For select neonates with borderline left ventricle, staged left ventricular (LV) recruitment is a promising strategy that avoids the morbidity associated with neonatal biventricular repair or long-term single-ventricle palliation.1-3 One recruitment maneuver that can increase the size of left heart structures through volume loading is the “Super Glenn” operation, which consists of a right-sided cavopulmonary shunt, a contralateral aortopulmonary shunt, separation of the branch pulmonary arteries, and fenestrated atrial septation.4 However, construction and takedown of the Super Glenn is technically challenging. Here, we present the case of a patient who initially underwent a Norwood-Sano operation, then received a simplified LV recruitment operation with Sano shunt upsizing and fenestrated atrial septation.

CASE REPORT

The patient is a male born with Shone complex variant including hypoplasia of the transverse aortic arch with juxtaductal coarctation, bicuspid aortic valve, mitral arcade with severe mitral regurgitation (MR), and restrictive atrial septal defect (Figure 1, A and B). Permission was granted by the patient’s guardians to publish this case report. This study received institutional review board approval (Pro00101549, May 2020). Echocardiography at birth demonstrated a small co-apex forming LV and normal biventricular function. The left ventricular end-diastolic volume index (LVEDVi) was 25 mL/m² (Pediatric Heart Network z score = -2.3) with a diastolic length of 27 mm (z score = -2.6) (Figure 1). The aortic valve annulus measured 5.3 mm (z score = -2.9) and the mitral valve annulus measured 8.2 mm (z score = -2.6). The proximal transverse aorta measured 2.9 mm (z score = -4.1). The atrial septum was restrictive with mean gradient 9 to 11 mm Hg. Two scoring systems, the Rhodes (1.12) and CHSS-1 (13.67), favored univentricular repair whereas the discriminant (0.07) and CHSS-2 (36.89) scores favored biventricular repair.

Given uncertainty regarding the reversibility of the MR, the decision was made to proceed with initial univentricular repair to assess for improvement in MR after relief of arch obstruction, and to simultaneously not rely on the small LV and abnormal mitral valve for systemic output. A Norwood operation with a 6-mm Sano conduit was performed on day of life 3 with sustained total all-region perfusion.5 An atrial septectomy was performed to prevent left atrial hypertension in the setting of severe MR. Due to
unstable arrhythmias coming off bypass, extracorporeal membrane oxygenation rest was used. The patient stabilized on amiodarone and extracorporeal membrane oxygenation decannulation was performed on postoperative day (POD) 3, followed by chest closure on POD5. The patient was discharged 4 weeks later.

Cardiac magnetic resonance imaging (MRI) at 4 months of age showed persistently small left-sided structures. There was no evidence of endocardial fibroelastosis. LVEDVi was 19.0 mL/m² (Figure 1, C) with mitral valve annulus measuring 9.9 mm (z score = −2.3) and native aortic valve annulus measuring between 5.9 and 7.1 mm (z score = −3.3 to −1.8). Mitral valve function was markedly improved on MRI and echocardiography, with mild regurgitation and no stenosis. Based on MRI estimates of native aortic valve, pulmonary valve, and Sano flows, the LV cardiac index was 1.9 L/min/m² and the LV/RV flow ratio was 0.43 (0.6 liters per minute/1.4 liters per minute). Cardiac catheterization demonstrated a normal LVEDP of 8 mm Hg with no gradient across the mitral and native aortic valves; however, the LV was incompletely loaded due to the atrial septal defect. Pulmonary vascular resistance index was normal at 1.30 Wood-units/m². Given the improved MR after relief of arch obstruction, LV recruitment was thought to be feasible. At 6 months of age, an LV recruitment operation was performed including 4-mm fenestrated atrial septation and Sano upsizing from 6 mm to 8 mm to volume load the LV. On bypass with the heart arrested, the 6-mm Sano was removed, the new 8-mm conduit was placed into the previous ventriculotomy site using a “dunk” technique, and the distal end was sewn to the pulmonary artery confluence without patch augmentation. Echocardiography on POD3 showed patent Sano, left-to-right shunt across the atrial fenestration, normal biventricular function, and unchanged mitral valve function. Postoperative saturations were 85% to 90% due to near-complete intracardiac septation and favorable streaming. The patient was discharged on POD4.

Follow-up MRI at 13 months of age showed excellent growth of the LV (LVEDVi of 57.4 mL/m²; Figure 1, D). The mitral valve annulus was 13 mm (z score = −1.2) and the aortic valve annulus was 10 mm (z score = +0.2) with mild stenosis of the native aortic valve. There was no interval change in mitral valve function. There was no residual atrial shunt on MRI. LV cardiac index increased to 3.3 L/min/m², and LV/RV flow ratio increased to 1.0 (1.3 LPM/1.3 LPM). Pulmonary valve flow decreased from 1.9 L/min/m² before recruitment to 0.8 L/min/m², which suggested preferential flow from the RV to the upsized Sano. The regurgitant fraction of the Sano did not change substantially during LV recruitment (13%-22%) and the RV end-diastolic volume index did not change (89 mL/m² to 88 mL/m²), suggesting no evidence of RV dilation as a result of Sano conduit regurgitation. Catheterization at this time revealed mild proximal narrowing of bilateral branch pulmonary arteries, normal LVEDP of 7 mm Hg, mild stenosis across the native aortic valve (10 mm Hg gradient), and an unobstructed aortic arch. Pulmonary vascular resistance index was normal at 1.30 Wood-units/m². These reassuring hemodynamics suggested the LV was handling an adequate

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**FIGURE 1.** Neonatal echocardiogram showing (A) small LV and severe mitral regurgitation as well as (B) juxtaductal aortic coarctation and arch hypoplasia. Cardiac MRI at (C) 4 months and (D) 13 months of age demonstrating interval LV growth from 19.0 mL/m² to 57.4 mL/m². MRI stills acquired at end diastole. LV, Left ventricle; MRI, magnetic resonance imaging.
cardiac output without elevated filling pressures. Thus, a biventricular conversion operation was performed including Damus–Kaye–Stansel takedown, Sano take-down, and reestablishment of aortic and pulmonary continuity including bilateral pulmonary arterioplasty (Video 1). Postoperative echocardiography revealed normal biventricular function, trivial aortic valve stenosis (14 mm Hg peak gradient), and trace MR and mild mitral stenosis (5 mm Hg peak gradient). The patient was discharged on POD5 after an uncomplicated hospital stay. At 3-month follow-up, routine echocardiogram revealed normal biventricular function, mild MR, and focal left pulmonary artery stenosis for which balloon angioplasty is scheduled.

COMMENT

There are many strategies for recruitment of the borderline ventricle. This case illustrates the feasibility of staged LV recruitment by fenestrated atrial septation and upsizing of the Sano shunt, which together create favorable physiology with LV volume loading and high systemic saturations. This approach obviates the need for creation and takedown of Super Glenn shunts, leading to simplified recruitment and biventricular conversion operations. Although we did not observe detrimental effects on the RV with Sano upsizing, RV dilation due to upsized conduit regurgitation is a theoretical consideration, and our approach may be best for patients undergoing a shorter LV recruitment. Children undergoing a lengthier LV recruitment may instead be better served with a Super Glenn, which avoids this concern and may also lead to more stable oxygen saturations over time as the Glenn connection grows with the patient. Further study is required to determine the reproducibility and characteristics of the ideal patient for this approach.

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