Case Report – Persistent Fifth Arch

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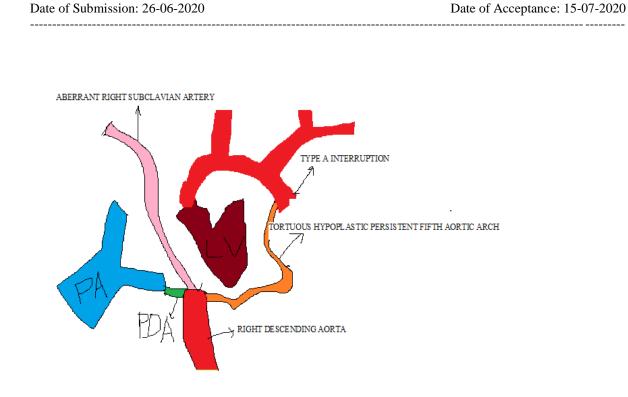

Abstract
INTRODUCTION-We report a 4 and 1/2 month male childwho had Nonrestrictiveperimembranous VSD Left to Right shunt, with Tortuous and Hypoplastic arch with Retroesophageal Right Subclavian artery originating from Right Descending Aorta.CT angiography was done which revealed similar findings.PDA was connecting from right descending aorta to RPA.
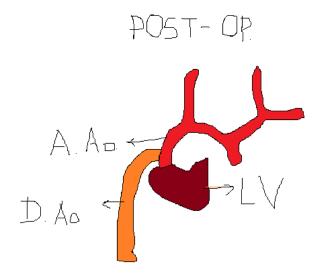*MATERIALS & METHODS-The the patient was taken for cardiac catheterisation.*
Catheter course was from Right Femoral artery to Descending Aorta to Transverse arch to Hypoplastic Tortuous arch to Ascending Aorta to LV.Significant gradient of 37mmHg was found between ascending and descending aorta.Hence patient was taken for surgery.VSD was closed with preformed oblong dacron patch with interrupted 5 -0 prolenepledgetted sutures along the posteroinferior margin &continous 5-0 sutures for rest of the margins, so as to dedicate the aorta across the VSD to the LV.Descending Aorta was mobilised so that it could be approximated to the ascending Aorta without tension and then it was anastomosed side to side to Ascending Aorta.
DISCUSSION-In type A interrupted left aortic arch, the arch interruption occurs distal to the origin of the left subclavian artery. In any of the 3 types, the right subclavian artery may arise normally or abnormally; the 2 most common abnormal sites are distal to the left subclavian artery (aberrant right subclavian artery) and from a right ductus arteriosus (isolated right subclavian artery).Thus in our case it was Type-I interruption with Right descending Aorta.Persistent fifth aortic arch (PFAA) has been subcategorised into three subtypes: (I) double-lumen aortic arch with both lamina patent; (II) atresia and interruption of the superior arch with patent inferior (persistent fifth) arch; and (III) systemic-to-pulmonary arterial connection arising proximal to the first brachiocephalic artery.
CONCLUSION-In our case it was PFAA with (II) and (III) subtypes i.e. there was interruption of the superior arch with patent inferior (persistent fifth) arch joining Ascending and Descending Aorta; and (III) systemic-to-pulmonary arterial connection was established by PDA to RPA.

I. Case Report-

TYPE A INTERRUPTION WITH ABERRANT RIGHT SUBCLAVIAN ARTERY WITH TORTUOUS HYPOPLASTIC PERSISTENT FIFTH AORTIC ARCH CONNECTING ASCENDING AND DESCENDING AORTAWITH RIGHT DESCENDING AORTA FEEDING RPA FROM PDA – SUCCESSFULLRECONSTRUCTION

