

Left Ventricular Rehabilitation for Biventricular Outcome in HLHS

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Background: Marked hypoplasia of the left ventricle (LV) or mitral valve is ordinarily managed with a surgical strategy resulting in single functional right ventricle and cavopulmonary connection. In a subset of these patients, however, the LV may be amenable to maneuvers to encourage its growth with the eventual goal of restoring biventricular circulation.

Methods: Beginning in 2009, six patients have been selected to undergo left ventricular rehabilitation (LVR) procedures in addition to single ventricle management. Clinical records of these six patients were reviewed.

Results: Procedures applied to encourage LV growth included resection of LV endocardial fibroelastosis (9 operations in 5 patients), mitral valvotomy/repair (4 operations in 1 patient), aortic valvotomy/repair (11 operations in 6 patients), fenestrated atrial septal defect closure (6 patients), and aortic root augmentation (5 operations in 4 patients). One patient underwent superior cavopulmonary connection and two underwent upsizing of the right ventricle to pulmonary artery connection; one had previous Fontan. Of the six, two have undergone takedown of the Norwood with restoration of biventricular circulation, one at 14 months after initial LVR operation, and one at 33 months after Norwood with LVR. One experienced failure of LV rehabilitation and proceeded with cavopulmonary connection. This patient died suddenly 19 months later. Three patients are currently alive with progressive LV growth, at 20 months, 8 months, and 1 month after Norwood/LVR operation.

Conclusions: In a subset of patients with HLHS, additions to standard single ventricle surgical management can result in growth of LV structures and may result in eventual biventricular circulation.