TYPE A INTERRUPTED AORTIC ARCH WITH TAUSSIG-BING ANOMALY: AN UNUSUAL INDICATION FOR STAGED REPAIR

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ABSTRACT

Complex transposition of great arteries (TGA) is when additional cono-truncal anomalies coexist with a TGA. Notable reasons for staged repair include the need to the need to adapt the left ventricle to the systemic high pressures. We report a case of staged repair of Type A Interrupted aortic arch with Taussig-Bing anomaly in a one month old male. Single stage repair could not be done for this patient because of technical difficulties, positive preoperative blood culture and associated airway problem. Patient had a successful two stage surgery involving an initial extended end-to-end anastomosis, PDA ligation with PA Banding and a delayed arterial switch operation done two months later.

KEY WORDS: Interrupted aortic arch, arterial switch operation, Taussig-Bing Anomaly, Transposition of Great arteries.

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INTRODUCTION

Complex transposition of great arteries refers to conditions in which there are additional cono-truncal anomalies in addition to a complete transposition of the great vessels across the ventricular septum. It includes patients with persistence of bilateral conus giving rise to the Taussig Bing anomaly. Interruption of the aortic arch (IAA) represents a critical congenital heart disease which necessitates surgical intervention during the first days of life.[1] The incidence of transposition of the great arteries in patients with interrupted aortic arch is put at four per cent.[2] The coexistence of Interrupted Aortic Arch with congenital cardiac defect is associated with a high mortality.[3] This mortality is worsened by concurrent sepsis.

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Here we report our challenges with the management of an unusual case of Taussig Bing anomaly with Type A interrupted aortic arch associated with positive preoperative blood culture that contraindicated cardiopulmonary bypass for single stage repair.

Our patient was a month old male baby who was delivered at term to a 39-year-old mother. Birth weight was 3kg. He presented with respiratory distress that commenced shortly after birth. Respiratory distress had progressively worsened over time, was associated with poor suck but no history of excessive drooling of saliva or cyanosis. On examination at presentation he weighed 3.5kg, had cardiovascular instability with significant difference in radio-femoral systolic pressures. He was resuscitated with Oxygen therapy and inotropes.

Echocardiographic evaluation revealed situs solitus heart with levocardia, normal systemic and pulmonary venous drainage

with, atrio-ventricular concordance. There was ventriculo-arterial discordance and a sub-pulmonic ventricular septal defect. Both ascending aorta and main pulmonary artery sizes were adequate for weight. However, the aortic arch was interrupted immediately distal to the origin of the right subclavian artery with the ductus arteriosus continuing as the descending aorta (Type A interrupted arch). A diagnosis of Taussig Bing anomaly with Type A Interrupted aortic arch was made.

Blood culture revealed a positivemixed bacterial and yeast culture. The surgical decision was to do an emergent repair of interrupted aortic arch, PDA ligation and PA banding to protect pulmonary vasculature and, a delayed corrective arterial switch and VSD closure.

Patient had an emergency left thoracotomy. Aortic arch, arch vessels, PDA and descending aorta were dissected. Findings were same as identified on echocardiography. Occlusion clamps were applied to distal aortic arch, proximal descending aorta and proximal ductus arteriosus. PDA was transected and ligated, undersurface of aortic arch was opened and an extended end to end anastomosis was made between the arch and descending aorta. Clamp time was 15 minutes. Good descending aortic flow was confirmed by palpable pulsation and improvement of femoral pressures. PA banding was done.

Delayed arterial switch was done two months later to allow for patient optimization for surgery. Access was via a sternotomy. Intra-operative findings were mild pericardial adhesions, PA band in-situ, main pulmonary artery was twice the size of the ascending aorta. Following adhesiolysis and commencement of standard cardiopulmonary bypass, patient was cooled to 28 degrees centigrade. Antegrade blood-crystalloid cardioplegia was administered following aortic cross-clamp. Oblique right atriotomy was done, Left atrium was vented through the patent foramen ovale.

Aortotomy was done just above the sino-tubular junction, the coronary buttons were harvested. Pulmonary arteriotomy was done. The coronary buttons were implanted into the neo-aorta via the trap door technique. Ventricular septal defect was closed using a Goretex patch via the pulmonary valve. Then, the aorta was then brought posterior to the Pulmonary artery (Le Compte's maneuver). Aortic anastomosis was done using Polypropylene stitches. The neo-pulmonary artery stump was reconstructed using fresh pericardial patch. Patient was re-warmed, Left atrium was de-aired, and clamp was taken off, pulmonary artery anastomosis done. Right atrium was closed in two layers. Pacing leads, chest drains were inserted. We were unable to close the chest primarily because of raised intra-thoracic pressure, hence chest was covered with sterile band. Chest closure was done after 24 hours following normalization of intra-thoracic pressures. Post-operative period was uneventful. Post-operative period was uneventful. Patient has been having uneventful follow up in the outpatient clinic. Weight gain and milestones are achieved as expected.

DISCUSSION

The association of Interrupted Aortic Arch and intra-cardiac anomaly portends a high mortality in the absence of surgical correction.[3] Two stage procedure, with initial surgery addressing the aortic interruption and the subsequent surgery addressing the intra-cardiac lesion, had been the preferred treatment modality.[4] Recently, Single stage repairs were proposed.[2]

Experience with patients who have perioperative sepsis and cardiopulmonary bypass early in infancy suggest that sepsis is associated with adverse outcome.[5] We

considered that our patient could not undergo a single stage correction because he had sepsis with positive yeast cultures preoperatively. The irrelevance of isolated positive cultures from cardiopulmonary bypass prime and blood as well as its non-correlation to incidence of postoperative infections has been documented.[6] However, a pre-operative positive blood culture with clinical evidence of airway obstruction and systemic inflammatory response, that was the case in our patient, was considered to be a contra indication to cardiopulmonary bypass. This is because the cardiopulmonary bypass could serve as a second hit and has been shown to have adverse outcome.[5]

The cardio-respiratory instability was most likely as a result of several reasons, to include: the ductal dependent lower body circulation, the systemic inflammatory response to chest infection and a possible airway compression by the enlarged duct. Following the initial surgery – extended end to end anastomosis, inotropic requirements waned. However, ventilator requirement were high in the immediate post operative period. This is thought to be due to stiffness of the lung from consolidation and mucus accumulation resulting from bronchial obstruction by the enlarged duct. With positive end expiratory pressure ventilation and chest physiotherapy, patient was weaned off the ventilator within 48 hours.

The extended end – to – end anastomosis was also considered a rescue surgery to forestall gut malperfusion resulting from duct dependent lower body circulation.

CONCLUSION

In the presence of systemic inflammatory response syndrome in a patient with Taussig – Bing anomaly and Interrupted aortic arch, an initial minimal salvage surgery may be preferred to a single stage correction.

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