Staged treatment of a premature newborn with interrupted aortic arch and aorta-pulmonary window using intraoperative hybrid procedure before subsequent total correction – Long-term follow-up

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ABSTRACT

Interrupted aortic arch (IAA) associated with an aortopulmonary window (APW) is a rare cardiac malformation that needs an immediate diagnosis after birth and surgical treatment to avoid irreversible pulmonary lesions. Herein, we describe a case of successful staged treatment of premature neonate using many operative procedures. The first stage was stenting of patent ductus arteriosus with banding of pulmonary artery branches using a hybrid approach. At the age of one, he underwent total correction of malformation using Gore-Tex patch. Two procedures of implantation and redilatation of the previously implanted stents were performed in the following years. In the 8-year follow-up, the patient has been in good clinical condition, without cardiac symptoms. To the best of our knowledge, this is the first case report representing such a long-term follow-up of treatment of IAA accompanied by APW.

Keywords: Aortopulmonary window, catheterization, congenital heart diseases, hybrid procedure, interrupted aortic arch

INTRODUCTION

Interrupted aortic arch (IAA) accounts for about 0.9% of congenital heart defects (CHDs) and rarely occurs in isolation.[1] Aortopulmonary window (APW) is seen in about 0.2% of CHDs. In nearly 50% of the cases, APW is isolated and in the rest, it is accompanied by different CHDs.[2] Coexisting IAA (usually type A) with APW (usually proximal type) is a very rare critical CHD (7.3% of all cases of IAA and 32% of APW).[3] Such a complex defect requires urgent treatment because of duct-dependent systemic perfusion, pulmonary hypertension, and excessive pulmonary blood flow rapidly leading to congestive heart failure and death.

We present a successful multi-stage repair of this malformation in a low-birth-weight premature newborn.

CASE REPORT

We present a premature male neonate (pregnancy 1 terminated by cesarean section at 35 weeks of gestation because of maternal gestosis, birth weight 2.1 kg, length 46 cm). A heart murmur was noticed postnatally. X-ray of the chest showed cardiac enlargement and signs of pulmonary congestion. Detailed echocardiography revealed interruption of the 7 mm segment of aortic arch between the origin of the left subclavian artery and descending aorta (Celoria and Patton type A).[4] The main pulmonary artery and pulmonary artery branches were dilated, and APW was present (Richardson type II).[5]
Ductus arteriosus was wide with dominating right-to-left shunt. Prostaglandin E1 was started (0.03 µg/kg/min).

Due to prematurity, low body weight and overall deterioration, the patient was considered for a hybrid approach performed at the age of 2 weeks. Under general anesthesia, the chest was opened. Through the incision of the pulmonary trunk, the stent mounted on the balloon catheter was introduced into the patent ductus arteriosus (PDA) and expanded. Since the used stent did not cover the entire length of the PDA in the control angiography, an additional stent was inserted. In a further angiography, the location of both stents was correct [Figure 1]. Subsequently, banding was performed on the proximal segments of both pulmonary artery branches. The postoperative course was uneventful, and the patient was discharged in the good condition at the age of 4 weeks on aspirin 3 mg/kg per day.

The child was readmitted for the evaluation at 7 months of age in good condition. The peripheral pulse was symmetrical. Oxygen saturation in pulse oxymetry on his right hand was 92% and on the left foot 87%. Cardiac catheterization revealed the IAA [Figure 2], large APW 10 mm in diameter, normal SaO₂ (99%) in the ascending aorta and desaturation (67%) in its descending part, proximal stenosis resulting from banding of the two pulmonary artery branches with normal dimensions on the periphery [Figure 3].

He was readmitted at 13 months of age. A total correction was performed through median resternotomy. The banding of both pulmonary arteries was removed. The pulmonary artery trunk was longitudinally dissected and APW, as well as vascular stents within the arterial duct were visualized. The stents were removed from the duct and all the remnants of the duc tal tissue were removed. APW was closed with a Gore-Tex patch. The proximal part of the aortic arch and a part of the ascending aorta were dissected and augmented with a tailored aortic homograft. A segment of the whole circumference (conduit) of homograft was used to augment the gap between the descending aorta and aortic arch. The proximal homograft was incised along external curvature after removing all homograft arch branches. The distal part of the incised native aortic arch was anastomosed to the homograft segment's external circumference, and the arch was augmented with the proximal part of the homograft. The method of aortic arch reconstruction is presented in Figure 4. The whole body perfusion was restored. The right atrium was opened, and ASD was closed with mattress suture. During re-warming, the left and right pulmonary arteries were incised longitudinally beyond the narrowing caused by bands. Pulmonary branches and pulmonary trunk in the area of APW were augmented with suitably trimmed pulmonary artery homograft.

The postoperative course was complicated by pneumonia and left pleural effusion. After discharge, the patient was thriving showing normal weight gain. At the age of 5 years, he was admitted to the hospital for routine evaluation. Heart catheterization was performed which revealed stenosis of the right pulmonary artery. Subsequently, a stent was implanted enabling normal blood flow. Two years later, another routine catheterization revealed stenosis of both pulmonary arteries. A stent was inserted in the left pulmonary artery and the one implanted previously was redilated. Both procedures had no complications. Currently, the patient remains in the good condition without cardiac symptoms nor exercise limitations. In future, further redilation of the stents inserted in pulmonary arteries is considered as well as another hybrid procedure with aim to reinsert stent in LPA.

**DISCUSSION**

The association between IAA and APW is rare and represents a life-threatening malformation. Clinical
symptoms result from postnatal closure of PDA supplying the systemic circulation and excessive pulmonary blood flow in consequence of decreasing pulmonary resistance. In an echocardiography, the presence of both arterial valves differentiates APW from the common trunk. Establishing the distance of the arterial valve rings and the origin of coronary arteries from the proximal edge of the APW and the distance of its distal edge to the pulmonary bifurcation is essential. Possibility of early total correction has been postulated, but this is a treatment option appropriate only for term infants.\(^3\)\(^6\)\(^7\) According to Murin et al.,\(^3\) the early mortality rate reaches 15.1% (95% confidence interval: 6.8%–27.6%). Prematurity and low birth weight are the significant risk factors. Therefore, it is relevant to consider staged treatment with the hybrid approach as the first phase. It stabilizes the patient’s condition, allows the child to grow and makes it possible to delay and perform the total correction more safely. PDA stent placement has been shown to have a lower incidence of adverse events if performed intraoperatively using a direct rather than a percutaneous approach.\(^8\)

Hybrid techniques are appropriate in the situations where neither surgery nor catheterization alone offer optimal treatment, or when the combination of both techniques reduces invasiveness. Various hybrid procedures have become increasingly common in the management of selected CHD patients. In our center, we also perform hybrid single ventricle palliation when Norwood stage I surgery for hypoplastic left heart syndrome is too risky. This technique can be considered in patients with muscular ventricular septal defects that cannot be treated with percutaneous closure, including children who have an additional cardiac defect requiring conventional surgical repair and infants with small body weight.

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