

# Right aortic arch with isolation of the left innominate artery in a case of double chamber right ventricle and ventricular septal defect

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## ABSTRACT

Herein, we report an unusual case of right aortic arch with isolation of the left innominate artery in a case of double chamber right ventricle with ventricular septal defect. The blood supply to the innominate artery was by a collateral arising from the descending aorta. The embryological development of this anomaly can be explained by the hypothetical double aortic arch model proposed by Edwards with interruption of the arch at two levels.

**Keywords:** Right Aortic arch, double interruption of embryonic aorta, isolation of left innominate artery

## INTRODUCTION

Right-sided aortic arch with isolation of the innominate artery is a very rare congenital anomaly, in which the innominate artery loses its connection with the ascending aorta and is supplied by either a patent ductus or mediastinal collaterals. The origin of this anomaly can be explained by the hypothetical double aortic arch model proposed by Edwards with interruption of the arch occurring at two levels. Herein, we present a case where this anomaly was present in a child with double chamber right ventricle and ventricular septal defect.

## CASE REPORT

A 10-year-old male child presented to us with chief complaints of shortness of breath while playing. There was no history of cyanosis or difficulty in swallowing. There was no history suggestive of cerebral ischemia or vertebral steal. On clinical examination, pulse was slightly weaker on the left radial artery and with a delay compared to right radial pulse. Blood pressure in the left upper arm was 85/58 mmHg while in the right upper

arm it was 98/60 mmHg. Auscultation of bilateral lung fields revealed absence of wheeze or abnormal sounds. Echocardiography showed ventricular septal defect (VSD), double chamber right ventricle (DCRV), right aortic arch (RAA) with normal sized pulmonary arteries and normal pulmonary valve. There was no evidence of co-arcuation.

Patient also underwent cardiac catheterization [Figure 1a and b] and multi-detector computed tomography (MDCT) scan [Figure 1c-e] with virtual bronchoscopy to look for tracheal compression and barium esophagogram. Investigations revealed right common carotid and right subclavian arteries originated separately from the ascending right aortic arch with right vertebral artery branching from right subclavian artery. Right vertebral artery was larger in size compared with left vertebral artery. There was no evidence of tracheal compression.

Left innominate artery (LIA) did not have any direct connection with the aortic arch and was filled with a delay of contrast through a tortuous vessel opening in left subclavian artery (LSCA). The tortuous vessel originated from upper descending aorta, travelled in the posterior mediastinum, came out of the thoracic outlet and joined the 1<sup>st</sup> part of LSCA. During the course it gave off first few intercostal arteries on the left side. The left vertebral artery was arising from 1<sup>st</sup> part of subclavian artery and was smaller compared to the right vertebral artery.

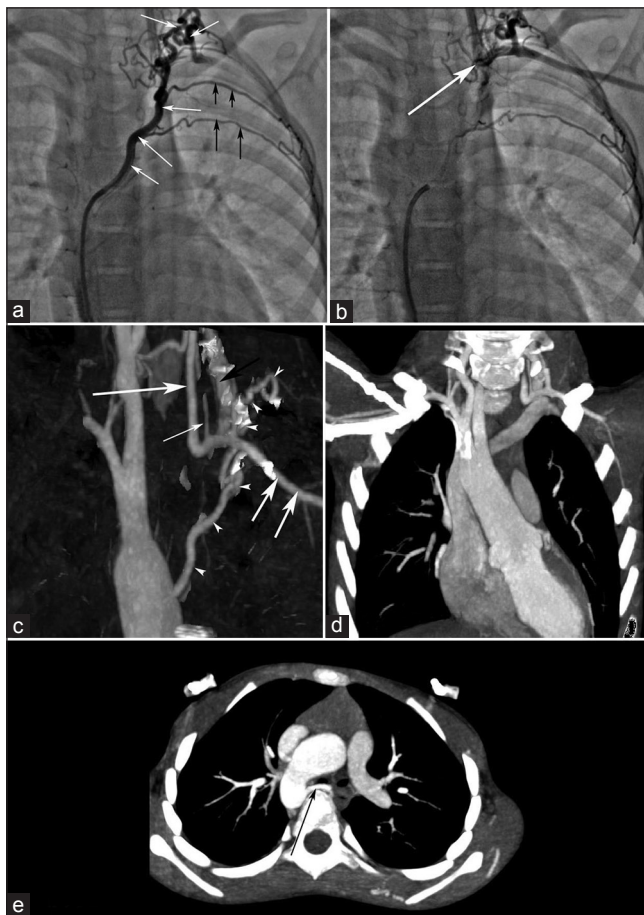
Magnetic resonance (MR) brain revealed normal study, while MR angiogram [Figure 2] described a reduced size