We report a 4 and 1/2 month male child first child of nonconsanguinously married couple diagnosed to have heart disease when admitted in local facility for fever and lethargy. Referred to us after treatment for surgical management of the heart disease. Echo from outside revealed Large subaortic and inlet VSD,small PDA with mild coarctation small Ascending Aorta and severe Pulmonary Hypertension.On examination,thinchild lookingalert, HR 150/min regular, PP+ CRT <3 sec,RR- 32/min, retractions, 100% on Oxygen 1L via nasal cannula, Respiratory system-Air entry good, occasional crepitations, CVS, S1S2 normal, systolic murmur +3/6 best heard LLSB,P/A- soft non tender,Hepatomegalymild,CNS- Alert, AF level,no focal deficits noted.Plan was to do routine investigations continue nasal cannula Oxygen, Furosemide 4mg PO TDS,continue RT feeds,

Echo revealed Situssolitus,LevocardiaAV-VA concordance, NRGA, Intact IAS,Nonrestrictiveperimembranous VSD Left to Right shunt,Laminar flow in LVOT/RVOT,NO TR/MR,Normal ventricular function,Tortuous and hypoplastic arch with retroesophageal Right subclavian artery originating from right descending aorta,NormalCoronaries,Retroaortic innominate vien.CT angiography was done which revealed similar findings.PDA from right descending aorta to RPA.Then the patient was taken for cardiac catheterisation. Catheter course- RFA-->D.Ao-->TRANSVERSE ARCH-->HYPOPLASTIC TORTUOUS ARCH-->AAo-->LV.Significant gradient of 37mmHg was found between ascending and descending aorta.Hence patient was taken for surgery.

STEPS OF SURGERY-
- Aorta and left arch with its branches extensively mobilised.PDA dissected out and traced back to the descending Aorta.Azygous vein doubly ligated and divided and the Descending Aorta mobilised so that it could be approximated to the ascending Aorta without tension.
- Patient heparinised and cardiopulmonary bypass established by cannulating the arch distal to the RtCA origin. Venous return via IVC and SVC cannulation.Patient cooled to 26 deg.PDA doubly ligated and divided and the ends oversewn.
- Aortic root antegradecardioplegia given after cross clamping the aorta at the root of the RCA origin. Cavae snugged. Heart arrested well.
- RA opened,stab PFO dilated ,heart vented
- Clamp applied on the descending Aorta and the feeding channel doubly
ligated and oversewn. Remaining channel mobilised to relieve the ring.
- Arteriotomy made on the posterior wall of the ascending aorta proximal to the RCA origin.
- Fibrous tissue transected off the descending Aortic end and the incision extended into the RSCA origin. The two Aortic ends were approximated and an end to side anastamosis done with 6/0 prolene suture.Deairing done and descending aortic clamp removed.
-Cross clamp repositioned on the ascending aorta proximal to the anastamosis, to perfuse the descending aorta.

- VSD anatomy discerned. VSD closed with preformed oblong dacron patch with interrupted 5 -0 prolenepledgetted sutures along the posteroinferior margin &continous 5-0 sutures for rest of the margins, so as to dedicate the aorta across the VSD to the LV. Suture line reinforced with interrupted 5-0 sutures in between.
- Rewarming started and PFO closed using 5-0 prolene suture.
- RA closed with 5-0 Prolene continuous suture. Heart deaired& cross clamp removed. Patient reverted to sinus rhythm.
 Patient rewarmed fully and gradually weaned off bypass. gradients of 5mm Hg between the upper and lower limb pressures(pre-op 35 mm Hg). Patient decannulated after reversing heparin.
- One retro cardiac and one right pleural drain inserted. Two atrial and ventricular wire.
- Hemostasis achieved and sternum reapproximated with No. 2 Ethibond after ensuring correct counts.
 Incision closed in layers.
- Patient shifted to ICU on IPPV with stable hemodynamics on vasoactive support .

CPB Time - 125 minutes, Aortic cross clamp time -76 min,Cooling - 26C
Cannula - 10 Fr,SVC - 12 Fr angled,IVC - 14 Fr angled,Oxygenator - Capiox Rx 05,Cardioplegia - aortic root,CUF - 400 ml.IntraopECHO revealed No residual VSD, No LVOTO,Normal ventricular function,LVOT Well opened. Trivial TR.
Post operatively child had Hypertension which was controlled with clonidine hydralazine and envas. Child was extubated on POD 9.

