

Obstructive left heart disease in neonates with a "borderline" left ventricle: diagnostic challenges for choosing the best outcome

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Background: In most newborns with left heart obstruction, the choice between a single ventricle or biventricular management pathway is clear. However, in some neonates with a "borderline" left ventricle, this decision is difficult. Existing criteria do not reliably identify neonates who will have a good long term outlook after biventricular repair (BVR).

Objectives: Prospective assessment of the outcome after BVR of newborns in whom the left ventricle (LV) was considered "borderline" by an expert group.

Methods: Prospective follow up of neonates with obstructive left heart disease associated with a "borderline" LV who underwent biventricular management between January 2005 and April 2011.

Results: Thirteen of 154 (7.8%) neonates who required intervention for left heart obstruction met echocardiographic (ECHO) inclusion criteria. At first and last ECHO, mitral valve Z score was $-1.76 (\pm 1.37)$ and $-0.66 (\pm 1.47)$ ($p=0.013$) respectively, aortic valve $-1.02 (\pm 1.57)$ and $-0.23 (\pm 1.78)$ ($p=0.056$), and LV end-diastolic volume $13.77 (\pm 5.8)$ and $20.85 (\pm 8.9)$ ml/m² ($p=0.006$). All 12 survivors are clinically well. However, LV diastolic dysfunction and pulmonary artery hypertension was present in 5/12 (36%). Endocardial fibroelastosis (EFE) was detected in 5 patients at last echo follow up, but only in 2 preoperatively. Cardiac MRI did not confirm EFE in any of assessed patients.

Conclusions: We were unable to reliably predict outcome after BVR of neonates with left heart obstruction and a "borderline" LV. The presence of EFE with consequent diastolic dysfunction is more important than LV volume in determining the outcome. Prospective identification of EFE remains challenging.