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Surgical Correction of Aorto-Pulmonary Window: A rare and lethal cause of pulmonary hypertension

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Abstract

Aorto-pulmonary window is a relatively rare congenital cardiac malformation with an overall incidence of 0.1%. Pulmonary hypertension develops quickly if the lesion is left untreated hence early surgical intervention is warranted after diagnosis. The surgery for correction of APW has evolved over years, currently an open repair on cardiopulmonary bypass (CPB) with a single patch technique yields good results. Mortality is affected by association of pulmonary hypertension and other cardiac malformations. We present a case of an infant with a large type II APW with a relatively low pulmonary vascular resistance. Hospital stay was complicated because of pulmonary arterial disease making it an important reason for correction in the first few months of life.

Keywords: Aorto-pulmonary window, Surgical repair, Pulmonary hypertension.

Introduction

Aorto-pulmonary window (APW) is an uncommon congenital anomaly arising in the great vessels of the heart. It is characterized by a communication between ascending aorta

and the pulmonary trunk. The development and structure of the aortic and pulmonary semi lunar valves is normal.¹ Various sizes and shapes of APW have been documented, yet the classification depends upon the location of the APW rendering it proximal, distal, total or intermediate.^{2,3} Surgical correction of the APW, immediately after diagnosis, is imperative to avert irreversible pulmonary hypertension which develops earlier as compared to other left to right shunts. Once significant pulmonary hypertension has set in, surgical intervention of the APW leads to pulmonary hypertensive crises and right sided heart failure, thus substantiating early surgical rectification.² We present a case of APW in a seven month old child where postoperative course was complicated because of pulmonary hypertensive disease- a manifestation of pulmonary vascular disease- hence reinforcing the earliest possible diagnosis and closure of the defect.

Case Report

A 7 month old male infant was electively admitted with a heart rate of 132 beats per minute, respiratory rate of 36 breaths per minute, O₂ saturation of 93% on room air and blood pressure of 90/50 in both arms. The child was

cyanosed, dehydrated and pale. Precordium examination revealed a soft pan-systolic murmur in the lower left sternal border. Previous medical history included Acute Respiratory Distress Syndrome (ARDS) and per-rectal bleed on 1st day of life, along with multiple chest infections during the last 7 months. Chest radiograph demonstrated Cardiomegaly with pulmonary plethora. An echocardiogram demonstrated normal systemic, pulmonary venous and cardiac connections. The left ventricle was enlarged. There was, mild tricuspid regurgitation indicating low pulmonary pressures. The aortic arch was on the right side with no Coarctation. A tiny patent foramen ovale (2mm), with left to right shunt and a large distal APW (7mm), were illustrated on colour Doppler. Mild pulmonary regurgitation was present. With a diagnosis of Type II APW and mild pulmonary hypertension and a PFO the infant was scheduled for elective corrective surgery. Pulmonary hypertension was a very significant concern in this patient. More than one pharmacological technique was used to reduce pulmonary hypertension. Hydrocortisone 25 mg (3mg/kg) at the time of induction was used to reduce the inflammatory response of Cardiopulmonary bypass. Other pharmacological management was done with infusion of Milrinone 0.8µ/kg/min started after induction and continued peri and post operatively. Infusion of PGE1 (Aloprost) 0.05µ/kg/min and Magnesium sulphate 0.4 meq/Kg on CPB was also used. The non-pharmacological measures included mild hyperventilation, 100% oxygen and head up tilt.

The patient was operated upon by a median sternotomy; and thymectomy pericardiotomy was performed. The heart was found to be hyper-dynamic. The APW was identified as the junction of the left lateral wall of the aorta and the right wall of the proximal pulmonary artery. Cardiopulmonary bypass was established by right atrial and high aortic cannulation just beneath the innominate artery, this allows for placing the cross clamp and provides space to open the APW along its entire length. Patient was cooled to 32°C. Aortic cross clamp was applied and myocardial protection was achieved by snaring the branch pulmonary arteries (to prevent the cardioplegia from entering the pulmonary circulation) and instilling cold blood cardioplegia into the aortic root which resulted in prompt diastolic arrest.

