The Heart Surgery Forum #2006-1141
10 (1), 2007 [Epub December 2006]
doi: 10.1532/HSF98.20061141

Online address: http://cardenjennings.metapress.com

Rastelli and Norwood Combination for the Treatment of Type I Truncus Arteriosus and Hypoplastic Aortic Arch: A Case Report

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ABSTRACT

Although truncus arteriosus is often treated with low mortality and morbidity rates, truncal valve patency and aortic arch and coronary artery anomalies are factors that can contribute to a worse outcome. In this report, we present our experience with the combination of Rastelli and Norwood procedures for the treatment of Type I truncus arteriosus that was complicated by a hypoplastic aortic arch.

INTRODUCTION

Truncus arteriosus (TA) is a rare congenital cardiac disease, and 15% of the cases include further aortic pathology such as hypoplasia, coarctation, or interruption. The combination of TA with an additional aortic pathology severely deteriorates a patient's status and requires early surgical treatment in the infancy period. After the first successful repair described by Gomes and McGoon in 1971, there have been sporadic cases reported, but the literature lacks larger series explaining a promising surgical repair for TA combined with aortic arch anomalies [Konstantinov 2006].

CLINICAL SUMMARY

Our patient was an 8-day-old 2950 g female infant, born to term, referred with the diagnosis of Type I TA (Collett and Edwards classification) associated with a hypoplastic aortic arch. On admission, she was in cardiac failure and required cardiac inotropic and mechanical ventilatory support. The truncal valve had 4 leaflets and mild regurgitation on echocardiographic evaluation. Ventricular septal defect was subarterial with a diameter of 9 mm. The aortic arch was 3 to 4 mm and the ductus arteriosus was patent (Figure 1). Surgical treatment was planned after initial stabilization and performed at the age of 10 days. A combination of Norwood and Rastelli procedures were used for the treatment (Figure 2). The sternum was closed on the third postoperative day; however, the patient could not be extubated and died due to necrotizing enterocolitis on the 16th postoperative day.

Surgical Technique

The operation was performed through median sternotomy at moderate hypothermia. Myocardial protection was achieved with antegrade cold crystalloid cardioplegia and topical ice slush. An arterial cannula was placed into an expanded polytetrafluoroethylene graft that was anastomosed to the origin of the brachiocephalic trunk and bicaval cannulation was performed (Figure 1). A left ventricular vent was placed via the right superior pulmonary vein. After the initiation of cardiopulmonary bypass, the ductus arteriosus was ligated and divided. Right and left pulmonary arteries were separately snared, and the main pulmonary artery was dissected below the pulmonary artery branches. The patient was cooled to 25°C. The arterial cannula was elongated into the brachiocephalic trunk and flow was reduced to one third. The descending aorta was clamped, and the left common carotid and left subclavian arteries were snared. A longitudinal incision was made to the inferior surface of the aortic arch, extending to the descending aorta. The transected main pulmonary artery was anastomosed to this region (Figure 3). The clamp at the descending aorta and the snares of the left common carotid and subclavian arteries were removed, the arterial cannula in the brachiocephalic trunk was withdrawn into the expanded polytetrafluoroethylene graft, and bypass flow was returned to normal. A large cross clamp covering the truncus was placed, and cardiac arrest was established. A vertical right ventriculotomy was performed, and the ventricular septal defect was closed with a dacron patch in a fashion directing the left ventricular flow to the truncal valve. Then the cross clamp was removed and the right ventricular outflow tract was reconstructed with a 14-mm Contegra (Medtronic, Minneapolis, MN, USA) (Figure 2). Weaning from cardiopulmonary bypass was possible with dopamine and low-dose adrenalin support. The sternum was not closed. The cross clamp, selective cerebral perfusion, and cardiopulmonary bypass times were 13 minutes, 23 minutes, and 29 minutes, respectively.

Received September 16, 2006; accepted September 27, 2006.

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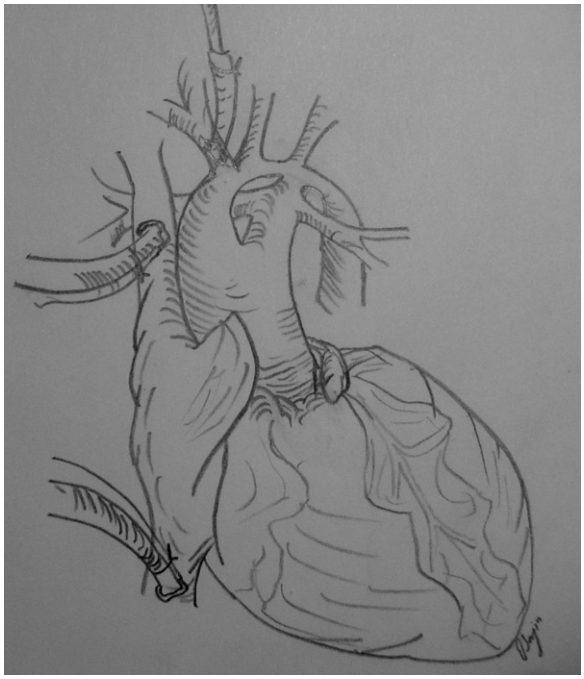

Figure 1. Schematic drawing of the anatomy of the patient's cardiac pathology.

