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Successful surgical repair of berry syndrome with severe biventricular dysfunction

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ABSTRACT

Background - Absence of the aortopulmonary septum with the presence of two separate semilunar valves, interrupted aortic arch, aortic origin of the right pulmonary artery, intact ventricular septum, and patent ductus arteriosus is a rarely reported association known as Berry syndrome. This abnormal right pulmonary arterial origin may lead to "steal" from the aortic flow during embryogenesis and cause hypoplasia of the aortic arch. Most patients present in infancy or earlier with symptoms of cardiac failure. Scattered reports in the literature confirm the possibility of surgical correction of this complex anomaly.

Case Report – We report a case of 7 week old cyanotic child who presented in emergency with features of congestive cardiac failure. Two dimensional echocardiography and CT scan confirmed the diagnosis of interrupted aortic arch with large aorto-pulmonary window. Single stage repair of the defect was done with the use of PTFE patch and autologous pericardium.

Conclusion – Berry Syndrome is a rare congenital cardiac anomaly with patients presenting early in infancy with congestive cardiac failure. Early single stage correction of the defect is indicated. Different surgical options have been tried but we used an intra aortic PTFE patch to baffle the RPA towards MPA without detaching the RPA and an autologous pericardium to augment the aorta and it translated into a shorter clamp time.

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Introduction

Berry syndrome is a rare cardiac malformation characterized by the absence of the aortopulmonary septum with the presence of two separate semilunar valves, interrupted aortic arch, aortic origin of the right pulmonary artery, intact ventricular septum, and patent ductus arteriosus.[1] Aortopulmonary window (APW) is a communication between the two great vessels, occurring above 2 normally formed semilunar valves. APW is accompanied by other cardiac anomalies in around 52% of cases, of which Interrupted aortic arch (IAA) is the most frequently associated anomaly.[2] APW can be classified into 4 types: proximal, distal, total and intermediate.[3] Patients may be asymptomatic or present with symptoms of pulmonary hypertension and congestive heart failure in the first few weeks of life depending on the size of the defect.[4] IAA is defined as a loss of luminal continuity between the ascending and descending portions of the aorta, with Patent Ductus Arteriosus (PDA) and systemic collaterals supplying the Descending Thoracic aorta (DTA) that occurs in 3 per one million live births. It is classified into type A, B and C depending on the site of interruption.[5,6] We present the case of a 7-week old child of Berry Syndrome with severe Mitral Regurgitation (MR), Tricuspid Regurgitation (TR) and severe biventricular dysfunction, who underwent a successful single-stage repair, which is yet to be reported.

Case report

A 7-week old cyanotic child presented to the emergency department with complaints of respiratory distress and fever. The chest x-ray revealed an increased cardiothoracic ratio of 60% and increased pulmonary vascular markings. The patient was intubated on admission and managed with diuretic therapy and antibiotics. 2D-Echocardiography (Figure-1) revealed a large APW (Type-II), severe MR and severe TR with severe biventricular dysfunction, interrupted aortic arch just distal to the

origin of the left subclavian artery (Type-A) and DTA being supplied by a large PDA originating from distal main pulmonary artery (MPA).

