Complete Repair of Interrupted Aortic Arch with Ventricular Septal Defect in a Premature Weighing 1600 Grams — A Case Report

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A premature neonate with gestational age of 32 weeks and birth body weight of 1650 grams was diagnosed to have type A interrupted aortic arch, perimembranous type ventricular septal defect, atrial septal defect, and patent ductus arteriosus. One-stage complete reconstruction of aortic arch and concomitant repair of intracardiac defects was performed at age of 13 days with body weight of 1600 grams. Through innominant artery and retrocardiac descending aorta cannulation for continuous cerebral and lower body perfusion, deep hypothermic circulatory arrest was avoided. Postoperative renal function was excellent, and there were no neurological sequelae after 24 months’ follow-up.

Key Words: Premature • Interrupted aortic arch • Ventricular septal defect

INTRODUCTION

Low birth weight is a risk factor for poor outcome in corrective surgery for many cardiac defects.1 Congenital cardiac defects in low birth weight infants are traditionally managed with supportive therapy or palliative operations, followed by a definitive repair later at a more conventional age and body weight. The morbidity with such approach is high. In addition, there is also a major concern among neonatologists regarding intracranial bleeding after cardiopulmonary bypass. Interrupted aortic arch is a severe form of cardiac anomaly with nearly 100 percent mortality if not recognized and treated early. The associated intracardiac lesions often lead to death if only the arch anomaly is repaired. We report a premature neonate with low body weight who underwent primary one-stage repair of interrupted aortic arch and intracardiac lesions without circulatory arrest during operation.

CASE REPORT

A 6-day-old neonate was transferred from a local hospital because of prematurity with gestational age of 32 weeks. Birth body weight was 1650 grams. He was born with vaginal deliver, and the Apgar scores were 7 and 8 at 1 minute and 5 minutes, respectively. After birth, shortness of breath was noted and grade I respiratory distress syndrome (RDS) was impressed. Respiratory support with nasal continuous positive airway pressure was given. Physical examinations revealed poor activity with coarse breat sounds, and grade II/VI continuous murmur was heard over the left upper sternal border. The chest roentgenography showed cardiomegaly and groundglass opacity over bilateral lung fields. ECG showed sinus tachycardia, with heart rate between 140-160 beats per minute. Transthoracic echocardiography (TTE) showed type A interrupted aortic arch (IAA), perimembranous type ventricular septal defect (VSD),
atrial septal defect (ASD), and patent ductus arteriosus (PDA) with right-to-left shunt. Prostaglandin E was infused after diagnosis. Operation was performed at the age of 13 days with body weight 1600 grams. Standard median sternotomy with partial thymectomy was performed. Arterial cannulation was made with an 8Fr. DLP cannula (Medtronic Inc., Minneapolis, MN, USA) through a 3.5-mm expanded-polytetrafluoroethylene (E-PTFE) graft connected to the innominate artery for cerebral perfusion, and a 6Fr. DLP cannula (Metronic Inc., Minneapolis, MN, USA) in the retrocardiac descending aorta for lower body perfusion. Venous drainage was obtained by bicaval cannulation with 8Fr. RMI angle cannula (Edwards Lifesciences, Irvine, CA, USA) and 14Fr. RMI cannula (Edwards Lifesciences, Irvine, CA, USA), respectively (Figure. 1A). Under high flow (cardiac index 3.0-3.5 L/M²BSA) and moderate hypothermic (rectal temperature 26 °C) cardiopulmonary bypass, the PDA was divided with ductal tissue excised. Aortic arch reconstruction and intracardiac repair were performed under cold crystalloid cardioplegic arrest. Direct anastomosis between ascending and descending aorta with continuous 6-0 polydioxanone suture in an end-to-side fashion was performed because of aortic arch hypoplasia, which was only 2.0 mm in diameter (Figure. 1B). VSD was repaired with Teflon patch anchoring with 6-0 polypropylene sutures interruptedly. Aortic cross-clamp time was 81 minutes, and cardiopulmonary bypass time was 231 minutes. No circulatory arrest was used during operation. Postoperative TTE showed no residual VSD or ASD, and only mild residual aortic stenosis at the anastomotic site, with pressure gradient of less than 20 mmHg. Brain sonography showed no intraventricular hemorrhage. Neurological examination showed no neurological deficit. Renal function was within normal limits throughout the postoperative course. After follow-up for 24 months, the patient grew well without any intervention.

