

Longitudinal assessment of global and segmental right ventricular deformation in children with hypoplastic left heart syndrome through their univentricular palliation

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Introduction: The univentricular palliation treatment in patients with hypoplastic left heart syndrome (HLHS) commonly consists of three stages; Norwood 1, superior cavopulmonary connection (SCPC) and total cavopulmonary connection (TCPC). Following each stage the loading conditions of the right ventricle (RV) change and these may impact on RV deformation. In this study, we evaluated longitudinal changes in myocardial deformation in patients with HLHS. Previous work from our group has shown that RV strain correlates with RV ejection fraction.

Methods. Patients with HLHS who have successfully completed the univentricular palliation at Evelina Children's hospital between 2007-2016 were recruited for this study. RV systolic myocardial function was assessed at pre and post Norwood 1, pre and post SCPC and pre and post TCPC stage. Speckle tracking analysis was performed on apical four chamber views to measure global longitudinal strain (GLS) and segmental deformation. Differences in RV function according to LV morphology (mitral and aortic atresia (MA/AA) vs mitral stenosis and aortic atresia (MS/AA)) were also explored. All measurements were analysed by one observer using Q-lab version 10.5 (Philips Medical Systems, Andover, USA).

Results: Fifty-one patients with HLHS were identified (20 MA/AA and 31 MS/AA). Norwood 1, SCPC and TCPC operations were performed at 0.13 ± 0.09 , 5.39 ± 1.45 and 40.16 ± 7.87 months respectively. None of the patients had significant tricuspid regurgitation. A trend towards highest values of GLS was noted prior to Norwood 1 (Table 1). No difference was observed between MA/AA, MS/AA groups with respect to GLS, whereas lower mid-septal strain was noted in the MS/AA group ($p:0.013$).

Conclusions: In this study we demonstrated for the first time that patients with HLHS with MA/AA, MS/AA followed through Fontan palliation have similar GLS through all three stages of surgical palliation. A trend towards higher GLS in pre Norwood stage was noted which is consistent with RV volume loading prior to initial surgical palliation. The lower mid-septal strain in MS/AA group, possibly reflects their diseased fibrotic myocardium of the LV. These findings indicate that speckle tracking assessment tracks with physiological changes and can be used to monitor RV function in patients with HLHS.

Table 1. Speckle tracking echocardiographic parameters at each stage

Variable	Pre-Norwood	Post-Norwood	Pre-SCPC	Post-SCPC	Pre-TCPC	Post-TCPC	P-value - time effect - group-time effect
Longitudinal							
Global strain (%)	-23.4 ± 4.8	-22.6 ± 2.9	-19.9 ± 3.4	-18.6 ± 3.5	-21.3 ± 3.2	-20.2 ± 3.5	0.051 0.928
PSSi (%)	2.2 ± 3.2	2.0 ± 4.1	1.7 ± 3.5	1.9 ± 3.9	1.4 ± 5.6	2.5 ± 5.5	0.811 0.479
MDI (%)	6.6 ± 8.0	5.1 ± 5.2	5.1 ± 5.4	7.8 ± 6.9	6.3 ± 5.9	7.3 ± 6.4	0.104 0.241

Mean \pm standard deviation

SR, strain rate; PSSi, post systolic strain index; MDI, mechanical dyssynchrony index