CASE REPORT

Berry syndrome: One-Stage Surgical Repair

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Berry Syndrome is characterized by the sum of several abnormalities; namely, aortopulmonary window, aortic origin of the right pulmonary branch, interruption of the aortic arch and intact interventricular septum. First described by Berry et al. in 1982. We present the case of a 21-day-old infant weighing 3.3kg, with a progressive increase in respiratory effort, vomiting, and irritability. Complementary studies were carried out reporting the anomalies that make up the Berry Syndrome.

Key words: Aortopulmonary window; Anomalous implantation of the right pulmonary artery; Berry syndrome; Congenital malformation; Interruption of the aortic arch; 3D vascular reconstruction.

El síndrome de Berry se caracteriza por la suma de varias anomalías; a saber, ventana aortopulmonar, origen aórtico de la rama pulmonar derecha, interrupción del arco aórtico, y septum interventricular íntegro. Fue descrito por primera vez por Berry y cols. en 1982. Presentamos el caso de un infante de 3.3 kg de peso, con con aumento progresivo del esfuerzo respiratorio, vómito, e irratibilidad. Los estudios complementarios llevados a cabo demostraron las anomalías que constituyen el Síndrome de Berry.

Palabras clave: Ventana aortopulmonar; Implantación anómala de la rama derecha de la arteria pulmonar; Síndome de Berry; Malformación congénita; Interrupción del arco aórtico; Reconstrucción vascular en 3D.

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Berry syndrome is characterized by the sum of several abnormalities: aortopulmonary window; aortic origin of the right pulmonary branch (ROAAPD); interruption of the aortic arch (IAA) and intact interventricular septum. First described by Berry et al. in 1982 [1,2], it is an extremely rare pathology with an incidence of 0.04% reported in the literature [3,4]. Timely treatment after birth is very important because most patients die shortly after birth, while surviving patients will develop pulmonary hypertension rapidly. This complex anomaly has been reported in around 100 patients [1]. Mortality rate was 100%, and the median age at death was one month [3,4]. Therefore, it is a challenge for the cardiovascular team to perform an early diagnosis and successful surgical correction in the neonatal period.

We present the case of a 21-day-old neonate with the aforementioned characteristics, treated in a single surgical procedure at our institution.

CLINICAL CASE

A 21-day-old infant with no maternal history, weight 3.3 kg, presented 10 days before admission, increased work of breathing with intercostal retractions, irritability, vomiting of gastric content, and generalized pallor.

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He was assessed by the pediatric cardiology service showing evident dyspnea, tachypneic, with left parasternal grade III ejection murmur and generalized pallor with SaO2 92%, RR 70 rpm, HR 111 bpm, right upper limb blood pressure 50/38 and right lower limb 54 /36, decreased pulses in lower extremities. In the admission, chest x-ray with levocardia, levoapex, bronchial situs solitus and increased pulmonary flow were observed, a cardio-thoracic ratio of 0.64. Advanced airway management was required.

The echocardiographic study (**Fig. 1**) (**Fig. 2**) showed atrial and abdominal situs solitus, levocardia, levoapex, lateralized systemic and pulmonary venous returns, concordant perforated mode atrioventricular connection, perforated mode concordant ventricle-arterial connection, presence of a 2mm patent foramen ovale with shunt from left to right, intact interventricular septum. Site of communication of the ascending aorta with the trunk of the pulmonary artery of 7.2 x 9.7 mm with shunt from left to right, located 12 mm away from the pulmonary valve, reporting type II pulmonary aortic window, non-confluent pulmonary branches, the right pulmonary branch arises from the ascending aorta, the left pulmonary branch arises from the trunk of the pulmonary artery. Aortic arch to the left with interruption of the type A arch distal to the subclavian artery and flow of the ductus arteriosus with flow from right to left. Systolic function of the right ventricle with TAPSE of 9.6 mm (Z score +0.43) area shortening fraction of 32.6%. Left ventricular systolic function with biplanar LVEF of 58%.

