

**Impact of aortic elastance on right ventricular function in Hypoplastic Left Heart Syndrome after Fontan palliation.**

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Introduction: Systemic right ventricular (RV) function is the most important determinant of long term outcome in patients with palliated hypoplastic left heart syndrome (HLHS). Abnormal elastic properties of the reconstructed aorta, may negatively impact on ventricular function. We therefore aimed to assess systemic RV function and aortic arterial elastance in palliated HLHS patients to describe the relationship between the two.

Methods: Fifty-six HLHS patients (median age 5.4 (range 2.9-14.2) years) were studied at a median of 2.6 (range 0.8-12.7) years after completion of the Fontan circulation with the pressure-volume conductance system.

Results: Arterial elastance of the reconstructed aorta (Ea) was higher in palliated HLHS patients compared to normal values from the literature [Chen et al., JACC 1998] for the unoperated aorta in a biventricular circulation ( $3.1 \pm 1.1$  vs.  $2.2 \pm 0.8$  mmHg/ml;  $p < 0.01$ ). Ea correlated negatively with RV ejection fraction as an afterload dependent measure of systolic ventricular function ( $r = -0.31$ ,  $p = 0.02$ ). However, load independent systolic RV function measured as end systolic elastance (Ees) increased with increasing Ea ( $r = 0.29$ ,  $p = 0.02$ ). Therefore, ventriculo-arterial coupling (Ea/Ees) remained within the physiological range. End diastolic stiffness (Eed) of the systemic RV showed a positive correlation with Ea ( $r = 0.6$ ,  $p < 0.001$ ).

Patients who needed an intervention for significant coarctation after aortic reconstruction showed higher Ea and Eed even four years after successful coarctation treatment (Ea:  $3.4 \pm 1.2$  vs.  $2.8 \pm 1.0$  mmHg/ml,  $p = 0.04$  and Eed:  $0.67 \pm 0.44$  vs.  $0.45 \pm 0.3$  mmHg/ml,  $p = 0.04$ ).

Conclusion: The elastance of the reconstructed aorta of palliated HLHS patients is abnormally high. Aortic elastance, a determinant of afterload, negatively impacts on ejection fraction but not on load independent systolic myocardial function. Diastolic stiffness is increased when aortic elastance is high. The effects of abnormal aortic elastance on intrinsic systolic function and diastolic stiffness were more pronounced in patients who underwent successful treatment for aortic coarctation.