DISCUSSION

Recent advances in neonatal cardiac surgery have dramatically improved the outcomes of surgery in the past decade. However, low birth weight is still considered by many as a contraindication to cardiopulmonary bypass and anatomic repair of even relatively simple cardiac defects. In recent years, some centers reported excellent results after early complete repair of congenital cardiac defects in low birth weight and very low birth weight premature patients.2,3 Pawade and associates2 reported on a series of 60 patients with low birth weight who underwent operations for various congenital heart defects, with a 16.5% early mortality rate and a 13.3% late mortality rate. In their experience, the risk factors for early death after the operation in patients smaller than 2500 grams were preoperative metabolic acidosis, univentricular palliation, duration of cardiopulmonary bypass, and postoperative low cardiac output. Reddy

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**Figure 1.** (A) Cannulation at innominante artery by means of E-PTFE graft for cerebral perfusion and direct descending aorta cannulation for lower body perfusion. E-PTFE = expanded polytetrafluoroethylene; Innom A = innominate artery; Des Ao = descending aorta; MPA = main pulmonary artery; PDA = patent ductus arteriosus. (B) Direct anastomosis between ascending aorta and descending aorta due to aortic arch hypoplasia.
and associates reported on a series of 102 infants no larger than 2500 grams with a 10% early mortality rate and 9% late mortality rate. They believed delay in intervention was associated with increased complications including ventilatory dependency, failure to thrive, absence of weight gain, sepsis, chronic pulmonary disease, necrotizing enterocolitis, and acute renal failure.

IAA is a rare congenital heart disease in which the aortic arch is atretic or interrupted. The prognosis for untreated infants is extremely poor, and survival beyond infancy is uncommon. The surgical approach to this condition in different centers varies considerably. Most reports advocate two-stage repair, while other centers prefer a one-stage repair of the defect and associated anomalies. In recent years, the results of both primary and two-stage repair of IAA have improved, and the early mortality in some centers is about 10%. In the majority of cases, the mortality and morbidity after surgical repair are a result of total preoperative and intraoperative ischemia (operative circulatory arrest time) and systemic or organ-related failure (low cardiac output syndrome, sepsis, acidosis, coagulopathy, renal, hepatic or multi-organ failure syndrome). Our patient was a case of prematurity with gestational age of 32 weeks and birth body weight only 1650 grams. He had severe congenital cardiac defects, with type A IAA, aortic arch hypoplasia, VSD, ASD, and PDA. We used the policy of early intervention and one-stage complete repair. During operation, we cannulated the innominate artery through a 3.5-mm E-PTFE graft for cerebral perfusion and retrocardiac descending aorta just superior to the diaphragm for lower body perfusion. Arch reconstruction and repair of VSD and ASD were performed under continuous perfusion to the brain and lower body. Postoperative renal function was excellent, and there was no neurological deficit. The technique of descending aorta cannulation for aortic arch reconstruction was first reported by Imoto and associates, and was believed to be an effective method to avoid circulatory arrest which had been proved to have detrimental effects on renal and neurologic function. During the short-term follow up of 24 months, the patient was doing well and grew well. The long term result needs to be further evaluated.

In summary, one-stage repair of congenital heart defects can be performed safely and effectively in premature and low birth weight infants, as in the patient we have reported herein. Special cannulation technique as we mentioned above to avoid circulatory arrest may be a useful alternative in aortic arch reconstruction surgery.

REFERENCES

1600 公克早產兒主動脈弓斷離合併心室中膈缺損之完全矯正 — 一病例報告

潘俊彥*1 謝凱生2 康沛倫1 盧麗芬1 吳東和1 鄭國琪1
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一例 32 週早產兒出生體重 1650 公克，診斷為 A 型主動脈弓斷離，心室及心房中膈缺損，開放性動脈導管。病患於 13 天大體重 1600 公克時接受一期主動脈弓重建及心內修補手術，藉由無名動脈及心後下行主動脈持續灌注，深低體溫循環停止可以完全避免。術後病患腎功能正常，且無神經學方面之缺損。

關鍵詞：早產兒、主動脈弓斷離、心室中膈缺損。