The computed tomography (**Fig. 3**) reported dilatation of the aortic ring, at the level of the ascending aorta, 15.7 mm

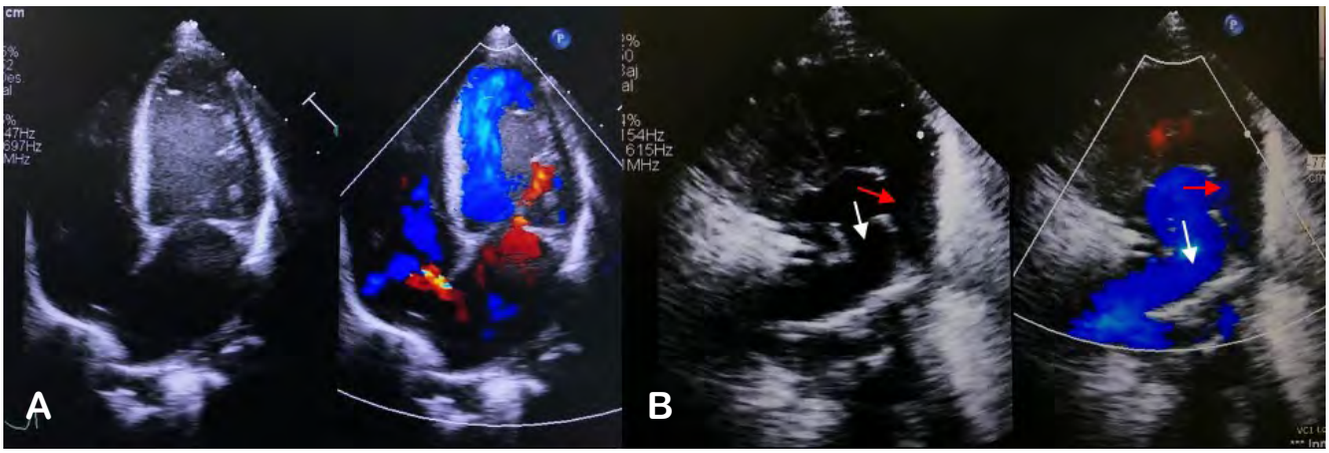

Figure 1.Two-dimensional transthoracic echocardiogram. Two-dimensional transthoracic echocardiogram: a) 4-chamber axis with patent foramen ovale with shunt from left to right, intact interventricular septum, and mild mitral regurgitation. B) axis of the great vessels showing the aortopulmonary window (red arrow) and the origin of the right pulmonary branch of the ascending aorta (white arrow).

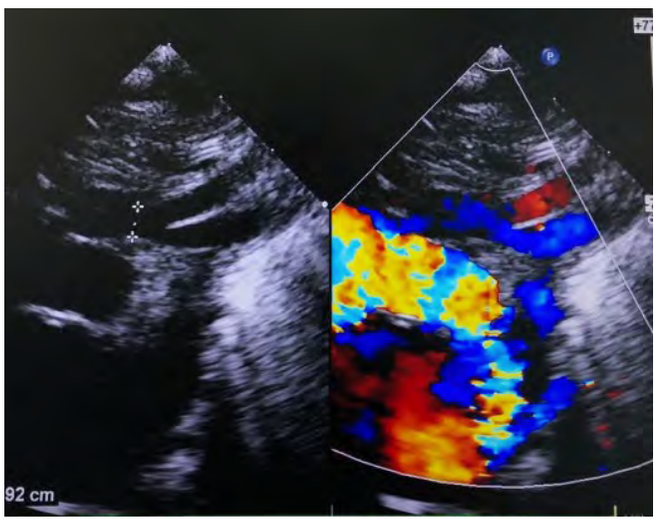

Figure 2. Two-dimensional transthoracic echocardiogram showing interruption of the aortic arch after emergence of the subclavian artery (white arrow) and the presence of the ductus arteriosus that reaches the pulmonary artery.

from the valvular plane, presence of aortic-pulmonary window (type II), the right pulmonary branch originating from the posterior wall of the ascending aorta to 12 mm from the valvular plane, interruption of the type A aortic arch, dilated pulmonary trunk.

Concluding as Berry Syndrome, emergency surgical correction was performed in a single stage through a median sternotomy (**Fig. 4**) under extracorporeal circulation. Moderate hypothermia was used for 96 minutes and selective cerebral perfusion for 14 minutes. Surgical Section and suture of patent ductus arteriosus was performed. Direct closure of the aortopulmonary window, aortic advancement and reimplantation of the right pulmonary artery in the trunk of the pulmonary artery were also performed. Weaning from extracorporeal circulation was successful at the first attempt. Leaving with deferred sternal closure, performing the definitive closure at 48 hours. He was progressed favorably in intensive care, extubation after 10 days, stay in intensive care for 14 days and in-hospital discharge was after 22 days.

