

Berry syndrome: One stage surgical repair in a neonate

Somia Razzaq, Nadeem Aslam, Waris Ahmad, Muneer Amanullah

ABSTRACT

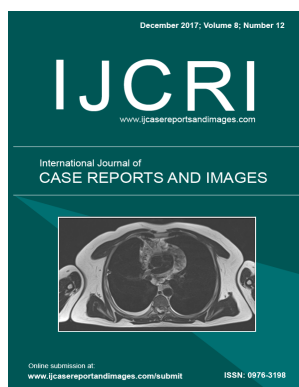
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Conclusion: Early clinical recognition with prompt surgical correction is associated with good outcomes.



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CASE REPORT

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Introduction: Interruption of aortic arch, aortopulmonary septal defect, patent ductus arteriosus and anomalous origin of right pulmonary artery is a rarely encountered constellation of anomalies. **Case Report:** We present a case of a 14-day-old male neonate with persistent respiratory distress. Echocardiogram and computed tomography angiography confirmed the constellation of Berry syndrome. A one stage surgical correction was successfully performed. Hypoplastic aortic arch was reconstructed using end to end anastomosis. Aortopulmonary septal defect was closed using single patch technique and the right pulmonary artery was re-implanted into the main pulmonary artery. **Conclusion:** Early clinical recognition with prompt surgical correction is associated with good outcomes.

Keywords: Anomalous right pulmonary artery, Aortopulmonary window, Berry syndrome, Interrupted aortic arch

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INTRODUCTION

Berry et al. [1] in 1982 described the constellation of distal aortopulmonary septal defect, aortic origin of right pulmonary artery, interruption of aortic arch, intact ventricular septum and patent ductus arteriosus as a syndrome (Figure 1). Aortopulmonary window has been classified based on the location of the septal defect [1, 2]. Type I comprises proximal communication with normal origin of pulmonary arteries. Type IIA consists of distal defect with straddling right pulmonary artery that originates from the aortopulmonary communication but maintains its continuity with the left pulmonary artery. Type IIB consists of distal communication with origin of right pulmonary artery solely from the aorta. Type III consists of multiple aortopulmonary communications. Of all the published reports Berry syndrome has not been found to have been associated with chromosomal abnormalities except in one patient with trisomy 13 [3].

Prenatal diagnosis of this rare complex congenital anomaly is related with improved outcomes due to prompt initiation of prostaglandin infusion [4]. Detailed anatomic depiction using echocardiogram and other diagnostic

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modalities with early surgical repair is essential, thereby avoiding pulmonary hypertension associated with pulmonary over circulation and subsequent heart failure [5–7].

A number of reports in literature have described the utilization of either one-stage or two-stage surgical correction of this complex congenital cardiac anomaly. We describe a case of successful one-stage surgical repair in a neonate.

CASE REPORT

A male baby was born by cesarean section with a birth weight of 4.2 kg at 37th week of gestation. Shortly after birth, the neonate developed respiratory distress. The perinatal history was unremarkable. On physical examination the baby was tachypneic with a respiratory rate of 65 breaths/min and heart rate of 136 beats/min. The blood pressures of both arms were equal. The blood pressures of the lower limbs were lower by 15 mmHg. Differential cyanosis was detected by pulse oximetry measurement with upper limb oxygen saturation of 94% and lower limb oxygen saturation of 87%. On auscultation a grade 3/6 systolic murmur was heard at the left sternal border. Femoral pulses were barely palpable. Chest X-ray demonstrated borderline cardiomegaly with subtle pulmonary plethora. Transthoracic echocardiogram revealed a Type A interrupted aortic arch, a 1.5-mm patent ductus arteriosus, a 12-mm Type IIB aortopulmonary window and possibility of anomalous origin of right pulmonary artery from descending aorta (Figure 2). Doppler study showed supra systemic pulmonary artery and right ventricular pressures. A computed tomography angiography confirmed the presence of interrupted aortic arch, anomalous origin of right pulmonary artery from the proximal part of right aortic wall and a patent ductus arteriosus which continued as descending aorta (Figure 3A–C). Prostaglandin infusion was initiated to maintain patency of ductus arteriosus.