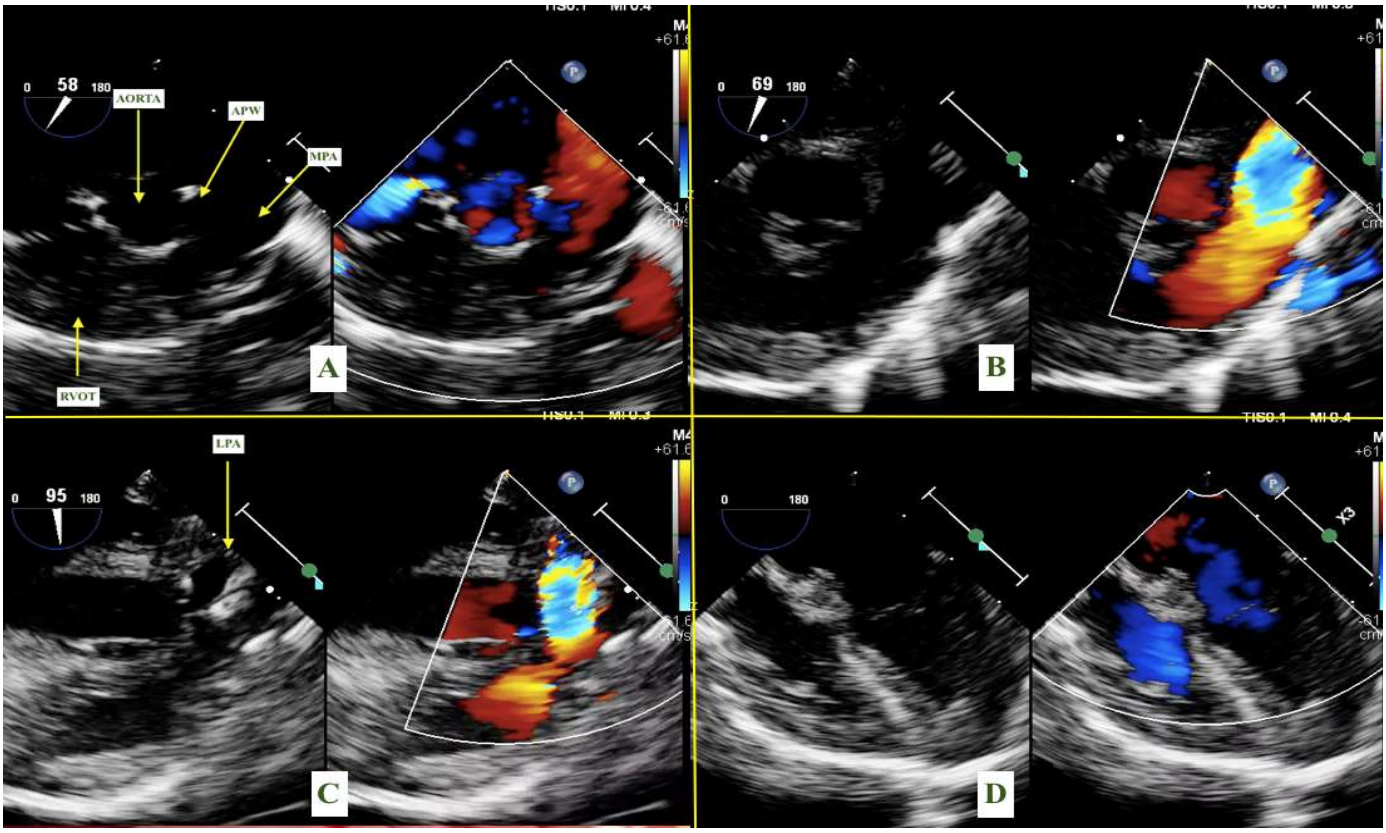

Figure-1-2 D-Echocardiography Images-(A)Pre-operative aortic valve short axis view showing a communication between ascending aorta and MPA with bidirectional shunt, (B)Post-operative aortic valve short axis view showing closed communication between ascending aorta and MPA, (C) Post-operative long axis view showing MPA connected through a baffle across ascending aorta to LPA, (D) Post-operative four chamber view showing no MR/TR

CT-Aortogram(Figure-2) in addition to the above findings, revealed an aortic origin of the Right Pulmonary Artery(RPA).

