorta. CT scan and MRI complement each other in diagnosing aortic arch abnormalities. Acquired conditions are commoner than congenital anomalies. MR angiogram gives definitive diagnosis in aortic arch abnormalities. Double aortic arch is most common symptomatic vascular ring. Right arch supplies Right common carotid and Right subclavian arteries. Left arch supplies Left common carotid and Left subclavian arteries. Aortic dissection needs urgent imaging. Site, extent; vital organ involvement should be assessed. True, false lumen characterisation should be done. Multislice CT, MRI has replaced conventional angiogram.

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## REFERENCES

1. Türkvtan A, Büyükbayraktar FG, Ölçer T, Cumhuri T. Congenital Anomalies of the Aortic Arch:

- Evaluation with the Use of Multidetector Computed Tomography-Korean J Radiol. 2009;10(2):176-84.
2. Kau T, Sinzig M, Gasser J, Lesnik G, Rabitsch E, Cedin S, et al. Univ.-Doz.- Aortic Development and Anomalies-Semin Intervent Radiol. 2007;24(2):141-52.
3. Crawford ES, Stowe CL, Crawford JL. Aortic arch aneurysm. A sentinel of extensive aortic disease requiring subtotal and total aortic replacement.- Ann Surg. 1984;199(6):742-52.
4. Goor DA. Congenital malformations of the heart. Embryology, anatomy and operative considerations. New York: Grune & Stratton, 1975.
5. Ramos-Duran L, Nance JW, Henzler T. Developmental Aortic Arch Anomalies in Infants and Children Assessed With CT Angiography. AJR. 2012;198:W466-74.
6. Noguchi K, Hori D, Nomura Y. Double aortic arch in an adult. Interact Cardiovasc Thorac Surg. 2012;14(6):900-2.
7. Ramos-Duran L, Nance JW, Schoepf UJ. Developmental aortic arch anomalies in infants and children assessed with CT angiography. AJR Am J Roentgenol. 2012;198(5):W466-74.
8. Macura KJ, Corl FM, Fishman EK. Pathogenesis in acute aortic syndromes: aortic dissection, intramural hematoma, and penetrating atherosclerotic aortic ulcer. AJR Am J Roentgenol. 2003;181(2):309-16.
9. Pereles FS, McCarthy RM, Baskaran V. Thoracic aortic dissection and aneurysm: evaluation with nonenhanced true FISP MR angiography in less than 4 minutes. Radiology. 2002;223(1):270-4.
10. Ka-tak W, Lam WW, Yu SC. MDCT of an aberrant right subclavian artery and of bilateral vertebral arteries with anomalous origins. AJR Am J Roentgenol. 2007;188(3):W274-5.
11. Epstein DA. Abnormalities Associated with Aberrant Right Subclavian Arteries: A case Report, vasc endovascular surg. 2002;36(4):297-303.

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