Rudimentary right ventricle to pulmonary artery shunt in the Norwood procedure

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Abstract We describe here successful palliative repair of tricuspid atresia, hypoplastic right ventricle, transposition of the great arteries, and hypoplastic aortic arch in a neonate. The repair consisted of the Norwood procedure with a rudimentary right ventricle to pulmonary artery shunt, which was located on the right side of a neo-aorta. This procedure could be a useful adjunct to avoid left ventriculotomy and its subsequent dysfunction.

Key words Norwood procedure · Right ventricle to pulmonary artery shunt · Rudimentary right ventricle · Tricuspid atresia

Introduction

Ventriculotomy in the Norwood procedure with right ventricle to pulmonary artery shunt poses a concern with regard to systemic ventricular dysfunction, tricuspid valve regurgitation, and ventricular arrhythmias. Technical difficulty with a high risk of stenosis at the proximal anastomosis site is another obstacle in left ventricle to pulmonary shunt in single left ventricle variants. A rudimentary right ventricle to pulmonary artery shunt on the right side of a neo-aorta in the Norwood procedure could be a useful adjunct to avoid these issues.

Case report

A 2,972-g male neonate was referred to our hospital after scheduled cesarian section based on fetal echo diagnosis of tricuspid atresia. Echocardiography after birth led to the diagnosis of tricuspid atresia, hypoplastic right ventricle, transposition of the great arteries, hypoplastic aortic arch, coarctation of aorta, patent ductus arteriosus (PDA), and nonrestrictive ventricular septal defect. A chest X-ray showed cardiomegaly (with a cardiothoracic ratio of 57%) and increased pulmonary vascularity. Lipo-PGE1 was used to maintain ductus arteriosus patency, and carbon dioxide was added to a head box to control pulmonary blood flow.

Surgery was performed on day 3. The ascending aorta was located in front of the huge main pulmonary artery. The right anteriorly located rudimentary right ventricle gave rise to the small ascending aorta. After dissection of the proximal and distal aortic arch and bilateral pulmonary arteries, cardiopulmonary bypass was established with cannulas in the innominate artery and PDA, and a venous cannula in the right atrial appendage. After the distal ascending aorta was cross-clamped and bilateral pulmonary arteries ligated, crystalloid cardioplegia was administered. The arch was directly reconstructed under isolated cerebral perfusion via the innominate artery, and the cannula via PDA was removed. The atrial and ven-
tricular septal defects were enlarged and trabeculation in the right ventricle was resected through right ventriculotomy under deep hypothermic circulatory arrest. A rudimentary right ventricle to pulmonary artery shunt using a 5-mm Polytetrafluoroethylene (PTFE) graft was constructed on the right side of a neo-aorta (Fig. 1). Cardiopulmonary bypass was terminated with arterial systolic blood pressure 52 mmHg and left atrial mean pressure 8 mmHg under inotropic supports.

Nitric oxide inhalation was discontinued on the 3rd postoperative day. Scheduled delayed sternal closure was performed on the 5th postoperative day, and extubation was performed on the 15th postoperative day. The subsequent postoperative course was uneventful, and the patient was discharged home on postoperative day 51 without any complications. A computed tomography scan at 1-month follow-up demonstrated an unobstructed right ventricle to pulmonary shunt without kinking (Fig. 2). The patient underwent angiography 5 months later due to a hypoxia of SpO2 62%–68%, which demonstrated nonrestrictive flow across the ventricular septal defect and stenosis at the proximal anastomosis site of the shunt (Fig. 3). Echocardiography showed the size of the ventricular septal defect was 5.4 mm in four-chamber view. Left ventricular function was preserved. A bidirectional cavopulmonary shunt was subsequently performed. The patient is currently waiting for total cavopulmonary anastomosis at an outpatient clinic and is in a fair condition.

Discussion

To improve postoperative hemodynamic status, a right ventricle to pulmonary artery shunt in place of a modified Blalock–Taussig shunt has been used in the Norwood procedure. A ventriculotomy in the Norwood procedure with right ventricle to pulmonary artery shunt in