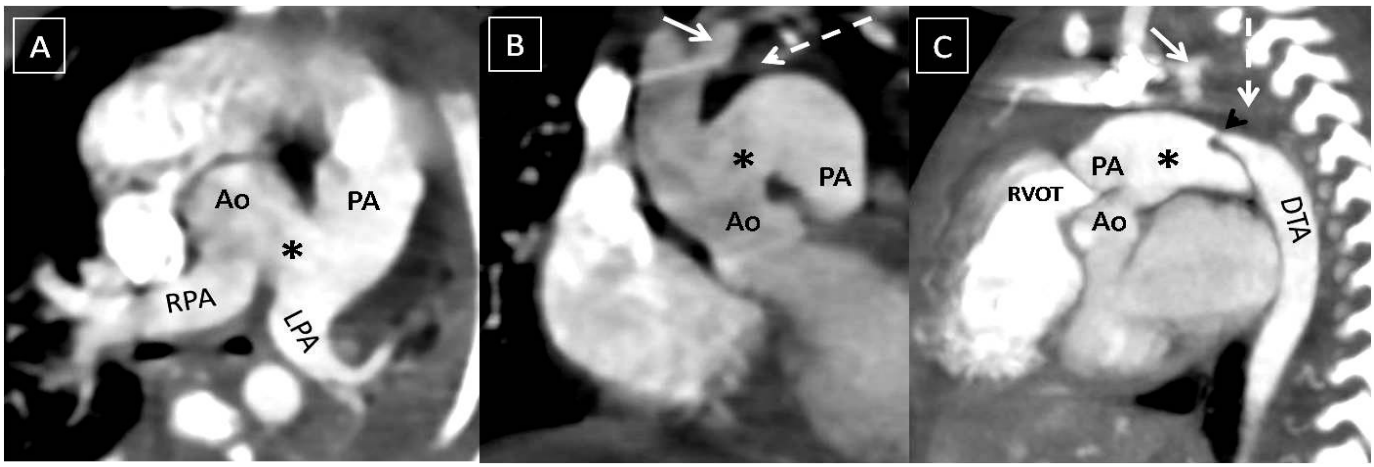

Figure-2-CT Angiography- (A) Distal large APW (*) with anomalous origin of RPA from aorta, (B,C) Type A-IAA*(region of interruption shown by dashed arrow in B,C*), distal to left subclavian artery(*arrow in B,C*) and reformation of DTA via PDA(*arrowhead in C*)

The patient was taken up for a single-stage repair. After median sternotomy, the pericardium was harvested and prepared. Arch vessels, bilateral Pulmonary arteries and DTA were dissected and looped (Figure-1A). Dual-aortic cannulation was planned, using one aortic purse string at the base of the innominate artery and another purse string on the distal MPA near the origin of the PDA(Figure-1B, C). Bicaval cannulation was done and Cardiopulmonary bypass was initiated after systemic heparinization with the child being cooled to 28℃. A vertical incision was made on the anterior surface of the ascending aorta near the lesser curvature (Figure-1D). Separate orifices of RPA and MPA were identified on the posterior aspect of the distal ascending aorta.

