

Variable Myocardial Response to Load Stresses in Infants with Single LV Anatomy: Influence of Initial Physiology and Surgical Palliative Strategy

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Introduction: Initial surgical strategies in neonates with single left ventricular (LV) anatomy vary based on adequacy of pulmonary and systemic blood flow. Differing myocardial responses to these strategies, as reflected in indices of systolic function, ventricular size, and mass have not been well defined. We hypothesized that the initial surgical palliation based on presenting physiology will impact post-operative indices of LV size, mass, and function.

Methods: Infants presenting with single LV physiology (double inlet LV or tricuspid atresia) from January 1999-June 2007 were divided into groups based on neonatal palliation: no palliation/PA band (NO group); BT shunt only (BT); and Norwood procedure (NP). Echo measures were obtained at presentation, early post-bidirectional Glenn (BDG), late post-BDG follow-up, and post-Fontan procedure if available. Measures included ejection fraction (EF), indexed LV mass (LVMI) and end diastolic volume (EDV), and mass/volume (M/V) ratio.

Results: The cohort included 38 children, with 13 in the NO group, 13 in BT group, and 12 in NP group. Thirty-two subjects had adequate studies for measurement at all four time points. EF was similar but depressed in all groups at all stages, with a significant decline in all groups early after BDG followed by gradual improvement to mildly depressed function (mean EF 48-55%) post-Fontan. LVMI increased significantly in the NP group early post-BDG and remained elevated at late post-BDG follow-up compared to the NO and BT groups, but this difference resolved by the post-Fontan study with mass estimates in the normal range for all groups (LVMI 38-40 g/m^{2.7}). LV EDV was similar with mild chamber dilatation in all groups after BDG. This dilatation was most striking in the NP group, with significant and progressive LV chamber enlargement late after BDG (from 70 mL/m² at presentation to 100 mL/m²) that regressed to a similar size compared to the other surgical groups after Fontan. Those with limited pulmonary blood flow (the BT shunt group) had a significantly higher M/V ratio than those with less restricted blood flow (the NO and NP groups) at presentation. There was a significant increase in the M/V ratio in the NO group from presentation to early post-BDG, but the ratio progressively decreased through late BDG follow-up and remained stable post-Fontan. The NP group had the highest M/V ratio and was significantly higher than the NO group (p= 0.03) at late post-BDG.

Conclusions: Ventricular remodeling can be identified by echocardiography in children with single LV physiology, despite variable initial surgical palliative strategies. Importantly, these initial surgical strategies do not result in significant differences in EF, indexed LV volume, LVMI, or M/V ratio after Fontan palliation during early childhood. LVMI regresses to the normal range after the Fontan surgery in all groups, but single LVEF is reduced initially and remains mildly depressed throughout the palliative procedures. LV chamber dilatation is seen early after BDG in all groups but significantly increases only in the NP group. Further longitudinal echocardiographic assessment of single LV size, mass, and systolic function in this group of patients will be important as they remain at long-term risk of myocardial dysfunction given their abnormal physiology and history of multiple surgical procedures.