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of left carotid and vertebral system in cervical region [Figure 2a] along with absence of left internal carotid artery at the level of circle of Willis [Figure 2b]. Left anterior cerebral artery received supply from right internal carotid artery through anterior communicating artery. There was intense contrast opacification from the right sided carotid-vertebral system filling up the whole of the circle of Willis with very little contribution from left carotid and vertebral artery.

Patient underwent surgery for VSD closure and infundibular band resection with preservation of pulmonary valve under moderately hypothermic cardiopulmonary bypass. The ligamentum arteriosum was identified to be attached to the left pulmonary artery traced cephalad into the depths of mediastinum. Postoperative stay was uneventful and the patient was discharged 8 days after surgery.



**Figure 1:** (a and b) Shows pictures of selective angiogram during cardiac catheterization. (a) Multiple white arrows show tortuous course of longitudinal artery, multiple black arrows show intercostal arteries given off by longitudinal artery. (b) Single large white arrow shows 'V' junction of the LSCA and LCCA; (c and d) Show images of CT angiography. (c) Large white arrow shows LCCA, while small white arrow shows left vertebral artery. A black arrow shows left innominate vein. (d) Shows separate origin of RCCA and RSCA from the RAA. (e) Retro-esophageal course of the longitudinal artery

## DISCUSSION

D'Cruz<sup>[1]</sup> and associates and Levine *et al.*, in 1966 reported the first cases of RAA and isolation of the LIA from the aorta. Later very few similar cases are reported, in which vertebro-basilar insufficiency was observed by few authors,<sup>[2,3]</sup> while others did not.<sup>[4]</sup> Re-implantation of innominate to the aorta is an attractive option whenever feasible.<sup>[5]</sup>

RAA occurs when a segment of the left arch of the primitive double aortic arch undergoes complete absorption at an early stage of embryologic development.

The interruption of the left arch may occur in 4 different zones, resulting in type 1, 2, 3 and 4 RAA anomalies. The concept of 'hypothetical double aortic arch plan' was first given by Edwards in 1948.<sup>[6]</sup> This hypothetical double arch model is very useful in understanding these malformations [Figure 3].

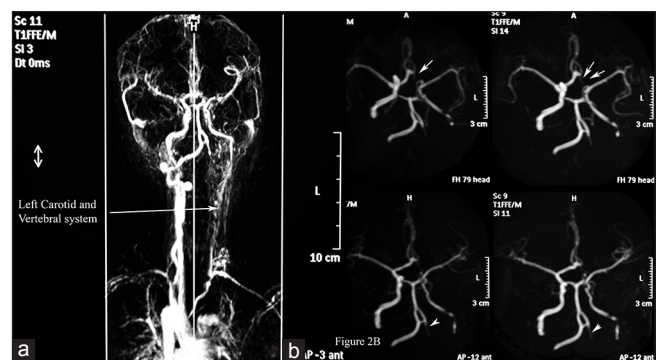
Type 1 RAA without retro-esophageal segment results from interruption of the embryonic left arch distal to the ductus arteriosus, in which the anterior ligamentum courses from the brachiocephalic artery to the proximal left pulmonary artery creating a mirror image branching pattern.

Type 2 RAA — Mirror-image branching and retroesophageal ligamentum arteriosum results from interruption of the left arch proximal (upstream) to the ductus arteriosus with the left-sided ligament arising from Kommerell's diverticulum.

Type 3 RAA — Retroesophageal left subclavian artery and ligamentum arteriosum results from interruption of the left arch between the LSCA and left common carotid artery (LCCA).

Type 4 RAA — Retroesophageal left brachiocephalic artery results from interruption between the left common carotid and the right arch.

