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Case Report Open Access

Berry syndrome: A rare aortopulmonary malformation

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ABSTRACT

Herein, we present a case of Berry syndrome with successful surgical repair in an infant. A definitive diagnosis was established with appropriate investigative modalities, and a single-stage repair was performed with good results.

Keywords: Aortopulmonary window, Berry syndrome, interrupted aortic arch.

Berry syndrome is an extremely rare aortopulmonary malformation accounting for 0.046% of congenital heart defects.[1] First illustrated by Berry et al.[2] in 1982, it consists of a distal aortopulmonary window (APW), an aortic origin of the right pulmonary artery, an intact ventricular septum, and patent ductus arteriosus (PDA) with hypoplastic or interrupted aortic arch (IAA). Surgery remains the mainstay of treatment. The choice of a single or staged approach is still controversial.[1,3] Herein, we present a case of Berry syndrome with successful surgical repair in an infant.

CASE REPORT

A nine-month-old female child weighing 5 kg presented to our clinic with failure to thrive and recurrent respiratory tract infections. The cardiovascular system examination revealed normal first and second heart sounds. A grade 3/6 continuous murmur was heard in the left second intercostal space.

Chest X-ray revealed cardiomegaly (cardiothoracic ratio: 0.68) and a right ventricular type of apex with plethoric lung fields. Electrocardiography demonstrated a normal sinus rhythm, a heart rate of 145/min, right axis deviation, incomplete right bundle branch block, right ventricular hypertrophy, and left ventricular volume overload. Transthoracic echocardiography (TTE) showed a large APW (1.2 cm) with a bidirectional shunt (Figure 1a), moderate tricuspid regurgitation, and features of pulmonary artery hypertension (PAH). Right ventricular systolic pressure (RVSP) was 97 mmHg, with normal right ventricular function and a left ventricular ejection fraction of 60%. Cardiac catheterization study was performed, which revealed a large APW of 1.6 cm (Figure 1b), arising from the ascending aorta, and a large PDA of 8 mm, supplying the descending thoracic aorta with type B IAA. The ratio of pulmonary blood flow to systemic blood flow (Qp/Qs) was 3.93, and the pulmonary vascular resistance index was 2.81 post oxygen administration.

Intraoperative anatomy was assessed. Type III APW[4] with type B IAA was noted (Figure 2a). Patent ductus arteriosus was observed supplying the descending aorta, and the right pulmonary artery was arising from ascending aorta (Figure 2a).[5]

Cardiopulmonary bypass was established with the high ascending aorta and bicaval venous cannulation. Aorta was cross-clamped, the core was cooled to 18°C, and antegrade cold blood cardioplegia was administered. After satisfactory arrest, the right atrium was opened, and a patent foramen ovale (PFO) was created to vent the left side of the heart. An APW was vertically opened, and the anatomy was noted. The aorta and the main pulmonary artery (MPA) were separated. Once the nasopharyngeal

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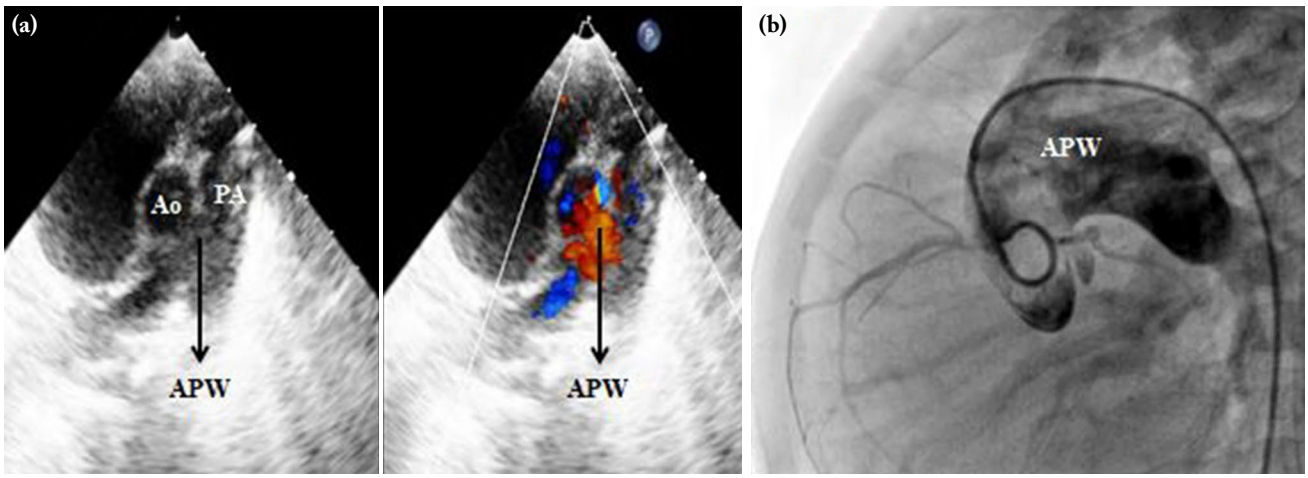