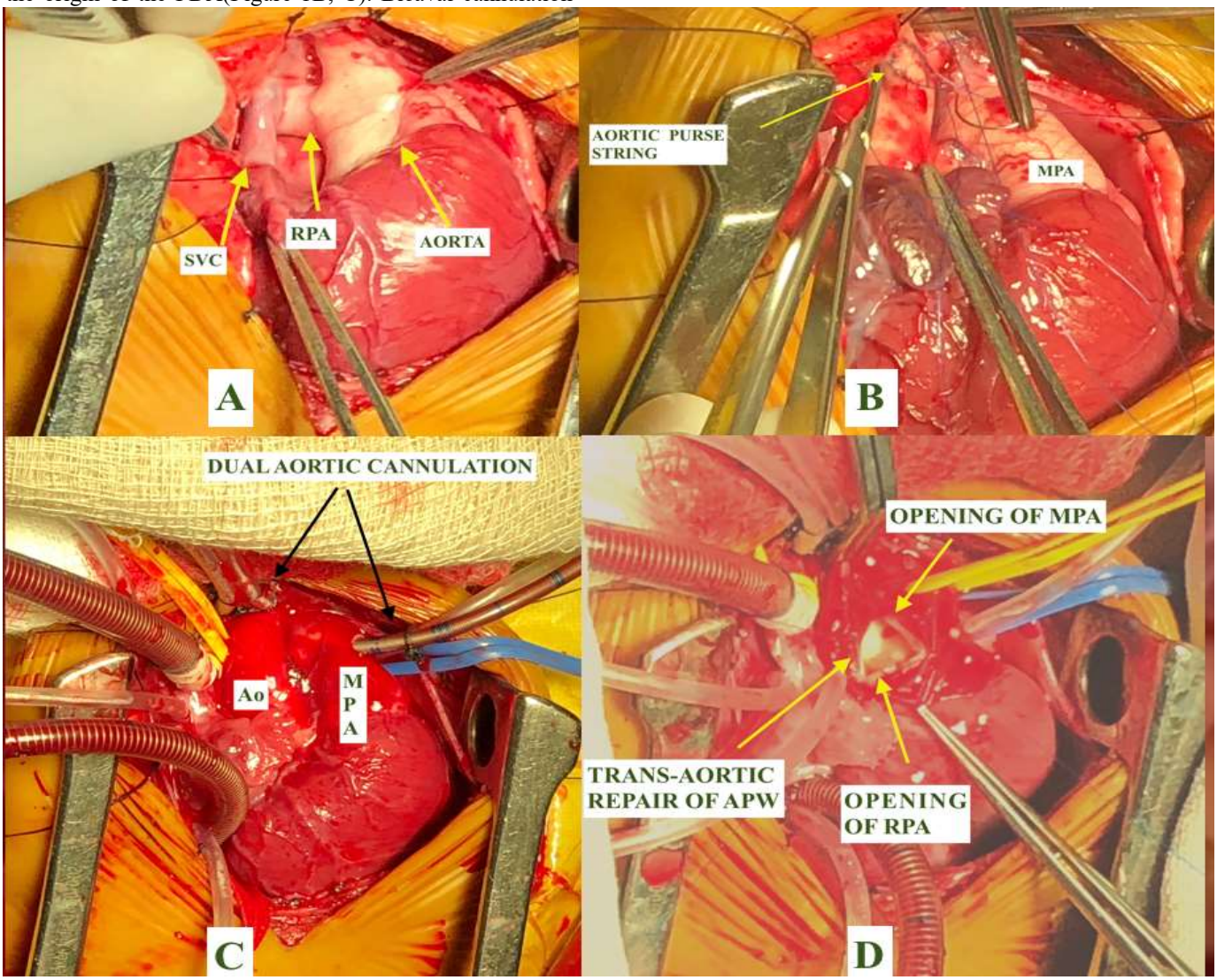

Figure-1-Intra-Operative Pictures–(A)Origin of RPA from Aorta, (B)Aortic purse string being taken as distally as possible, (C)Dual-Aortic Cannulation with cannula in Ascending aorta and MPA, (D)Trans-aortic opening of APW with openings of both MPA and RPA being visualized

An intra-aortic Polytetrafluoroethylene (PTFE) patch was sutured in such a way to baffle the RPA and LPA orifice towards the MPA, taking care that the origin of both coronary arteries was on the aortic side of the patch. The aortic cannula placed in MPA was removed, PDA was divided just proximal to its junction with the DTA and DTA was anastomosed to the Arch

of Aorta. The posterior layer of the ascending aorta and DTA was sutured directly while the anterior layer required an autologous fixed pericardium to create a wide anastomosis such that the post-operative systolic gradient was 5mmHg (Figure 4). Tricuspid and Mitral Valve were found to have normal anatomy and were competent on the saline injection test. A small PFO was left open. Aortic cross-clamp time and CPB time were 86 and 112 minutes respectively. Post-operative TEE did not reveal any MR or TR with mild LV dysfunction. The child is currently doing well in the follow-up clinic.