DISCUSSION

Treatment of TA associated with an aortic pathology has been a challenge for pediatric cardiac surgeons and requires early surgical intervention in the first few days of life. Although there are various case reports in the literature from different authors explaining their techniques, there is not an accepted single concept for the treatment of the pathology [Konstantinov 2006].

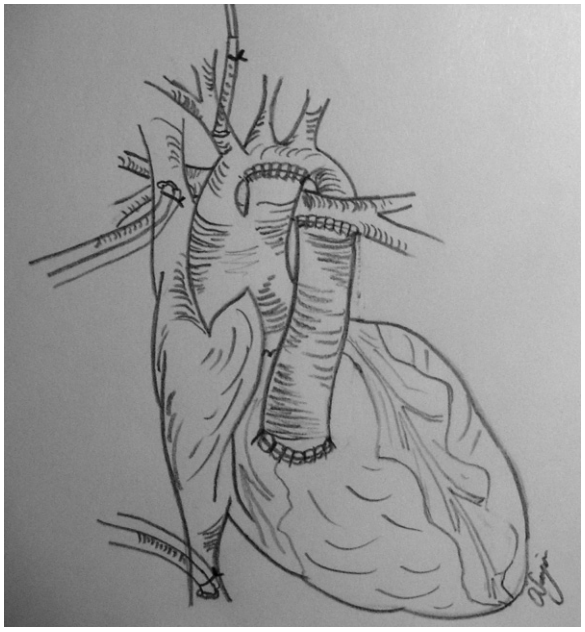

Figure 2. Schematic drawing of the completed operation, for which we used combined Norwood and Rastelli procedures for the treatment of Type I truncus arteriosus associated with a hypoplastic aortic arch.

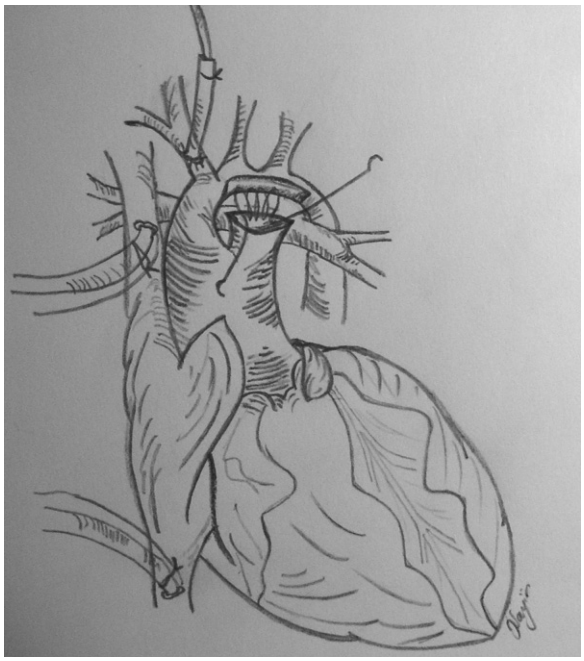

Figure 3. Schematic drawing of the hypoplastic aortic arch reconstruction. The ductus was ligated and divided. The pulmonary artery was transected just below the right and left pulmonary branches. The inferior margin of the aortic arch was incised to the descending aorta, and the pulmonary artery was anastomosed to this region of the aortic arch.

The treatment of TA associated with aortic interruption could be performed in a 2-stage manner, first the repair of the interruption followed by the treatment of the TA in the next step; the unfavorable results of this method directed many surgeons to single-stage complete repair of the cardiac defects, including truncal valvuloplasty if necessary [Jahangiri 2000]. Still, the operation has high mortality rates [Miyamoto 2005].

During the repair of aortic interruption, continuity of the aorta can be achieved with direct anastomosis or by the aid of a conduit. In the case of a hypoplastic aortic arch, reconstruction with an autologous pulmonary artery patch seems to be the best option and is widely used by many surgeons due to its capacity for growth and similarity to the aortic tissue in nature. On the other hand, during TA repair the most crucial point of the treatment is keeping the valves in good condition with minimal regurgitation and stenosis. Generally in TA, the truncal valve is dysplastic and may have various grades of stenosis and regurgitation. However, if the patency of the valve is not highly distorted, repair of the valve may not be necessary during the initial surgery. It is well known that mild to moderate dysfunction of the truncal valve is well tolerated after surgical correction

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[Brown 2001]. Therefore, TA repair must be performed with great precautions, especially in patients whose preoperative truncal valve functions are mildly deteriorated and in cases combined with an aortic pathology where aortic manipulations are necessary.

Our patient had a quadricuspid truncal valve with mild regurgitation, and to avoid compromising the valve's functions during surgery we preferred to keep the root untouched and to perform a modified Norwood procedure to reconstruct the arch. Following the arch repair, a Rastelli procedure was performed for further correction. The patient tolerated surgery, but died because of necrotizing enterocolitis.

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