Figure 1. **(a)** Two-dimensional echocardiography with color Doppler in short axis view shows echo dropout between the aorta and the pulmonary artery indicating APW. **(b)** Catheterization study (catheter course from descending aorta -PDA-through APW to ascending aorta) demonstrating an APW.
Ao: Aorta; APW: Aortopulmonary window; PA: Pulmonary artery.

temperature reached 18°C, total circulatory arrest was initiated. The PDA was ligated, and the proximal stump was transfixed. The distal end of the PDA and remaining ductal tissue was excised. The descending aorta was mobilized (Figure 2b). The left subclavian artery was ligated. The descending aorta was pulled up and anastomosed to the arch of the aorta, and the anterior wall was augmented with autologous untreated pericardium with 6-0 polypropylene sutures. The RPA was disconnected from the ascending aorta, and the proximal ascending aorta was augmented with autologous untreated pericardium. The arterial cannula was reinserted, and rewarming was started. Dearing was done after rewarming. The RPA was anastomosed to the MPA near its bifurcation with 6-0 polypropylene sutures and anteriorly augmented with autologous untreated pericardium (Figure 2c). Tricuspid valve repair was done through the right atrium. The PFO was closed with 5-0 polypropylene sutures, leaving behind a small PFO. The final image after the surgical closure is presented in Figure 2d. The patient was in sinus rhythm after cardiopulmonary bypass with milrinone at 0.5 minimum inhibitory concentration (MIC) and adrenaline at 0.05 MIC and was shifted to the intensive care unit with stable hemodynamics.

The patient was extubated on the first postoperative day and was doing well. She had to be reintubated on the second postoperative day for cardiorespiratory arrest due to a PAH crisis (RVSP=70 mmHg). The PAH crisis was conservatively managed with milrinone infusion, sildenafil, and hyperventilation. The patient was extubated on the 13th postoperative day. In view of low saturation with poor respiratory efforts and left lung collapse consolidation, the patient required prolonged continuous nasal positive airway pressure support, which was gradually weaned off. The rest of the postoperative period was uneventful.

Echocardiography at discharge demonstrated aortic arch continuity without any gradient, unobstructed RPA flow, pulsatile flow in the abdominal aorta, mild tricuspid regurgitation, RVSP of 38 mmHg, good biventricular function, left ventricular ejection fraction of 60%, and no residual APW.

On the last follow-up at one year, the patient was asymptomatic with TTE findings correlating with discharge TTE.

DISCUSSION

Berry syndrome is a rare and complex malformation requiring prompt diagnosis and well-planned, timely surgical treatment to restore normal circulation distal to the IAA supplying the lower part of the body and for the prevention of pulmonary vascular obstructive disease.[1,5]

The clinical presentation is an excessive left-to-right shunt, and most patients present early in life. The development of pulmonary hypertension

