

**Case Report**

Extra Anatomic Repair of Interrupted Aortic Arch

Anirudh Kumar Paidi¹, Palli Venkata Naresh Kumar¹, Madhava Rao Midhe²,
Ravi Kiran Mamidala¹, Venkat Reddy Navuluri³

¹Department of Cardiovascular and Thoracic Surgery, Yashoda Hospitals, Hyderabad, India

²Department of Cardiac Anaesthesia, Yashoda Hospitals, Hyderabad, India

³Department of Perfusion, Yashoda Hospitals, Hyderabad, India

Email address:

dranirudhkumar@gmail.com (A. K. Paidi), pvnaresh@hotmail.com (P. V. N. Kumar)

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Abstract: Interrupted Aortic Arch is a rare congenital anomaly with an incidence of 2 per 100,000 live births. It is almost always (95-97%) associated with structural abnormality like VSD. Interrupted Aortic Arch is usually diagnosed soon after birth, and when left untreated results in 90% mortality at a median age of 4 days. Isolated IAA is a very rare entity. Substantial collateral circulation must be present to maintain flow and enable survival. Conventional surgical repair is a challenge due to extensive collateral circulation in patients who have survived to adulthood. Here, we report a patient with an isolated IAA without any associated defect who survived into third decade. The patient presented with complaints of palpitations, uncontrolled hypertension, and weakness in lower limbs and on evaluation, he had isolated Interrupted Aortic Arch. We managed to do an Extra anatomical Bypass graft from ascending aorta to descending aorta on a beating heart with minimal risk. The procedure was done through median sternotomy to avoid the damage to collaterals when approached through thoracotomy.

Keywords: Interrupted Aortic Arch, Extra Anatomical Bypass, Sternotomy, Collaterals, IAA

1. Introduction

Interrupted Aortic Arch (IAA) is an uncommon congenital cardiovascular malformation. It is considered as the most severe form of aortic coarctation [1, 4, 7, 21]. In an IAA, there is an anatomical disruption between ascending and descending aorta. It was first described by Steidle in 1778 and is characterised by lack of luminal continuity between ascending and descending aorta [1]. It is almost always (95-97%) associated with structural abnormality like VSD. According to Van Mierop, about 68% of patients with IAA have DiGeorge syndrome [24, 28]. It is typically associated with Type B IAA. Ideal age of operate is within first week of life due to pulmonary overcirculation. Here, we report a patient with an isolated IAA without any associated defect who survived into third decade. The repair of IAA can be divided into two types as single staged repair and staged repair. A single stage repair is almost always advised in adults with

IAA. A direct anastomosis after adequate mobilisation of both ascending and descending aorta without patch augmentation is thought to provide satisfactory patency [18-22]. However, risk of recurrent obstruction due to tension at the anastomosis is found [20, 23]. Direct anastomosis with patch augmentation is considered by many surgeons to be an optimal method of repair [24, 20, 25, 27]. A homograft or autologous pericardial patch can achieve a tension free anastomosis with low incidence of recoarctation [24, 4, 25]. But there is every chance of collateral damage in these procedures. An extra anatomical bypass with a PTFE conduit can prevent thoracotomy related complications which may cause extensive collateral damage.

2. Case Report

A 23 year old male patient, a known hypertensive, presented with complaints of palpitations, uncontrolled hypertension, and weakness in lower limbs. On examination,

arterial pulses were feeble in both lower limbs when compared to both upper limbs with a radio-femoral delay. His Blood pressure in upper limbs is 145/82 mm of Hg and in lower limbs is 108/67 mm of Hg. On further evaluation, 2D Echo showed Good LV function (EF=62%) with concentric LVH, no structural abnormality. CT-Thoraco abdominal aortogram revealed interruption of proximal descending aorta at D6 level for a length of 2.7mm with mild narrowing of distal aortic arch, distal to left subclavian artery. Multiple collaterals noted in chest wall and para spinal region from dilated intercostals and internal mammary arteries, reforming descending thoracic aorta - Interrupted Aortic Arch Type A (Figure 1).



Figure 1. Preoperative CT Thoracoabdominal Aortogram showing Type A Interrupted Aortic Arch.



Figure 2. Intraoperative picture showing graft connecting Ascending Aorta to Descending Aorta. a=ascending aorta; b=right atrium; c=right ventricle; d=extra anatomical bypass graft from ascending aorta to descending aorta.