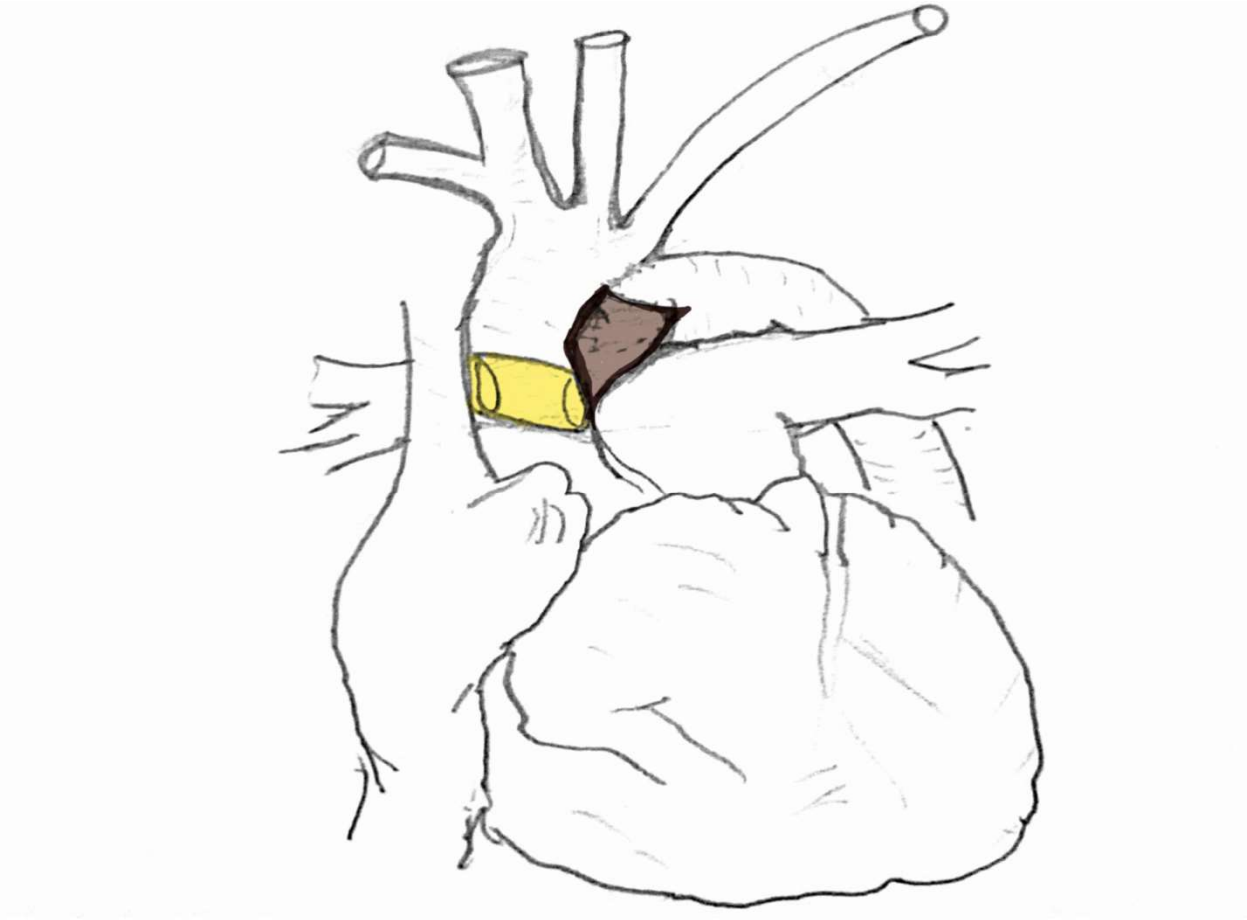

Figure-4-Post-operative Schematic diagram showing PTFE patch baffling of RPA to MPA (long arrow) and pericardial patch augmentation of aorta (short arrow).