Operative Procedure

On the 14th day of life the neonate underwent one-stage surgical correction. The heart was exposed via a median sternotomy. The anatomy was recognized to be consistent with constellation of Berry syndrome (Figure 4). Extensive dissection was done in order to mobilize the arch vessels, ascending and descending aorta and patent ductus arteriosus. The pulmonary arteries were dissected well into the hilar branches. After heparanization, arterial cannula was placed below the innominate artery to leave enough space above the aortopulmonary window to place the aortic cross clamp. A single venous cannula was placed in the right atrium. Cardiopulmonary bypass was initiated and the patient was cooled to 18°C. Aortic cross clamp was applied and crystalloid cold blood cardioplegia was infused. The pulmonary arteries were snugged.

Deep hypothermic circulatory arrest was established. Aortic arch vessels were snugged. Aortic cross clamp was removed. The ductus arteriosus was divided and transected (Figure 5A). All ductal tissue was excised until normal aortic tissue. The aortic arch was incised and an end-to-end anastomosis was constructed using 7-0 polypropylene (Figure 5B). Anterior aspect of the aortic anastomosis was augmented with bovine pericardial patch (Figure 5C). Aortic cross clamp was reapplied and aortic arch vessels were unsnugged. Cardiopulmonary bypass was reinstituted and a second dose of cardioplegia was infused. During the rewarming period a vertical incision over the aortopulmonary window was made and the septal defect was closed using bovine pericardial patch with a 6-0 polypropylene. Aortic cross clamp was removed. The patient spontaneously converted to sinus rhythm.

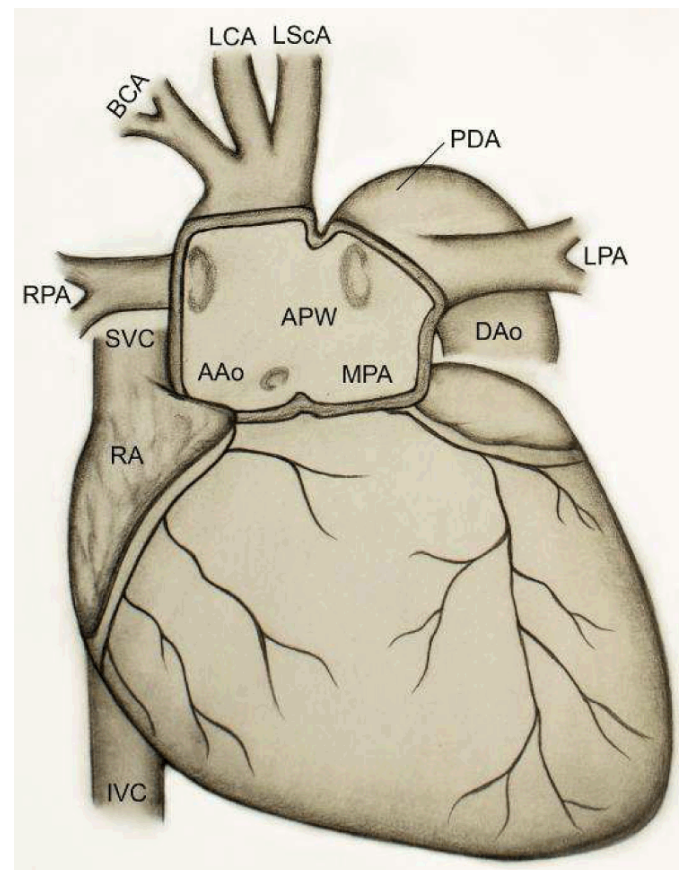


Figure 1: Berry syndrome consisting of aortopulmonary septal defect, interrupted aortic arch, patent ductus arteriosus and aortic origin of the right pulmonary artery.

