Biventricular Repair After Stage II Univentricular Surgery: Palliation Is Not a One-Way Path
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Surgical decision in mild forms of hypoplastic left heart syndrome can be challenging. Once a univentricular pathway has been chosen, it can be difficult to reconsider a biventricular repair. A commitment to a palliative pathway is usually considered irreversible after initial univentricular repair. We present this case as an example in which the primary surgical palliation pathway was altered, and eventually a successful biventricular repair was performed in a mild variant of hypoplastic left heart syndrome, despite the fact that maneuvers to promote left ventricular growth were not recruited at the time of initial surgery.

A 2.54-kg premature infant was born at 34 weeks’ gestation with a prenatal diagnosis of hypoplasia of the left ventricle, mitral valve, and aortic valve (Fig 1). The patient was intubated, and prostaglandin E2 infusion was started because of persistent acidosis, poor peripheral perfusion, and respiratory distress. The transthoracic echocardiogram demonstrated the following: mitral valve hypoplasia (annulus, 6 mm; Z-score, -3.24), moderate left ventricular hypoplasia with non-apex-forming left ventricle (left ventricular end-diastolic volume [LVEDV], 3.5 mL; Z-score, -3.64), aortic valve hypoplasia (annulus, 4.5 mm; Z-score, -3.03) with bicuspid morphology, and severe discrete coarctation in juxtaaductal area with a narrowest diameter of 1.4 mm.

Given the left ventricular and mitral valve hypoplasia, at day 7 of life, the patient underwent a Norwood procedure with right ventricle to pulmonary artery shunt (6 mm) placement, aortic arch reconstruction, coarctectomy, and atrial septectomy. Maneuvers to promote left ventricular growth were not used, such as resection of endocardial fibroelastosis, aortic or mitral valvuloplasty, atrial septal defect restriction, or addition of accessory pulmonary blood flow. Upon discontinuation of cardiopulmonary bypass, atrial septum was unrestrictive; however, severe tricuspid regurgitation was noted on the transesophageal echocardiogram. Cardiopulmonary bypass was re instituted, and tricuspid valve repair was performed in the form of posterior annuloplasty and anterior septal commissuroplasty. The patient received extracorporeal membrane oxygenator support for 4 days. The hospital stay was protracted to 70 days because of respiratory insufficiency, renal failure, and nutritional concerns with prematurity.

At 4 months of life, echocardiogram and cardiac catheterization showed a hypoplastic left ventricle with persistent mitral valve (annulus, 8 mm; Z-score, -2.55) and native aortic valve stenosis (annulus, 6 mm; Z-score, -2.4; LVEDV, 8.2 mL; Z-score, -3.0). Because of worsening cyanosis and persistent concerns for hypoplasia of
left heart structures, the patient underwent stage II palliation in the form of bidirectional cavopulmonary anastomosis and takedown of the right ventricle to pulmonary artery shunt. The atrial septum remained unrestrictive. At 3 years of age, catheterization and echocardiogram showed normal left ventricular size and function. The mitral valve annulus measured 12 mm (Z-score, −2.16) and the native aortic valve measured 10 mm (Z-score, −1.62; LVEDV 25.2 mL, Z-score, −2.3. In light of the left ventricular growth, a decision was made to undergo biventricular repair. The patient underwent takedown of Norwood anastomosis and reconstruction of the ascending aorta, reconstruction of the main pulmonary artery, and atrial septal defect closure leaving a 3-mm fenestration. The cavopulmonary anastomosis was left in place. The patient was discharged home on the ninth postoperative day. Postoperatively, the patient did well clinically. At the time of discharge, the diastolic mitral valve gradient was a mean of 6 mm Hg. The interatrial communication had continuous flow, with a mean gradient of 7 mm Hg. There was no left ventricular outflow tract obstruction with maximum velocity by continuous wave Doppler interrogation across the left ventricular outflow tract of 1.6 m/s. The mean gradient across the aortic valve was 6 mm Hg. The left ventricular function was normal, with a cardiac index of 3.8 L/min/m² by echocardiogram.

Comment

The initial decision of univentricular palliation with Norwood procedure was made because of the presence of multiple-level obstructions and a left heart considered unable to sustain independent systemic circulation. It is important to note that during the first surgery, no attempt was made to promote left ventricular growth. Such attempts would include removal of endocardial fibroelastosis, aortic or mitral valvuloplasty, atrial septal defect restriction, or the addition of accessory pulmonary blood flow, such as a systemic-to-pulmonary shunt [1]. Despite this decision, the left heart structures had grown at the time of second surgery. However, biventricular repair was not deemed favorable in the presence of persistent mitral and aortic valve hypoplasia and left ventricular hypoplasia noted by echocardiography. The cardiac catheterization before the third surgery demonstrated further growth of left ventricle with near equally sized left and right ventricles. Therefore, biventricular repair was ultimately performed.

Biventricular repair has been performed successfully after initial Norwood procedure in patients with left ventricular outflow tract obstruction and interrupted aortic arch or severe coarctation [2, 3]. Initial Norwood procedure followed by biventricular repair has also been reported in double-outlet right ventricle with subpulmonary ventricular septal defect and aortic arch hypoplasia [4]. In hypoplastic left heart syndrome, there is an additional issue of left ventricle hypoplasia. During the interstage period, the blood flow through the left ventricle may help it to grow with time [1, 5]. Recently, Emani et al. [1] published a case series in which they surgically recruited borderline left ventricles in patients with hypoplastic left heart syndrome, in a stepwise manner in patients who were not good candidates for primary biventricular repair. Their approach included augmentation of left ventricular filling by various interventions. In contrast, in our case the left ventricular growth continued even in the absence of any surgical intervention to augment left ventricular filling. This result raises the possibility that perhaps there is another subset of hypoplastic left heart syndrome with borderline left ventricles, in which the left ventricle continues to grow even when a surgical attempt to promote its growth has not been made. This possibility underscores the importance of reevaluating cardiac anatomy before the next stage of surgical palliation. In such mild variants of hypoplastic left heart syndrome, initial univentricular palliation should not be considered a one-way path to Fontan procedure. The left heart structures can grow with time, creating a possibility of biventricular repair in the future.

References