Discussion

APW is a rare disorder accounting for up to 0.2% of all congenital heart disease. Mori classified APW into proximal, distal and total according to its location. Congenital Heart Surgery database Committee recommended the terms type I- proximal defect, type II- distal defect, type III- total defect, and intermediate defect. It is most commonly associated with IAA which can be attributed to the decreased blood flow in the aortic isthmus during prenatal period.[7]

IAA is a rare congenital malformation that requires early surgical treatment. IAA has been classified into 3 groups, by the Celoria and Patton classification, according to the position of interruption: In type A (43%), the interruption is distal to the left subclavian artery, as in our case; In type B (53%), the interruption is between the left common carotid and left subclavian arteries; In type C (4%), the interruption is between the innominate and left common carotid arteries.[8]

In 1982, Berry and colleagues described the association of absence of the aortopulmonary septum, aortic origin of the right pulmonary artery, intact ventricular septum, patent ductus arteriosus, and hypoplasia of the aortic isthmus. They used different techniques in their patients including RPA detachment and reimplantation into MPA, tube graft interposition between the aortic annulus to the proximal arch. Subclavian artery and Dacron graft were also used to repair the IAA.[1]

Ding et al. reported the repair of the anomaly by a different technique wherein they used a Dacron graft to baffle the RPA to MPA through the AP window and direct repair of IAA. The graft was fashioned in such a way to create an intra-aortic baffle to direct the blood directly from MPA to RPA. Post repair PA pressures were less than half the systemic pressures.[7]

Burke et al detached the RPA from the ascending aorta and anastomosed it with the MPA anteriorly whereas the APW was divided completely and the defect on each side was closed separately with the pericardium. They suggested that an intra aortic baffle may partially obstruct the left ventricular outflow and a small baffle may compromise the flow to RPA.[9]

Boonstra et al reported a different technique where the aortopulmonary window was closed transversely by direct suture, creating a tunnel posterior to this suture that became the origin of the right pulmonary artery, now originating directly from the main pulmonary artery. They also suggested that anastomotic stenosis between the aortic arch and descending aorta is a potential complication.[10]

In our case, a 7-week old child presented with features of cardiac failure. The patient was managed with diuretics and antibiotics. The patient got intubated on admission and was diagnosed to be a case of an APW with IAA. Poor cardiac function added to the critical condition of the patient. The patient was stabilized further and taken up for corrective repair. We repaired the type III AP window via the trans aortic approach and the RPA was baffled towards MPA by a PTFE patch. Type A IAA was corrected by an end to side anastomosis of the DTA to arch of aorta with an autologous glutaraldehyde treated pericardial patch augmentation of the anastomosis. Postoperative TEE revealed good flow in bilateral pulmonary arteries as well as in the arch and good biventricular function with no MR. Later 2D TTE also revealed similar findings.

Conclusion

Berry syndrome is a rare congenital cardiac anomaly. Patients present with congestive cardiac failure. One-stage surgical correction has demonstrated acceptable outcomes and is now considered the procedure of choice. Early surgical treatment is however indicated to prevent the complications of pulmonary hypertension. Various surgical strategies have been described to correct the anomaly. We used an elliptical PTFE patch to baffle RPA and LPA towards the MPA and an autologous treated pericardium to reconstruct the aortic interruption. Shorter aortic clamp time and adequate intra-operative myocardial protection are essential to prevent post-operative cardiac dysfunction.

Abbreviations

IAA - Interrupted Aortic Arch, APW - Aortopulmonary Window, PTFE - Poly Tetrafluro Ethylene, RPA - Right Pulmonary Artery, LPA - Left Pulmonary Artery, MPA - Main Pulmonary Artery, PDA - Patent Ductus Arteriosus, DTA - Descending Thoracic Aorta, MR - Mitral Regurgitation, TR - Tricuspid Regurgitation, TEE - Trans Esophageal Echocardiography.

Conflicts of interest

The authors have no conflict of interest to declare.

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