Abbreviations: AAo: Ascending Aorta, APW: Aortopulmonary Window, BCA: Brachiocephalic Artery, DAo: Descending Aorta, IVC: Inferior Vena Cava, LCA: Left Carotid Artery, LPA: Left Pulmonary Artery, LScA: Left Subclavian Artery, MPA: Main Pulmonary Artery, PDA: Patent Ductus Arteriosus, RA: Right Atrium, SVC: Superior Vena Cava.

Partial occluding clamp was applied to the aorta and the right pulmonary artery was excised from the aorta. The aortic defect was closed with 6-0 polypropylene. The right pulmonary artery was anastomosed to the right lateral aspect of the main pulmonary artery with 6-0 polypropylene hence obtaining right pulmonary artery to main pulmonary artery continuity (Figure 5D). The patient was easily weaned off from cardiopulmonary bypass with minimal inotropic support. The total cardiopulmonary bypass time was 140 minutes, with a cross clamp time of 80 minutes. The circulatory arrest time was 46 minutes. Elective delayed sternal closure was performed 36 hours later.

The patient was extubated on postoperative day-6 and discharged on postoperative day-12. Postoperative echo and Doppler study demonstrated patched aortopulmonary window with no significant residual defect, mild turbulence in the right pulmonary artery with pressure gradient of 20 mmHg, mild tricuspid regurgitation with maximum gradient of 18 mmHg.

DISCUSSION

Berry syndrome is a very rare complex congenital cardiac anomaly that is amenable to surgical repair. The pathogenesis of this anomaly is unknown, however, proposed hypothesis states that the failure of formation of aortopulmonary septum leads to anomalous origin of pulmonary arteries from the undivided truncal segment. This anomalous origin of the pulmonary arteries induces a steal phenomenon hence reducing aortic blood flow during embryogenesis resulting in hypoplasia of the aortic arch [1].

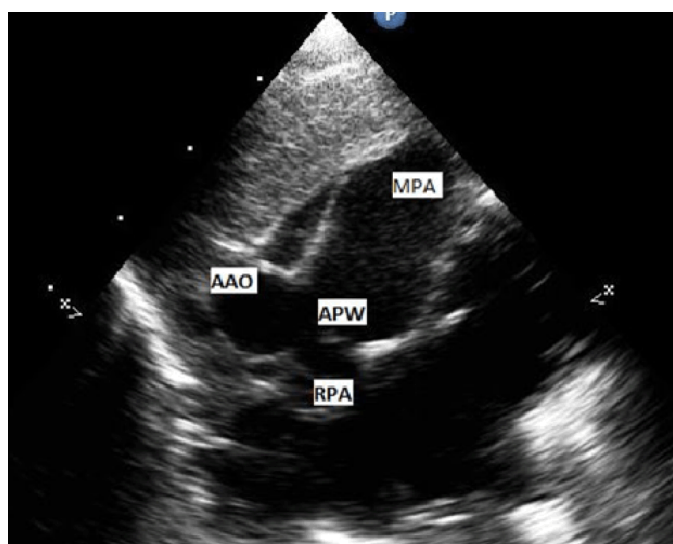


Figure 2: Echocardiography parasternal short axis view showing aortopulmonary window and anomalous origin of right pulmonary artery from ascending aorta.