II. Discussion

During the *fourth and fifth* weeks of embryological development, when the pharyngeal arches form, the aortic sac gives rise to arteries – the aortic arches. The aortic sac is the endothelial lined dilation just distal to the truncus arteriosus; it is the primordial vascular channel from which the aortic arches arise. Each pharyngeal arch has its own cranial nerve and its own artery, hence we can conclude that the growth of the the aortic and pharyngeal arches are very closely related. The aortic arches terminate in the right and left dorsal aortae. The dorsal aortae remain paired in the region of the arches, however below this region they merge to form a single vessel (the descending/thoracic/abdominal aorta).

The pharyngeal arches and their vessels appear in a *cephalo-caudal* order, so they are not all present at the same time. As a new arch forms the aortic sac contributes a branch to it. In the initial stage there are pairs of aortic arches, which are numbered **I, II, III, IV, and V**. This system becomes altered in further development.

The **truncus arteriosus** divides into the **ventral aorta** and **pulmonary trunk** by the aortic-pulmonary septum. They represent the outflow channels of the heart. After this, the aortic sac then forms right and left horns. The right horn becomes the **brachiocephalic artery** and the left becomes the **proximal***(ascending)* part of the **aortic arch**.

The **1st Arch** and **2nd Arch** start to regress by approximately day 27, however portions of each persist as the **maxillary artery**, and the **hyoid** and **stapedial arteries**, respectively. By day 29 both these arches completely disappear. Around the time regression of the 1st and 2nd arches, the 3rd is large and 4th and 6th arches are forming. Soon the 3rd, 4th and 6th arches all appear large. Because of division of the truncus arteriosus, the 6th arches are now continuous with the pulmonary trunk, with the **primitive pulmonary artery** present as a major branch. As development continues, the aortic arch system loses its original symmetry. The following changes occur:

- **3rd Arch** : forms **common carotid artery**, **first *(cervical)* part of internal carotid artery** (*rest of internal carotid arises from dorsal aorta)*, and **external carotid artery**.
- **4th Arch**: on *LEFT* – forms **part of arch of aorta** (between the left common carotid and left subclavian arteries).

On *RIGHT* – forms proximal part of right subclavian artery, distal part of subclavian formed by right dorsal aorta and seventh intersegmental artery.
- **5th Arch**: either never forms or regresses after incomplete formation.
- **6th Arch (Pulmonary Arch)**: on *RIGHT* – forms proximal part of **right pulmonary artery**, *distal part of arch loses connection with dorsal aorta*.

On *LEFT* – forms **left pulmonary artery**, *distal part of arch persists* in intrauterine life as *ductus arteriosus*.
- The dorsal aorta between the third and fourth arches is obliterated. This part also known as the **carotid duct**.
- The right dorsal aorta disappears between origin of the *seven intersegmental artery* and the junction with the *left dorsal aorta*. *Brachiocephalic and common carotid arteries elongate due to folding and growth of embryo.

- The **left subclavian artery** arises from the **left intersegmental 6th artery** in the region of the 6th -7th cervical

- Persistent fifth aortic arch (PFAA) is a rare anomaly of aortic arch development consisting of a systemic-to-systemic connection through an arterial branch originating from the distal ascending aorta, proximal and opposite to the ostium of the innominate artery and connected to the descending aorta. The persistent fifth aortic arch is located between the true aortic arch (a derivative of the fourth embryological arch) and the pulmonary artery (a derivative of the sixth aortic arch artery) with no arterial branches originating from it. Thus, the aortic arch is subdivided into superior and inferior parallel channels completely separated by two adventitial layers (double-lumen aortic arch). Unlike the classic double aortic arch, which surrounds the trachea and oesophagus, this condition does not result in a vascular ring. The double aortic arch usually shows an anomalous pattern of epi-aortic vessels, with branching from both aortic arches .PFAA is thought to be an extremely rare congenital heart defect (CHD) because the fifth aortic arch usually involutes during normal embryologic development. In 1969, Van Praagh and Van Praagh first reported PFAA in a male patient with a double-lumen aortic arch with both arches on the same side of the trachea