Anterior incision into the wall of the APW was made to visualise the defect; It was a type II APW (Richardson et al);⁴ the sinus of valsalva and right pulmonary artery were directly communicating through the APW (Figure-1). After identification of branch pulmonary arteries and the right coronary artery a patch of grutaldehyde fixed autologous pericardium was sutured to the edges of the window to separate the common trunk into aorta and the pulmonary artery (Figure-2). The right atrium was opened and the PFO was primarily closed. After closing the right atrium the baby was re-warmed and the aortic cross clamp removed (Cross-

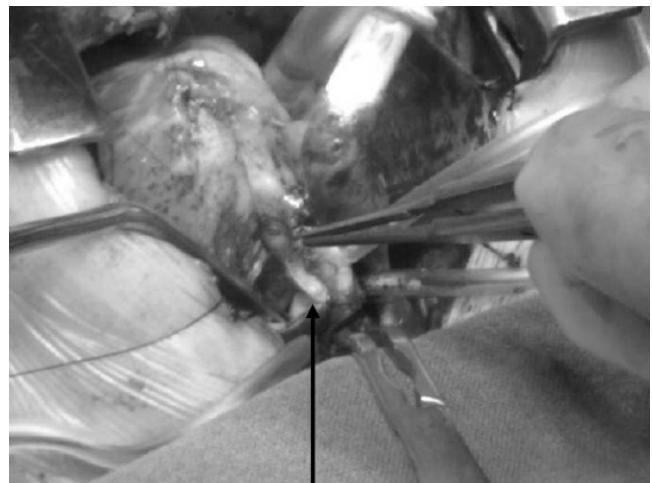


Right pulmonary artery opening

Figure-1: Pulmonary artery opening into the window.

clamp time 40 minutes). The patient came promptly into sinus rhythm and the cardiopulmonary bypass was discontinued with ease after 75 minutes, on minimal inotropic support. The post operative Aortic pressures were 60/32 (mean 45) and Pulmonary artery 29/15 (Mean 22) mmHg respectively. These low pulmonary pressures were achieved because of a combination of anaesthetic, surgical and CPB measures. Post-op stay in the CICU was stable. The baby was extubated after 21 hours. The chest tubes were removed on first post-op day (POD). Inotropic support was discontinued after 36 hours. Post-op, echocardiogram showed good biventricular function with no residual shunt.

The patient was shifted to a general ward on 4th POD. In the ward his reactive airway disease flared up with



Autologous pericardium being sewn

Figure-2: Autologous pericardial patch repair of the window.

excessive coughing, requiring intensive chest physiotherapy and bronchodilators. On 10th POD he developed sterile sternal dehiscence which required operative sternal rewiring. During the whole hospital stay he did not show any evidence of pulmonary hypertensive crisis. This could be attributed to management strategies adopted at the time of induction and in the early phase of post-operative recovery period.

Discussion

APW occurring as a solitary deformation has an incidence of 0.1% to 0.2% in congenital heart diseases⁵ hence the reported surgical experience is minimal. The embryogenesis of APW, pertinent from the fifth to eight week of intrauterine life, is associated with malalignment and incomplete fusion of the conotruncal ridges, on both sides. Therefore, septation of the truncus arteriosus - outflow tracts - fails to develop; resulting in a communication between right and left regions.⁶ Location of the defect and its relation to the ascending aorta and the pulmonary artery determines the type of the defect. Presently the defect is classified into three types. Type I is a simple defect between the ascending aorta and the pulmonary artery, type II defect is more distal with one of the branch pulmonary artery opening into it and type III defect is characterized by abnormal origin of the pulmonary artery from aorta.⁴ The defect can also be classified as simple (isolated or in association with an ASD or VSD) or complex (associated with IAA, TGA or TOF).⁷ Currently the diagnosis is confirmed with echocardiography, right heart cath being reserved for cases with doubtful diagnosis or to check for reversibility of pulmonary hypertension.⁷ Development of accelerated pulmonary hypertension is an important consideration with the result of surgery directly related to its development and the age at presentation.²

The first reported surgical closure of the defect was by Robert E Gross⁸ in 1946 after which the technique developed with the advancement in the field of congenital cardiac surgery from direct ligation without cardiopulmonary bypass to the presently established transaortic single patch technique. Treatment of choice after diagnosis is surgical while unrepaired cases have a dreadful outcome, with 40% of the infants dying during the first year of life. Moreover, a large proportion of the surviving infants will die from congestive

heart failure and pulmonary hypertension in childhood.⁴

However surgery is contraindicated in patients with PVR of more than 10 Woods units/ m², when Qp/Qs ratio is less than 1.5 and SaO₂ less than 90%.⁹ Surgical treatment early on in life is associated with low operative mortality, while concurrent cardiac abnormalities govern the prognosis in these cases. In older patients, prognosis solely depends on the degree of pulmonary vascular resistance.² Simple aortopulmonary window repaired before the development of significant pulmonary vascular disease carries a good long term prognosis.⁷ Association of pulmonary hypertension and other cardiac anomalies are the important determinants of peri-operative mortality.⁸

Conclusion

APW should be operated soon after birth or diagnosis to get best long term results. With proper peri-operative management, pulmonary hypertension can be controlled with good outcome. Since APW is rare it should be operated in a congenital cardiac surgery unit well versed with managing pulmonary hypertensive crisis in the neonatal and infant age groups.

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