Abbreviations: AAO: Ascending Aorta, APW: Aortopulmonary Window, MPA: Main Pulmonary Artery, RPA: Right Pulmonary Artery

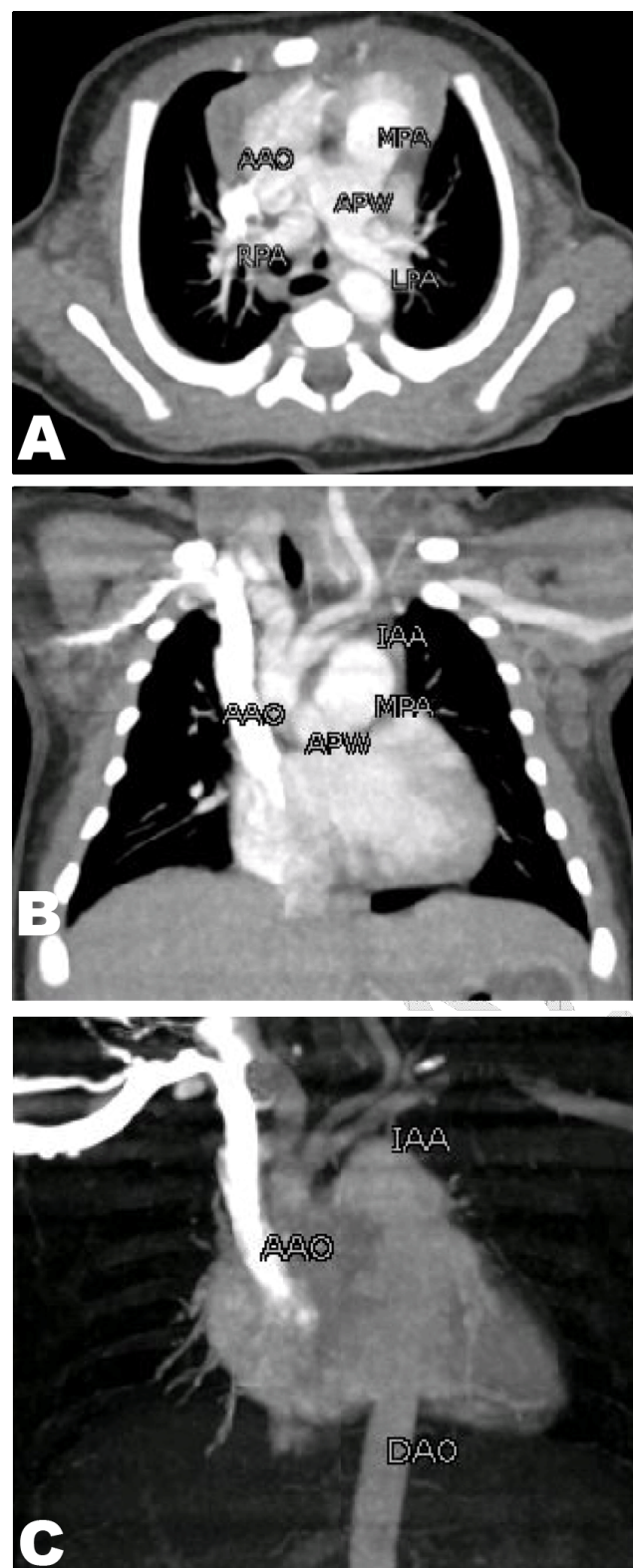


Figure 3: Computed tomography angiography (A) Axial view showing aortopulmonary window and anomalous origin of right pulmonary artery from ascending aorta, (B) Aortopulmonary window and interrupted aortic arch, and (C) 3D reconstructed image showing interrupted aortic arch.

Abbreviations: AAO: Ascending Aorta, APW: Aortopulmonary Window, LPA: Left Pulmonary Artery, MPA: Main Pulmonary Artery, RPA: Right Pulmonary Artery, DAO: Descending Aorta, IAA: Interrupted Aortic Arch