A portion of the left arch is transformed into a LIA in Type 1, 2 and 4 RAA [Figure 2]. The location of the innominate



**Figure 2:** MR angiogram of neck and brain perfusion pattern (a) shows markedly hypoplastic carotid and vertebral arterial system; (b) Deficient left internal carotid artery at the level of circle of Willis

artery in relation to the other branches of the right arch varies according to the site of interruption of the left arch during an early stage of embryologic development.<sup>[7]</sup> In Type 3 RAA, a left innominate artery is not formed.

In our case, there was neither LIA nor Kommerell's diverticulum. According to Edwards model, our case had 2 levels of interruption, one just distal to the ligament, another proximal to the origin of LCCA separating both LCCA and LSCA from main aortic artery, with left arch transforming into LIA.

**Fate of the inter-segmental arteries**

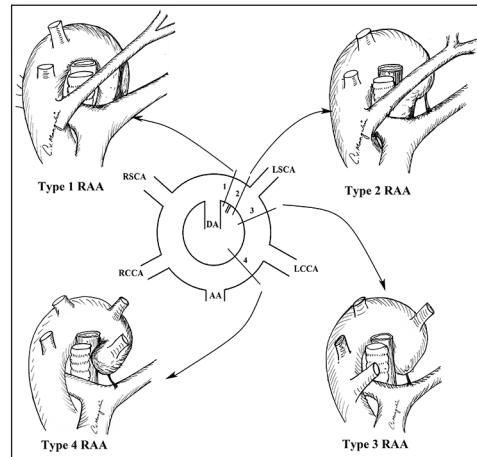
At the end of the third gestational week, inter-segmental arteries arise from the postero-lateral surface of the descending aorta and vascularize the somite derivatives. In the cervical region, longitudinal anastomoses connect the neighboring inter-segmental arteries. By the end of the 7<sup>th</sup> week, the cervical inter-segmental arteries are obliterated, with the exception of the 7<sup>th</sup> inter-segmental artery, which gives rise to the subclavian artery. The longitudinal anastomoses of the inter-segmental artery give rise to the deep cervical and vertebral artery.<sup>[7]</sup>

In the initial embryonic stages, the heart develops in cervical region. As the cervical region elongates, the heart gradually migrates into the thoracic cavity. This longitudinal anastomosis of the inter-segmental arteries also continues in the thoracic region creating an artery parallel to each dorsal aorta [Figure 4]. The artery also remains well connected with the 7<sup>th</sup> inter-segmental artery, which correlates well with the origin of both vertebral and internal mammary arteries from the first part of subclavian artery and also gives off first few superior inter-costal arteries [Figure 2e-Multiple black arrows].

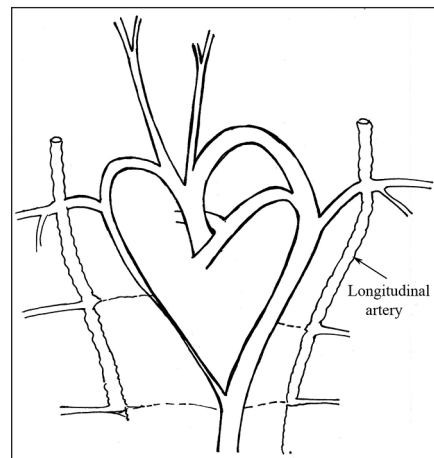
The ductus may be present to connect to the innominate artery with flows from the innominate artery to the pulmonary artery due to high systemic vascular resistance. However, the ductus may close off in later life and the innominate artery may get collateralized from vertebral or intercostal collaterals.<sup>[8]</sup> Usually the collaterals are multiple, very rarely a single large mediastinal 'collateral artery' is found as in our case as described by Gamillscheg et al.<sup>[9]</sup> This can best be surmised to be the longitudinal anastomosis of thoracic inter-segmental arteries.

