Case Report

DOUBLE AORTIC ARCH: AN UNCOMMON CAUSE OF STRIDOR IN AN INFANT

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A male infant six months of age presented with stridor. Echo, MRI, cardiac catheterization confirmed the diagnosis of double aortic arch. The child underwent successful surgery. The case is reported as an uncommon cardiac anomaly and an uncommon cause of stridor in an infant.

KEYWORDS: Stridor, Double Aortic Arch, Subclavian Artery, Thoracotomy, Ligamentum Arteriosum, Aortopexy, Stenosis, Trachea

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ETIOLOGY:

The classical double aortic arch anatomy develops when involution of the distal right fourth arch does not take place. The fourth right and left arches both persist and form the left sided descending thoracic aorta. The right and left arches encircle the trachea and oesophagus. The double aortic arch then forms a ring around the trachea and oesophagus. The defect is rarely associated with other intra cardiac defect. Factors responsible for aberrant persistence of double arch segment have not been clearly identified. Band 22q11 deletion was found in 3 of 22 patients (14%)

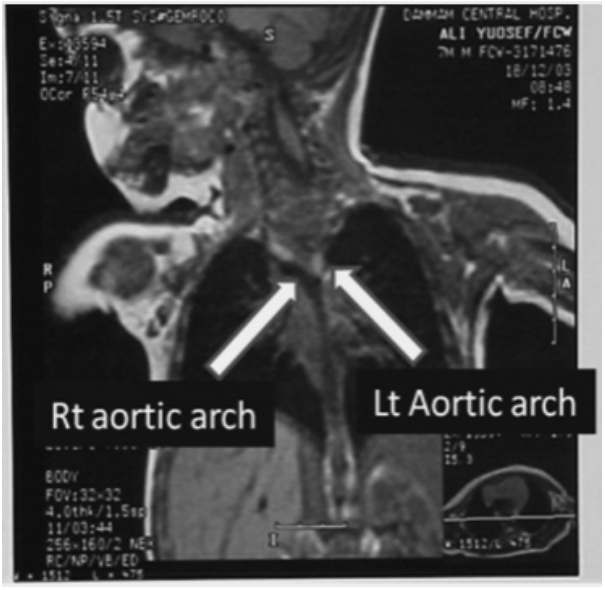

Fig. 1 MRI showing larger Rt. Aortic and smaller Lt. Aortic Arch

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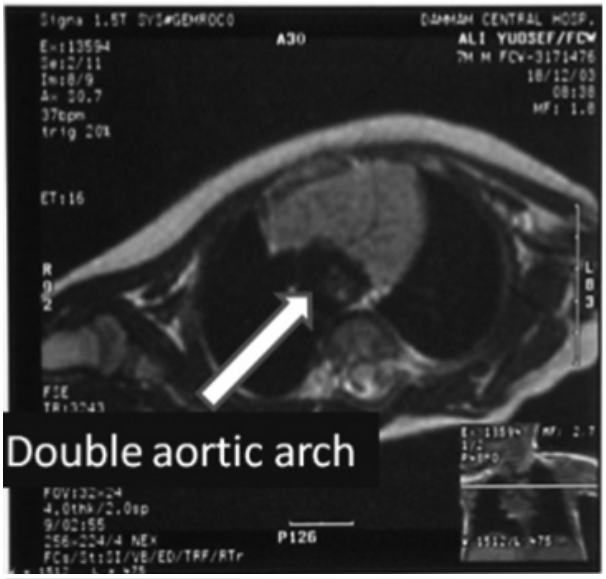

Fig. 2 MRI showing Double Aortic Arch forming a complete circle

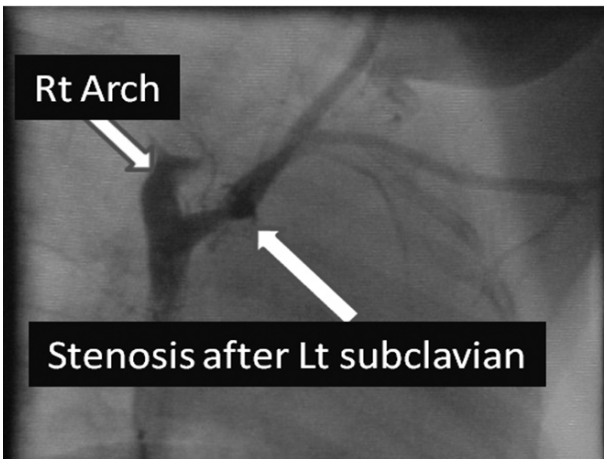

Fig. 3. Angiogram showing stenosis in Lt. Arch distal to origin of Lt. Subclavian Artery