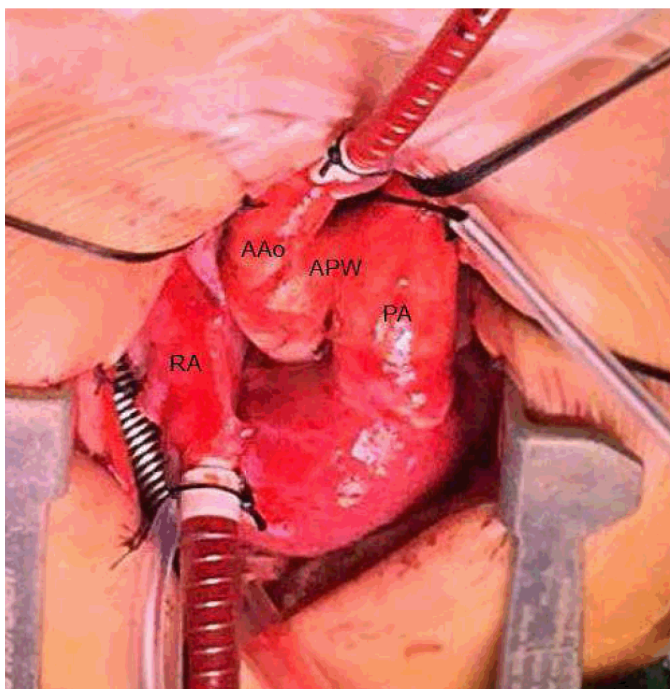


Figure 4: Intraoperative finding confirming the presence of constellation of Berry syndrome.

Abbreviations: AAo: Ascending Aorta, APW: Aortopulmonary Window, PA: pulmonary artery, RA: Right Atrium

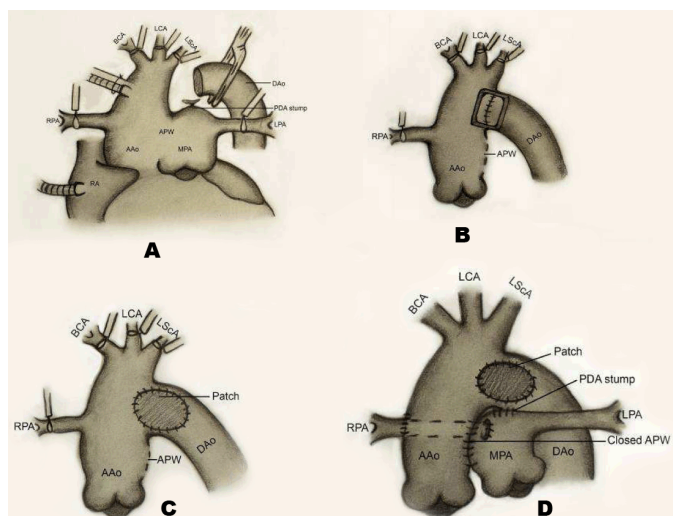


Figure 5: (A) After Arterial and venous cannulation the great arteries and pulmonary arteries were snugged. The ductus arteriosus was transected and ligated, (B) The aortic arch was incised and an end-to-end anastomosis was made with the posterior wall of the descending aorta, (C) The anterior aspect of the aortic anastomosis was augmented with bovine pericardial patch, and (D) The complete repair with anastomosis of descending aorta to the ascending aorta and the right pulmonary artery anastomosed to the main pulmonary artery running posterior to the aorta.

Abbreviations: AAo: Ascending Aorta, APW: Aortopulmonary Window, BCA: Brachiocephalic Artery, DAo: Descending Aorta, LCA: Left Carotid Artery, LPA: Left Pulmonary Artery, LSca: Left Subclavian Artery, MPA: Main Pulmonary Artery, PDA: Patent Ductus Arteriosus

Berry syndrome has also been reported to be associated with anomalous origin of left coronary artery from the main pulmonary artery [8]. Patients with Berry syndrome present with respiratory distress, metabolic acidosis, anuria, severe congestive heart failure or acute cardiovascular collapse with spontaneous closure of ductus arteriosus in the first few days of life. Early clinical diagnosis with detailed preoperative recognition of these unique anatomic features is therefore essential for good surgical outcomes [2, 3, 5–7]. Long-term follow-up is required after surgical correction as stenosis at the site of anastomosis is a potential complication. The stenosis can be relieved adequately by percutaneous balloon angioplasty [9, 10].

