Echocardiographic Assessment of Single Right Ventricular Systolic Function in Children with Hypoplastic Left Heart Syndrome and its Variants

Background:
The accurate assessment of cardiac function for patients with single right ventricles remains critical for their care yet reliable methods for the evaluation of right ventricular systolic function are still being developed. Unlike the left ventricle, which has well-defined tools to assess its systolic function, the right ventricle has long been a challenge for cardiologists due to its unique and complex shape. The purpose of this study was to evaluate the systolic function of the single right ventricle using two-dimensional echocardiographic parameters before and after palliative procedures and to assess if any of these parameters correlated with patient outcomes.

Methods:
Retrospective data analysis was conducted of patients born with single right ventricle anatomy between June 1, 2008 and February 1, 2014. Patients were included if they had any of the following lesions: Hypoplastic Left Heart Syndrome (HLHS), Unbalanced Atrioventricular Canal Defect with hypoplastic left ventricle (UAVCD) or a variant of HLHS. The assessment of the right ventricle included measurement of the area at end-systole and end-diastole and calculation of Fractional Area Change (FAC). Echocardiograms were reviewed at three time points: (1) initially after birth pre-surgical palliation, (2) pre-Stage II palliation and (3) pre-Stage III palliation. The two primary outcomes evaluated were the need for Extracorporeal Membrane Oxygenation (ECMO) and death.

Results:
Twenty-eight patients with single right ventricle cardiac anatomy were included in the study analysis. RV parameters were evaluated using Independent Samples t test. A large effect size was seen between RV end-diastolic area (EDA) pre-Stage II and mortality. Those patients who died were more likely to have a smaller EDA pre-Stage II compared to those that survived following Stage II palliation. Follow up ranged from 1 to 31 months with a median follow up time of 17 months.

Conclusion:
Echocardiographic assessment of the single right ventricle continues to be an on-going challenge. Following RV EDA for these patients during stages of palliation may be a useful tool in determining those with higher risk for mortality.