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One-Stage Repair of an Interrupted Aortic Arch with an Aortopulmonary Window in a Premature Neonate

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Interrupted aortic arch with an aortopulmonary window is a rare congenital entity that is associated with high morbidity and mortality, especially in premature low-birth-weight infants, and the proper timing of surgical correction remains a matter of debate. We present the case of a premature infant weighing 1.6 kg who successfully underwent one stage surgical repair to treat interrupted aortic arch with an aortopulmonary window. The therapeutic management of this patient is described below, and a review of the literature is presented.

Key words: 1. Aortopulmonary septal defect
2. Interrupted aortic arch
3. Neonate

CASE REPORT

A premature female neonate, delivered at 33 weeks and weighing 1.79 kg, who was a part of a twin gestation, was admitted to our paediatric cardiac intensive care unit (ICU) on her twelfth day of life with symptoms of cardiac failure. On clinical examination, the patient's weight was 1.6 kg, the blood pressure in all four limbs was equal (75/25 mmHg), and the patient had a sinus rhythm with a heart rate of 160 beats per minute. A 4/6 systolic murmur was noted at the left sternal border. The patient was in tachypnea, with an oxygen saturation of 91% on room air, and the following results were found when arterial blood gas analysis was performed: pO_2=55 mmHg, pCO_2=38 mmHg, and pH=7.37. Echocardiography and cardiac magnetic resonance imaging were used to diagnose the presence of a large aortopulmonary window (APW), an interrupted aortic arch (IAA, B type) with an aberrant right subclavian artery, a patent foramen ovale (PFO), a patent ductus arteriosus (PDA), and a bicuspid aortic valve (Fig. 1A, B). The patient was initially treated with 0.025 μg/kg/min of prostaglandin E1, 5 μg/kg/min of dopamine, furosemide, and total parenteral nutrition, with the goal of attaining a weight of at least 2 kg before surgery. Further investigation included brain and renal ultrasonography, with no pathological findings. After 10 days of treatment with total parenteral nutrition, the patient's weight remained constant. Therefore, the decision was made to perform one-stage total correction.

1) Surgical repair

A median sternotomy was performed and cannulation was carried out through a two-stage venous cannula at the right atrium and an arterial cannula through the innominate artery via a 3.5 mm Gore-Tex (WL Gore & Associates, Flagstaff,

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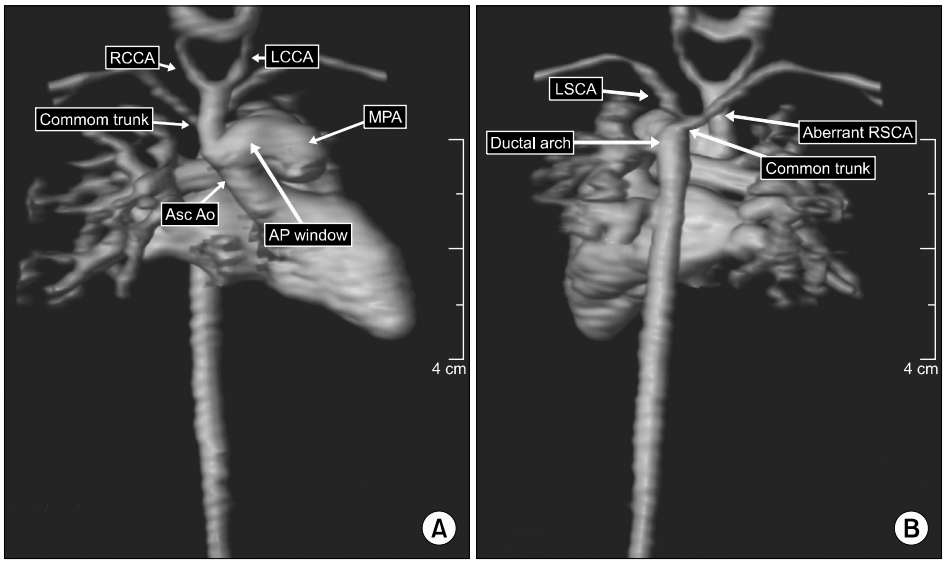

Fig. 1. (A) Anterior and (B) posterior views of cardiac magnetic resonance imaging depicting the interrupted aortic arch with an aortopulmonary window. RCCA, right common carotid artery; LCCA, left common carotid artery; MPA, main pulmonary artery; AP window, aortopulmonary window; Asc Ao, ascending aorta; RSCA, right subclavian artery; LSCA, left subclavian artery.

AZ, USA) synthetic graft. A type B interruption of the aortic arch was noted, as well as an aberrant right subclavian artery, which appeared to originate from a common vascular trunk arising from the posterior aspect of the ductal arch and aberrantly crossing to the right. Repair was performed using standard cardiopulmonary bypass, deep hypothermia to 18°C, and circulatory arrest.

Both pulmonary arteries, the PDA, and the ascending and descending aorta were dissected. After implementing cardiopulmonary bypass, both pulmonary arteries were snugged down. When circulatory arrest was initiated, the PDA was divided and the ductal tissue was excised, allowing further dissection of the descending aorta. The descending and distal aortic arch were anastomosed widely in an end-to-side fashion with a continuous 7/0 Prolene suture in the posterior wall and with interrupted 7/0 Prolene sutures in the anterior wall. The aberrant right subclavian artery was preserved, as the descending aorta was properly mobilized. The anterior wall of the APW was incised in a longitudinal fashion, and a large APW (type I) was recognised. A bovine pericardial patch was then used to close the aortopulmonary defect with a continuous 7/0 Prolene suture, using the sandwich technique. The total ischemic time in circulatory arrest was 40 minutes and the cardiopulmonary bypass time was 150 minutes. The patient was admitted to the ICU with a low dose of inotropes and an open chest.

