A rare type of interruption of the aortic arch: case report

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Abstract

Introduction
We present the first report of interruption of the aortic arch in our region.

Case report
A three-day-old neonate succumbed at a referral hospital with a history leading to diagnosis of a cyanotic heart malformation. Autopsy revealed a pulmonary trunk that arched then continued as the descending aorta after giving small pulmonary arteries to both lungs. The left ventricle was drained by an aorta that gave rise to both coronary arteries and then continued on to give the brachiocephalic trunk, the left common carotid and the left subclavian arteries. The latter was the last branch of this aorta. This arrangement falls into the type A classification of interruption of the aortic arch.

Conclusion
This case highlights the need for a systematic assessment of neonates at birth in order to pick and appropriately manage congenital cyanotic heart diseases.

Introduction

Interruption of the aortic arch (IAA) is an infrequent cyanotic heart disease and is reported to occur in three per million live births and constitutes 1% of congenital heart disease. It is defined as loss of luminal continuity between the ascending and descending parts of the aorta. In 97% of the cases, it is usually associated with other cardiac anomalies such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction, or aortopulmonary window. The IAA anomaly was first described by Steidele in 1778. Celoria and Patton later classified IAA into three types according to the site of discontinuity of the aortic arch. Type A was the IAA where the site of discontinuity was distal to the left subclavian artery, type B, between the left carotid and left subclavian arteries and between the innominate and left carotid arteries, type C. The most common type is B (53%), followed by A (43%) and C (4%). We describe a type A IAA that coexists with bilateral hydroureter and a foramen secundum.

Case report
A female neonate with birth weight of 2550 gm developed cyanosis and perioral twitching soon after birth. She was a third born in the family, with a favourable antenatal period. The mother was 29-years old with no history of contraceptive or other drug use during gestation. On examination, a systolic murmur was heard all over the left side of the chest with hypotonia in all limbs, weak Moro and grasp reflexes. The suck reflex was absent. She also had sutural diasthesis. There were no abnormal abdominal findings. During autopsy, the baby weighed 2605 gm; her head circumference was 35 cm, crown rump length 35 cm, crown heel length 51 cm. There were no external abnormalities. Examination of the thorax revealed a situs solitus organization of the intrathoracic organs with a heart in levocardia (Figure 1). Further examination of the heart showed four chambers. The inferior vena cava (IVC) and superior vena cava (SVC) drained normally into the right atrium (RA). The RA and appendage were normal in position. There was a large foramen secundum connecting the two atria (Figure 2). The right ventricle normal in its

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formation (tripartite) with a normal inflow, tricuspid valve, normal body, normal outflow tract (infundibulum and pulmonary trunk). The pulmonary trunk (PT) originated normally from the right ventricle with normal trunk, with no stenosis. This artery immediately gave two pulmonary arteries to the left and right lungs and was connected by large ductus, same diameter as PT, with the descending aorta (Figure 3). All the four pulmonary veins were found to enter normally into the left atrium (LA). The LA and its appendage appeared normal. There was normal left ventricular flow: mitral valve and the infundibulum. The left ventricle (LV) septum appeared intact with a normal body. The aorta was found to connect normally to the LV and the aortic valve was also normal. The aorta extended vertically, branching into the brachiocephalic trunk (BCA), Left common carotid (LCC) and the left subclavian artery (LSA) with an interruption after the LSA. The RCA and LCA arose from the aorta (Figure 4). Other findings comprised bilateral hydronephrosis and hydroureter (Figure 5), pulmonary oedema and brain oedema.

Discussion

The above findings were compatible with an IAA type A. IAA represents 1% of congenital heart diseases. This rare cyanotic heart anomaly is almost predictably accompanied by other congenital anomalies. The most common association recorded is being a patent ductus arteriosus and a ventricular septal defect. The most recent reported rare association is that of an ASD of a venosum type and anomalous pulmonary connection. In this report, the IAA coexists with an ASD of the secundum type.

The median age of death in IAA associated with cardiac anomalies has been reported to be 10 days. The cause of death being attributed to increased pulmonary hypertension and the resultant biventricular failure. Not with standing, there are cases that are detected late into adulthood. Krishna et al. reported IAA in several adults including a 64-year old. Collateral allows for survival of these patients into adulthood.

IAA is said to occur as a result of defective embryology of the primitive aortae and aortic arches. According to this author, type A IAA is formed by the abnormal regression of the left fourth arch segment late in development after the left subclavian artery has ascended to its normal position. A type B IAA is found when the left fourth arch segment regresses early, prior to cephalad migration of the left subclavian artery.

On the other hand, a type C IAA represents involution of the ventral portion of the left third arch and the left fourth arch (both of which arise from the left limb of the aortic sac) and persistence of the normally regressing ductus caroticus.

![Figure 3:](image-url) 3a and 3b illustrate ventricular outflow. The yellow arrow shows the right ventricular chamber. The glass rods (red arrows), show the right and left pulmonary arteries, respectively (3a). (3b) is close up view to show the pulmonary trunk (PT), the left pulmonary artery (glass rod), the ductus arteriosus (DA) and the descending thoracic aorta (DTA), the DTA has been unfolded. Note that the size of the DA is the same as that of the DTA and PT. The branches of the left ventricle can also be noted; 1 is the brachiocephalic trunc, 2 is the left common carotid, 3 the left subclavian. LL (left lung).

![Figure 4:](image-url) Showing the left ventricular outflow. 1 is the brachiocephalic trunc, 2 is the left common carotid, 3 is the left subclavian artery. RV, right ventricle; LV, left ventricle; DTA, descending thoracic aorta. Note that the probes are in continuation with the LV chamber.
The clinical pattern and chest X-ray are not diagnostic of IAA. Nonetheless, bounding carotid pulses and a weak peripheral pulse should heighten suspicion, but differential cyanosis is almost pathognomonic. With suspicion from the clinical signs, angiography is recommended. It is worth noting that Di George syndrome is associated with the highest incidence of IAA. Out of the seven patients with Di George syndrome studied by Finley et al., five of them had IAA. In our report, the thymus was present.

Association of congenital heart anomalies with urinary tract malformations is documented. According to these workers, atrial septal defects stood a higher chance of concurrence with a urinary tract malformation, while those with ventricular septal defects and tetralogy of fallot had an average incidence. Due to the coexistence observed in this case report, the possibility of a urinary tract malformation should be considered in the case where such an anomalies presents again.

**Conclusion**

This case highlights the need for a systematic assessment of neonates at birth in order to pick and appropriately manage congenital cyanotic heart diseases.

**Consent**

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**References**


**Figure 5:** Shows the dilated ureters, Ur, and renal pelvis (red arrows). B, is the bladder; K, kidney; C, colon. In 5b, note the dilated renal pelvis as shown by the red arrows.