- Although frequently associated with major cardiac anomalies, PFAA can be an incidental finding without clinical significance. Previously reported cases of PFAA have been associated with major CHDs, such as pulmonary atresia, tricuspid atresia, coarctation of aorta, interrupted aortic arch, ventricular septal defect, patent ductus arteriosus (PDA), atrioventricular septal defect, tetralogy of Fallot, and persistent truncus arteriosus.

PFAA is usually hemodynamically beneficial or much less harmful than the associated CHD and is diagnosed coincidentally. For example, double-lumen aortic arch is beneficial when associated with coarctation of the aorta or an interrupted aortic arch, and a systemic-to-pulmonary arterial connection is beneficial when associated with pulmonary atresia or tricuspid atresia. PFAA has been subcategorised into three subtypes: (I) double-lumen aortic arch with both lamina patent; (II) atresia and interruption of the superior arch with patent inferior (persistent fifth) arch; and (III) systemic-to-pulmonary arterial connection arising proximal to the first brachiocephalic artery . Type I PFAA is the most common form of these three subtypes; it can be diagnosed easily, although it is sometimes missed. Type III PFAA is extraordinarily difficult to differentiate from PDA or aorto-pulmonary window. Lee and his colleagues reported an infant with transposition of the great arteries, pulmonary atresia, and an artery arising from the origin of the innominate artery and connecting to the pulmonary arteries which they labelled as type III PFAA.

Left Aortic Arch with an Aberrant Right Subclavian Artery With Right Descending Aorta
An ARSA originating from normal left sided aortic arch is the most common aortic arch anomaly, with an incidence of 0.5-2%. This anomaly results from interruption of the dorsal segment of the right arch between the right carotid artery and right subclavian artery with regression of the right ductus arteriosus in the developing double aortic arch. In this anomaly, the right carotid artery arises as the first branch directly from the aortic arch, which is followed by the left carotid artery, left subclavian arteries and ARSA. The ARSA arises from the descending aorta as a last branch and crosses the mediastinum from left to right, passing behind the esophagus and trachea. An aortic diverticulum, also known as Kommerell's diverticulum, may be present at the origin of this vessel and has been reported in up to 60% of cases, representing the remnant of the distal RAA. An ARSA is generally asymptomatic and has been diagnosed incidentally, but about 10% of adults with this anomaly have symptoms of dysphagia due to extrinsic compression of the esophagus due to its retroesophageal course. Rarely, with failure regression of the right ductus, a loose vascular ring may be formed by the LAA with an ARSA, right pulmonary artery and right ductus arteriosus. Aneurysmal dilatation of the proximal portion of an ARSA is uncommon. Such an aneurysm may or may not be associated with Kommerell's diverticulum, and is believed to be the result of atherosclerotic disease. Generally, this anomaly is isolated, but may be associated with other cardiovascular anomalies, principally with coarctation of the aorta), a patent ductus arteriosus, intracardiac defects, anomalous pulmonary artery circulation and carotid or vertebral artery anomalies.
Interrupted aortic arch
Interrupted aortic arch has been classified into 3 types (A, B, and C) based on the site of aortic interruption. In type A interrupted left aortic arch, the arch interruption occurs distal to the origin of the left subclavian artery. In type B interrupted left aortic arch, the interruption occurs distal to the origin of the left common carotid artery. In type C interrupted left aortic arch, the interruption occurs proximal to the origin of the left common carotid artery.
In any of the 3 types, the right subclavian artery may arise normally or abnormally; the 2 most common abnormal sites are distal to the left subclavian artery (aberrant right subclavian artery) and from a right ductus

arteriosus (isolated right subclavian artery).Type B interruptions account for about two thirds of cases, type A occur in about one third of cases, and type C are present in less than 1% of cases.

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