Postoperative echocardiographic study reported absence of any obstructive gradient at the level of the aortic advance anas-

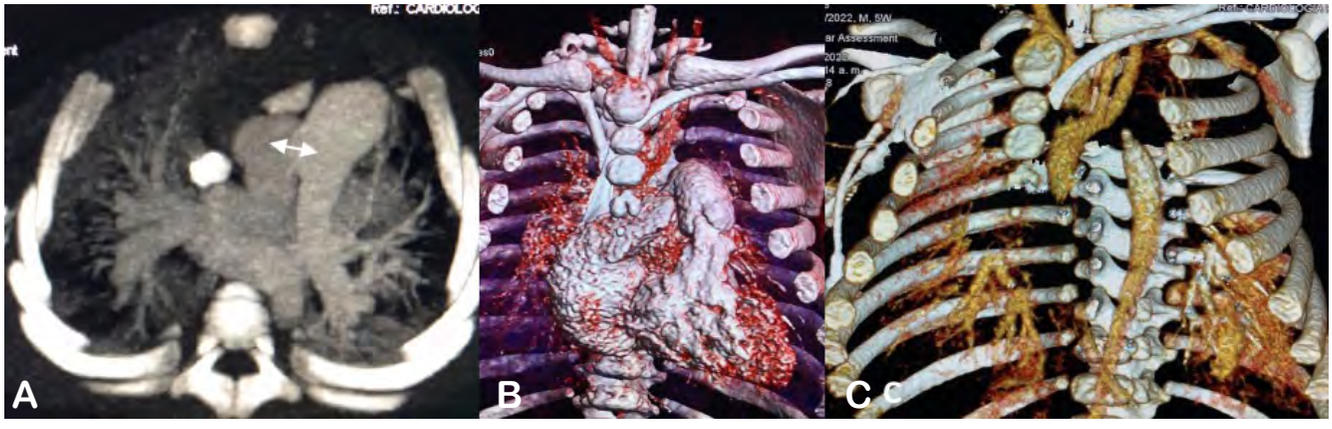

Figure 3. CT Angiography of the heart and great vessels; a) axial cut showing aortopulmonary window (white arrow), b) dilated 3D pulmonary reconstruction, c) reconstruction of the aortic arch showing interruption of the type A aortic arch, after emergence of the subclavian artery.

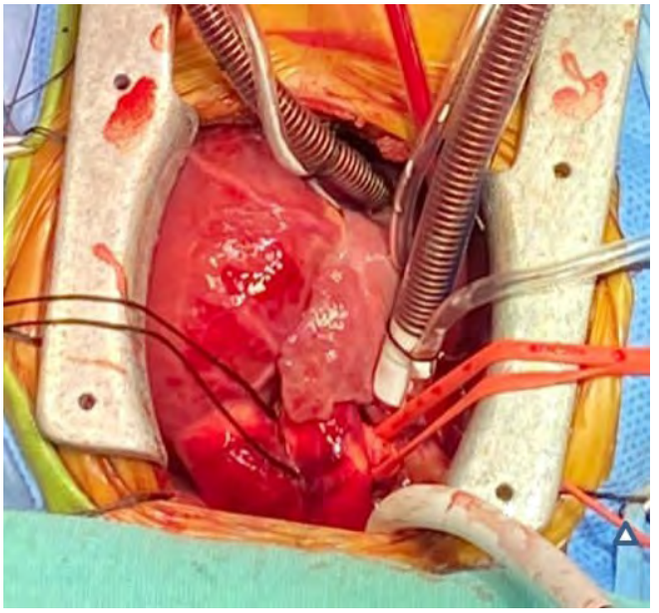

Figure 4. Intraoperative imaging. Presence of the aortopulmonary window (*), and the anomalous origin of the right pulmonary branch from the lateral aspect of the ascending aorta( )