Surgical repair can be done using either two-stage or one-stage approach. Two-stage repair involves reconstruction of aorta with ligation of patent ductus arteriosus followed by complete closure of aortopulmonary window in the next stage [11]. Ghelani et al. advocate the utilization of two staged surgical repair in case of premature infants and small for gestational age infants [4]. However, one stage surgical repair is considered superior, the rate of reoperation varies with development of complications.

Berry et al. reported eight cases out of which two patients underwent successful one-stage surgical repair, with reconstruction of aortic arch using either left subclavian or Dacron graft, closure of aortopulmonary window and reimplantation of right pulmonary artery to the main pulmonary artery [1].

Ding et al. reported a case of one-stage repair using Dacron baffle to obliterate the aortopulmonary septal defect and maintain continuity of right pulmonary artery with the main pulmonary artery with repair of hypoplastic aortic arch using end-to-end anastomosis [12]. The utilization of intra-aortic baffle is associated with obstruction of right pulmonary artery and left ventricular outflow tract. The size and growth potential of the baffle also pose as a problem to be utilized successfully in neonates [9].

Re-routing of right pulmonary artery by making a tunnel, either by suturing of native aortic wall tissue [13, 14] or utilization of glutaraldehyde fixed autologous pericardial patch [7] with simultaneous closure of aortopulmonary window is another reported successful surgical technique. Stenosis at the site of repair is a major complication of this method of repair [9].

Right pulmonary artery arterioplasty with aortic cuff formation is another method of repair in case of Berry syndrome as utilized by Abbruzzese et al. [15]. In this case, the aorta has to be transected which is associated with increased risk of bleeding along with difficult anastomosis [9].

Burke et al. [16] reported a case of successful arterial switch for the repair of Berry syndrome in a neonate. The aorta was separated from the pulmonary artery. The right pulmonary artery was anastomosed to the main pulmonary artery after being excised completely from

the right lateral aspect of the aorta. The descending aorta was connected via end-to-end anastomosis to the defect in the ascending aorta. This technique demonstrated easy anterior transfer of pulmonary arteries in neonates. This leads to decreased risk of compression of right pulmonary artery by the aorta and the airway [9].

Park et al. in 2008 [10] demonstrated two cases of single stage repairs. In this technique, the posterior wall of the ascending aorta was utilized to form confluence between the right pulmonary artery and the main pulmonary artery. The anterior half of the right pulmonary artery was repaired with an autologous pericardial patch. Utilization of this patch technique avoids undue tension and stenosis at the site of anastomosis.

We have implemented a surgical technique that demonstrates the reconstruction of hypoplastic aortic arch by end-to-end anastomosis with patch augmentation of anterior aortic wall. This eliminates the risk of postoperative aortic arch stenosis henceforth decreasing the risk of proximal segment dilation and compression of right pulmonary artery and the left main bronchus [9]. The detachment and anastomosis of the right pulmonary artery to main pulmonary artery behind the aorta maintains the geometry of the pulmonary arteries. This technique is preferred in neonates due to potential growth problems associated with patch repair or the utilization of intra-aortic baffles, thereby also reducing the risk of left ventricular outflow tract obstruction [9, 12, 16, 17].

CONCLUSION

Berry syndrome is a rare congenital anomaly. Echocardiography in an important diagnostic tool in the initial evaluation but computed tomography angiogram or cardiac magnetic resonance imaging scan is frequently required. One-stage surgical correction has demonstrated acceptable outcomes and is now considered the procedure of choice. Stenosis at the site of anastomosis is commonly encountered. Reoperations are mainly due to development of aortic or right pulmonary artery stenosis.

Author Contributions

Somia Razzaq – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Nadeem Aslam – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Waris Ahmad – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Muneer Amanullah – Substantial contributions to conception and design, Acquisition of data, Analysis and

interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

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None

Conflict of Interest

Authors declare no conflict of interest.

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