The patient was scheduled for surgical repair. Initially, the repair was planned through thoracotomy, but due to the presence of multiple collaterals in the chest wall, he was approached through median sternotomy. An extra anatomical (Ascending – Descending) aortic bypass was planned. High aortic, bi-caval (SVC, IVC) cannulation done and went on cardio-pulmonary bypass. Right pleura was opened. Heart

was mobilised into the right pleura for better view of posterior pericardium. The descending aorta was exposed through a longitudinal incision in the posterior pericardium just cephalad to the diaphragm. Side biting clamp applied to the exposed part of descending aorta and distal anastomosis was constructed using 16mm Woven-polyester graft prosthesis (Figure 2, Figure 3) using continuous 5-0 prolene suture. The graft was then passed anterior to the IVC on the right side along the free wall of RA before joining it to the ascending aorta. Side biting clamp was applied over the lateral aspect of the ascending aorta and proximal anastomosis was constructed with continuous 5-0 prolene suture. Patient was gradually weaned off from cardio-pulmonary bypass in normal sinus rhythm and stable hemodynamic. Throughout the operative procedure, femoral arterial pressures were maintained at least at 40mm of Hg. The complete procedure was done on cardio-pulmonary bypass without arresting the heart. The patient had an uneventful recovery and was maintained on minimal anti-hypertensive medication. The patient is doing well on follow up.

3. Result

Patient was extubated in 4 hours after surgery and shifted out of ICU on post operative day 2. He is discharged in 4 days. the patient had an uneventful recovery and follow up after 6 months, he is doing well. CT aortogram showed a patent graft (Figure 4).

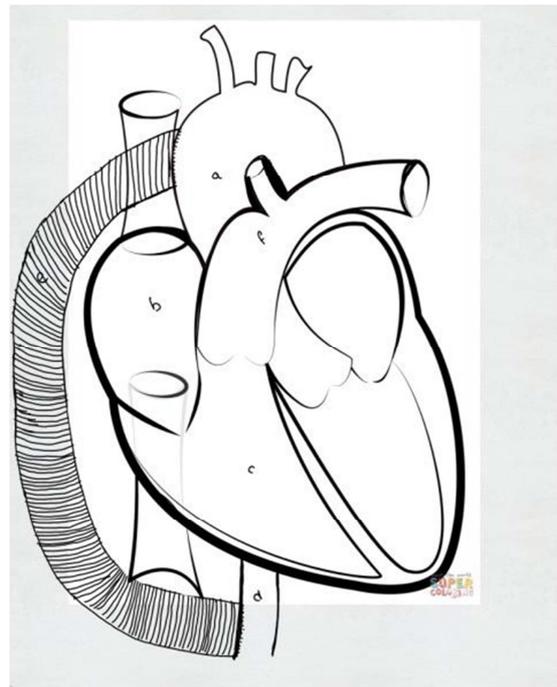


Figure 3. Post operative illustration showing extra anatomic graft positioned between ascending and descending aorta. Graft positioned on the right side along the free wall of right atrium anterior to Inferior vena cava.

a= Ascending aorta; b=Right atrium; c=Right ventricle; d=Descending aorta; e=graft; f=pulmonary artery.



Figure 4. Post operative CT aortogram showing patent graft.

4. Discussion

Interrupted Aortic Arch accounts for approximately 1.5% of all congenital cardiac anomalies. It is usually associated with ventricular septal defect, Patent ductus arteriosus, Truncus arteriosus and Transposition of Great arteries. Isolated IAA is a very rare entity. It has got a very poor prognosis when left untreated [2]. IAA is sub divided into 3 types based on site of interruption [3].

Type A: The interruption is beyond the left subclavian artery. 40% of all cases of IAA belong to this type.

Type B: The interruption occurs between the left common carotid and left subclavian arteries. It is most common type of IAA which accounts for 50-55% of reported cases.

Type C: The interruption occurs between innominate artery and left common carotid artery. Is is a rare form of IAA accounting for less than 5% of reported cases.

Interrupted Aortic Arch is usually diagnosed soon after birth, and when left untreated results in 90% mortality at a median age of 4 days [4]. In the few documented cases in adults, the presentation ranges from lack of symptoms to limb swelling with differential blood pressures in all extremities. Substantial collateral circulation must be present to maintain flow and enable survival.

This technique avoids extensive damage to collaterals which encounters in Thoracotomy. This technique reduces the complications of cardioplegia and conventional anatomic repair. It yields minimal intraoperative blood loss and early post operative recovery without any sequelae.

Consent

Patient gave informed written consent for this publication.

Conflicts of Interest

There are no conflicts of interest.

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