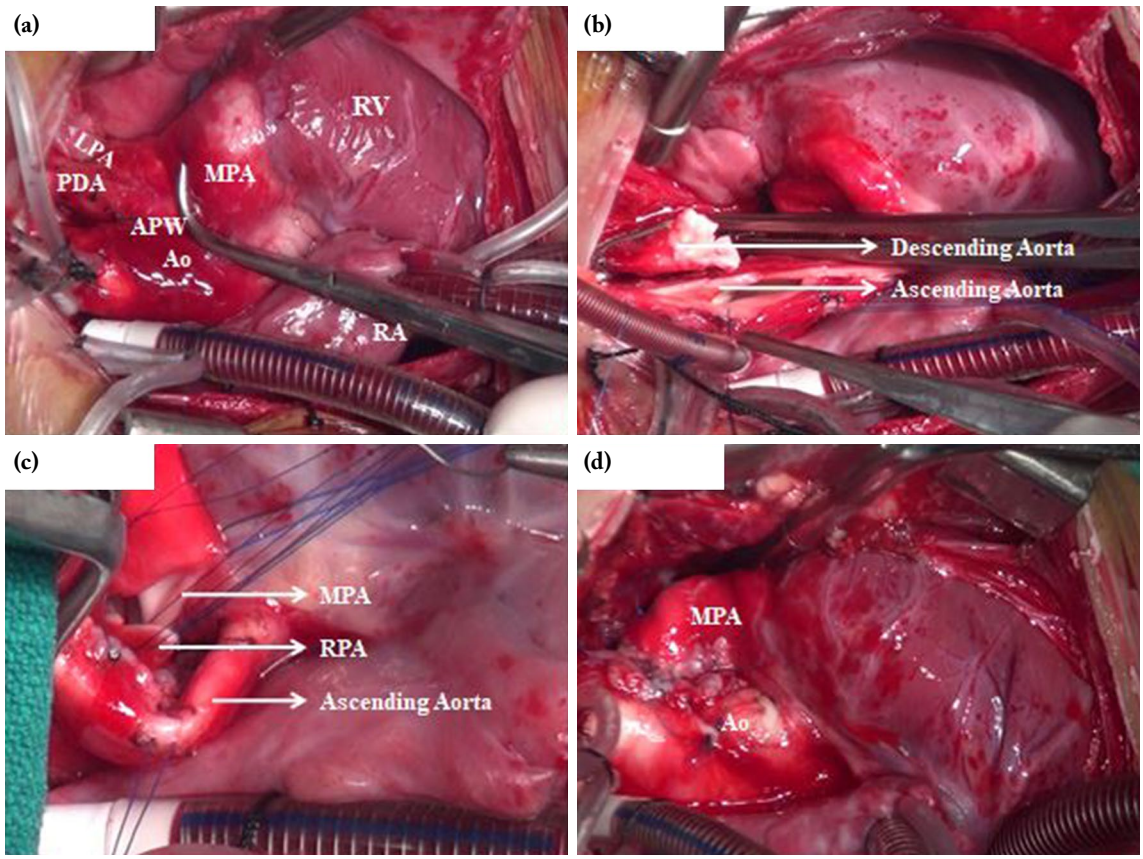

Figure 2. Operative image displaying **(a)** a large-sized APW and PDA; **(b)** mobilized descending aorta and ascending aorta; **(c)** anastomosis of the RPA to MPA; **(d)** the final picture after closure.

RV: Right ventricular; LPA: Left pulmonary artery; MPA: Main pulmonary artery; PDA: Patent ductus arteriosus; APW: Aortopulmonary window; Ao: Aorta; RPA: Right pulmonary artery.

and pulmonary vascular resistance is usually rapid. Fetal echocardiography and TTE are the mainstay of diagnosis in the prenatal and postnatal periods, respectively.[6] Cardiac catheterization provides anatomic details and assesses for pulmonary vascular obstructive disease.[1,2,5]

Single-stage surgery in the neonatal period and infancy is a widely accepted approach, although Ghelani et al.[6] suggested a staged repair in premature infants who present with excessive pulmonary shunts and congestive cardiac failure. In this category of patients, staged repair involves surgical pulmonary artery banding, followed by definitive surgical repair. Single-stage repair includes APW closure, anastomosis of the RPA to MPA, and maintaining the aortic arch continuity.[1] Postoperative PAH crisis is a known phenomenon that can be managed with pulmonary vasodilator therapy, in our case with milrinone infusion and sildenafil.[3]

In conclusion, Berry syndrome is an extremely rare aortopulmonary malformation that can be definitively diagnosed with prenatal fetal echocardiography and neonatal TTE, which warrants prompt and early surgical management. Surgical management can be tailored according to the patient presentation as staged or single-stage procedures. Postoperative PAH crisis is a known complication that can be successfully managed with pulmonary vasodilator therapy in most of the cases.

Patient Consent for Publication: A written informed consent was obtained from patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Idea, design, data collection and interpretation, literature review, writing the article, references, materials: M.W.A.; Data collection and processing: P.G.; Design, supervision, analysis, literature review, writing the article: S.M.H.; Supervision, critical review: N.D.

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