The postoperative course was uneventful and the patient made a full recovery. Delayed sternal closure was performed on postoperative day 3, and the patient remained haemodynamically stable. On postoperative day 7, the patient was extubated and inotropes were discontinued. The patient was first fed on postoperative day 8 and was discharged from the hospital on postoperative day 10. At a three-year follow-up, the patient showed normal growth. Echocardiography revealed no persistent aortopulmonary defect and continuous flow at the aortic arch with a gradient of 10 mmHg without left ventricular outflow tract obstruction, and the clinical status of the patient was excellent.

DISCUSSION

The association of APW with IAA is rare. Only 3.5%–4.2% of patients with IAA have APW, representing 0.046% of patients with congenital heart disease [1]. Thus, the cumulative experience with this combination of lesions is limited, mainly because it has been mostly described in isolated case reports. The largest multi-centre retrospective study, carried out by Konstantinov et al. [2], included twenty patients with APW among 472 neonates with IAA who were operated on from 1987 to 1997. In their study, the minimum weight of any neonate who underwent surgery was 2.1 kg at the time of the operation [2]. In this report, we present the case of a

premature underweight (1.6 kg) neonate with APW, IAA, PDA, and PFO, who was successfully operated on via a median sternotomy and total one-stage repair. To the best of our knowledge, this is the first reported case of this operation being successfully performed on such an underweight neonate.

Many infants with IAA present early with cardiac failure, shock, and severe acidosis. Initial management aims to stabilize the patient and involves prostaglandin E1 to maintain ductal patency, mechanical ventilation, avoidance of enteral feeding, and inotropic support. Early surgical treatment is required in infancy. However, a two-stage approach may be an alternative, depending on the specific defects observed in a given case. A coexisting ventricular septal defect could be initially approached through pulmonary artery banding, and an atrial septal defect with PFO may be corrected at a later time, but more complex cardiac anomalies, such as truncus arteriosus, transposition of the great arteries, or APW should be corrected in the same operation [3,4].

The combination of APW with IAA creates a particularly serious challenge because infants with this condition are critically ill at birth. Complete surgical repair can be achieved in one stage and includes the ligation and division of the PDA, reconstruction of the aortic arch, and the closure of APW and other defects with a pericardial, bovine, or synthetic patch. Postoperative mortality varies from 15% to 33%, decreasing significantly as postoperative time increases, and survival approaches 85% at 10 years. Approximately half of these patients will need at least one re-intervention in the future in order to deal with related complications, such as aortic arch obstruction (50%), left main bronchial compression (10%), or pulmonary artery stenosis (25%). If pulmonary or aortic stenosis occurs, balloon angioplasty is the first option [1,2,5]. The preoperative diagnosis is usually made using echocardiography. Major advances in cardiovascular magnetic resonance angiography allow the precise characterization of the anatomy and topography of an interrupted aortic arch and associated anomalies, while avoiding exposure to ionizing radiation [6].

Surgical repair, which takes place under cardiopulmonary bypass and circulatory arrest, includes division of the PDA, reconstruction of the IAA, and closure of the APW. Median sternotomy is the incision of choice. Reconstruction of the aortic arch with an end-to-side anastomosis is the best surgical technique and is usually feasible with adequate mobilization of the descending aorta. The use of the subclavian artery or pericardium for patch arterioplasty is rarely necessary. Patch augmentation, as a related complication, may be associated with bronchial compression. Direct anastomosis is more likely to result in a satisfactory lumen over the long term, compared with the use of prosthetic material or a vascular homograft, as the anastomosis will grow along with the patient. The APW may be accessed through a transpulmonary window or a transaortic incision.

Optimal cerebral and visceral protection is crucial in these cases. In addition to the traditional method of deep hypothermic circulatory arrest (DHCA), antegrade cerebral perfusion with moderate hypothermia has been used in children undergoing aortic arch surgery [7]. Although aortic arch reconstruction accompanied by selective antegrade cerebral perfusion has a lower risk of neurological complications than DHCA, selective antegrade cerebral perfusion is associated with a higher incidence of renal complications [8].

Several reports have observed that the presence of type B IAA and an anomalous right subclavian artery are risk factors for left ventricular outflow tract (LVOT) obstruction [5,9,10], probably due to alterations in blood flow across the LVOT. However, LVOT obstructions after IAA repair are highly heterogeneous. The detection of pre-existing LVOT obstructions through careful quantitative echocardiographic assessment can certainly assist surgical decision-making. Moreover, the heterogeneity of LVOT obstructions mandates individually tailored management [5]. In our case no LVOT obstructions were observed either preoperatively or during the three-year follow-up period, although we preserved the aberrant right subclavian artery.

In cases where total correction is unfeasible, staged repair may be the most preferable therapeutic choice. Bilateral pulmonary artery banding with continuous prostaglandin E1 infusion and bilateral pulmonary artery banding with stenting of the PDA in a hybrid procedure have been successfully employed in the management of low-birth-weight and unstable infants [11,12].

In conclusion, one-stage correction of IAA with concomitant APW in a premature, low-birth-weight neonate is a highly risky and complex operation, but it can be safely performed in ex-

perienced centres, given the concomitant psychosocial and economic advantages of the procedure. Early surgical treatment is a therapeutic option, even in low-weight premature neonates weighing less than 2 kg. These patients have a good prognosis and can achieve satisfactory early and late outcomes due to advances in anaesthesia, cardiopulmonary bypass, and perioperative management.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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