Many authors recommend vascularizing LIA whenever possible to treat steal and cerebral ischemia.<sup>[5,10]</sup> We electively chose to allow the child's left upper limb to be perfused by the natural collateral artery. The innominate artery was far away to allow mobilization and re-implantation required a synthetic graft with the prospect of graft out-growth, high re-occlusion rate and need for anti-coagulant. We did not anticipate subclavian steal or carotid steal because we felt that the majority

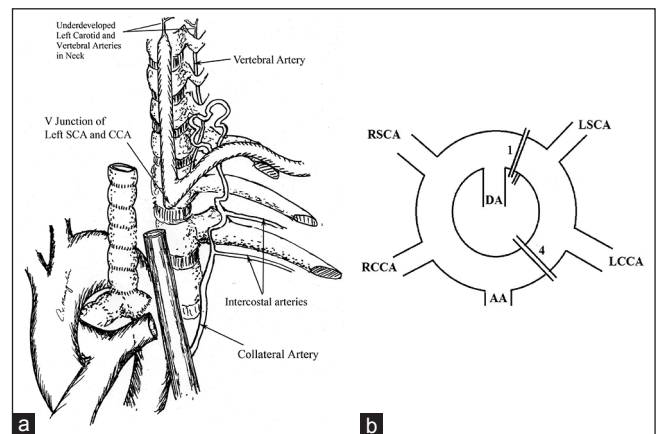
of brain was supplied by right carotid artery in the face of a grossly underdeveloped left carotid and vertebral system [Figure 5].



**Figure 3: Various types of RAA with Edward's model of DAA in center. Various interruption levels are well correlated with the types of RAA**



**Figure 4: Longitudinal artery as seen while embryonic development**



**Figure 5: (a) Illustration of the anomaly in this case. (b) Correlation with Edward's model shows double interruption i.e. at type 1 level and at type 4 level**

## SUMMARY

To summarize, our case is diagnosed as double chambered right ventricle with ventricular septal defect, right aortic arch (double interruption - type 1 and type 4), isolation of left innominate artery with persistent collateral artery feeding the left upper arm and left carotid-vertebral system hypoplasia. The root of this anomaly lies in the embryological development of aortic arches, major vessels, cervical and thoracic inter-segmental arteries. Edward's model of double aortic arch for classifying right aortic arch is extremely valuable in understanding such anomalies. Decision of revascularization should be taken considering target vascular tree anatomy and potential benefit.

## REFERENCES

- Hastreiter AR, D'Cruz IA, Cantez T, Namin EP, Licata R. Right-sided aorta. I. Occurrence of right aortic arch in various types of congenital heart disease. II. Right aortic arch, right descending aorta, and associated anomalies. *Br Heart J* 1966;28:722-39.
- Garti IJ, Aygen MM. Right aortic arch and isolation of the left innominate artery from the aorta. *Cardiovasc Intervent Radiol* 1982;5:235-7.
- Levine S, Serfas LS, Rusinko A. Right aortic arch with subclavian steal syndrome (atresia of left common carotid and left subclavian arteries). *Am J Surg* 1966;111:632-7.
- Martin EC, Mesko ZG, Griep RB, Haller JO, Gordon DH. Isolation of the left innominate artery, a right arch, and a left patent ductus arteriosus. *AJR Am J Roentgenol* 1979;132:833-5.
- Lee C, Kim TS, Lee CH, Kim SJ. Incomplete isolation of the left innominate artery in a patient with right aortic arch and double outlet right ventricle. *J Card Surg* 2010;25:232-4.
- Edwards JE. Anomalies of the derivatives of the aortic arch system. *Med Clin North Am* 1948;32:925-49.
- Garti IJ, Aygen MM, Vidne B, Levy MJ. Right aortic arch with mirror-image branching causing vascular ring. A new classification of the right aortic arch patterns. *Br J Radiol* 1973;46:115-9.
- Park MK. Right aortic arch with isolation of left innominate artery. *Chest* 1979;76:106-8.
- Gamillscheg A, Stein JI, Beitzke A. Ventricular and atrial septal defects, and right aortic arch associated with isolation of the left innominate artery from the aorta. *Heart* 2000;83:99-100.
- Gil-Jaurena JM, Ferreiros M, Zabala I, Cuenca V. Right aortic arch with isolation of the left innominate artery arising from the pulmonary artery and atrial septal defect. *Ann Thorac Surg* 2011;91:303.

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