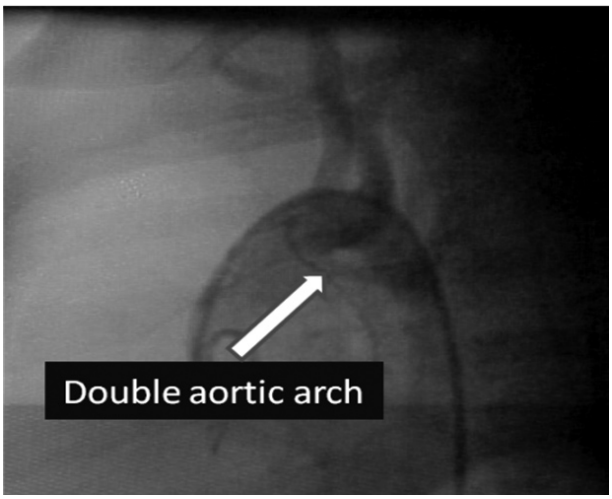

Fig. 4. Angiogram showing small Lt. Arch in lateral view

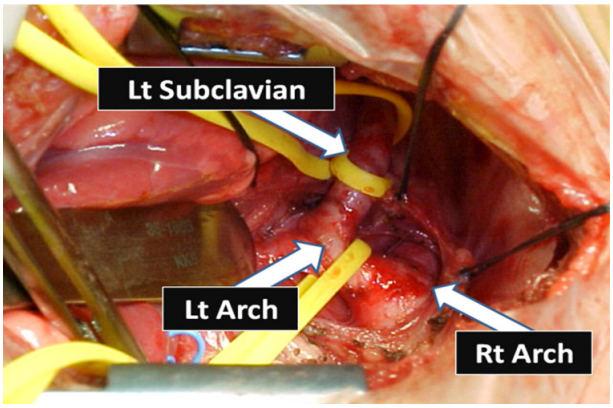

Fig. 5. Operation picture showing Lt. Subclavian Artery, Lt Aortic Arch and Rt. Aortic Arch

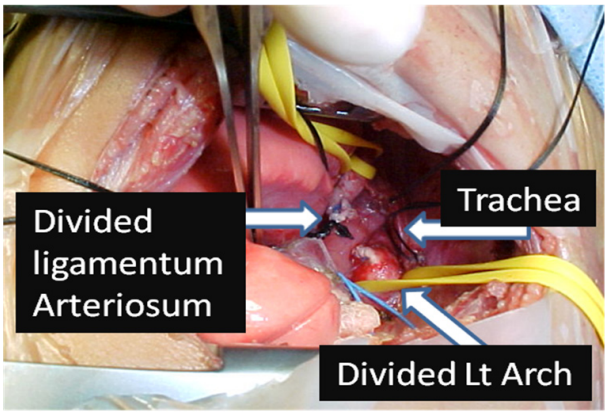

Fig. 6. Operative showing the division of Ligamentum Arteriosum and the Lt Aortic Arch

in double aortic arch[1].

ANATOMY:

In a double aortic arch both arches may be patent or an atretic segment may be present at one of the segments located in either arch. When both arches are patent one may be larger than the other or both may be similar in size.

In 75% of patients with double aortic arch the right arch is larger than the left.

Regarding associated cardiac anomalies, one fifth have other congenital cardiac defect; tetralogy of fallot, ventricular septal defect, coarctation of aorta[2], truncus areteriosus, patent ductus arteriosus and transposition of the great arteries[3].

CASE REPORT:

6 month old boy referred to us with stridor. Echo cardiography suggested double aortic arch. MRI (Fig. 1 & 2) and cardiac catheterization (Fig. 3 & 4) confirmed the diagnosis. In cardiac catheterization, there was stenosis distal to the origin of the left subclavian artery.

TREATMENT:

Under general anaesthesia, central line and bilateral radial arterial line were placed. Left lateral thoracotomy was done the Lt Aortic Arch, Rt. Aortic Arch and Lt. Subclavian Artery were identified (Fig. 5); the ligamentum arteriosum was divided after being ligation. The left arch was divided distal to the origin of the left subclavian artery and after minimal mobilization the trachea could be seen (Fig. 6). Chest drain was placed. In ICU post extubation the stridor disappeared completely and the child was discharged home four days later.

The risk of the procedure is extremely low[4]. Outcome is usually good. However in some patients obstruction of the airway persists. Symptoms may improve after aortopexy is performed[5]. Robotic assistance division has been performed[6], as well as video assisted thoracoscopic vascular ring division[7].

DISCUSSION:

The treatment of double aortic arch is undertaken by division of the non dominant arch after it is established as the cause of compression of the trachea. In our case the cardiac catheterization had shown stenosis distal to the origin of the left subclavian artery therefore the non dominant left arch was divided distal to the origin of the left subclavian to ensure that there is no compromise in blood flow. Thus the site of division is decided according to the anatomical variation as dominance of arch, stenosis or atretic segment ensur-

ing the blood flow to the head and neck vessels remains normal.

CONFLICT OF INTEREST:

The authors have no conflict of interest to declare.

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