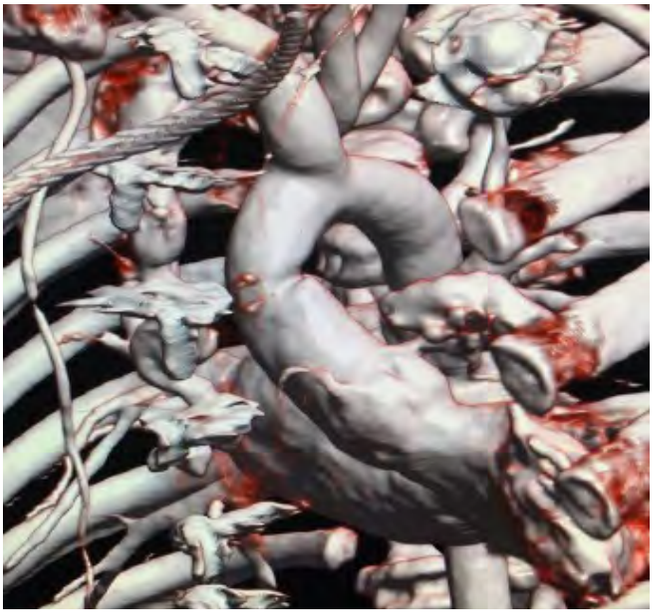

Figure 5. Postoperative CT scan. Post-surgical control at 5 months, observing adequate continuity of the aortic arch.

tomosis, but at the level of the right branch of the pulmonary artery with a maximum gradient of 10 mmHg, and a mean gradient of 5 mmHg without hemodynamic repercussion. In chest CT scan, it was possible to visualize adequate continuity of the aortic arch (**Fig. 5**).

COMMENT

Berry Syndrome is a rare heart disease, being a challenge for the echocardiographer and cardiac surgeon for the resolution of this anomaly in infants who are critically ill, with the need for urgent surgery.

Berry et al. [5] described the presence of interruption of the aortic arch, pulmonary aortic window, anomalous birth of the right pulmonary branch and intact interventricular septum in 5 patients. It also was described the possible embryological mechanisms of the aortopulmonary window that could affect the formation of the anomalous birth of the right pulmonary branch of the ascending aorta, at the level of the partition of the common trunk, where failure of trunk septation results in the aortopulmonary septal defect that varies in size from a small communication to the total absence of the septum; at the level of the pulmonary branches, the sixth arches join and form the pulmonary arterial bifurcation that normally merge with the pulmonary trunk in the posterior part , a failure of posterior trunk septation can alter the normal flow and cause anomalous origin of the right branch of the pulmonary artery from the aorta.

Bi et al. [1], reported that it is more frequent in males (67.1%). They generally present cyanosis, respiratory distress, heart murmur and symptoms of heart failure. Sixteen percent were diagnosed before 3 months.

The multidisciplinary approach requires an adequate echocardiographic assessment and 3D tomographic reconstructions. Berry et al. [5] recommended the use of angiography, which is still used in different centers [2]. However, it is currently possible to plan surgery with an adequate echocardiogram and CT angiography [1-4], which is the approach used in our center.

There are two surgical strategies: one stage, where all cardiac malformations are corrected, including the aortopulmonary window, reimplantation of the right pulmonary branch and repair of the aortic arch; the other strategy is the two-stage approach, which includes bandaging of the lung and then complete the repair after stabilizing the patient. However, the results have been unsatisfactory [6]. More recently, Shi et al. [4] and Choi et al. [7], recommend treating this group of patients in one single stage, with satisfactory results. This early correction is recommended even in the neonatal period due to pulmonary hypertension that appears early after birth, due to increased pulmonary flow to avoid irreversible damage to the pulmonary vascular bed [8].

In our center, we frequently use selective cerebral perfusion to treat the aortic arch, the reconstruction of the arch was achieved by aortic advancement performing an end-to-side anastomosis of the ascending aorta, as for the aortopulmonary window, direct closure of the aortic side was possible and on the pulmonary side, the right pulmonary branch was anastomosed, which was dissected and sectioned from the ascending aorta.

The postoperative course of our patient was uneventful. Complementary studies reported the absence of gradients at the level of the ascending aorta, pulmonary trunk, and aortic arch. A mean gradient of 5mmHg, measured by echocardiography at the level of the reimplantation of the right

pulmonary branch, stands out. Finding in the literature, the achievement of continuity and patency of the right pulmonary branch to the trunk is the key technical difficulty [4].

However, with the echocardiographic parameter at the level of the right pulmonary branch and what has been reported in the literature [1,4,9], postoperative stenosis of the right pulmonary branch continues to be the most frequent complication, due to the difficulty in reconstruction and the little space next to the ascending aorta. For this reason, the follow-up of our patient is mandatory and recommended.

In conclusion, Berry syndrome can be safely corrected in our environment in a single stage, paying close attention to the anastomosis of the right pulmonary branch. It requires a multidisciplinary approach for early diagnosis, adequate treatment and follow-up.

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