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). The RV is morphologically not capable of dealing with chronic exposure to the high afterload of the Fontan circulation. Theoretically this creates a mismatch between ventricular function and increased afterload: Abnormal elastic properties of the reconstructed aorta in HLHS may further impact negatively on ventricular function. Therefore we aimed to assess systemic RV function and aortic arterial elastance in palliated HLHS patients to describe the relationship between the two. To delineate the impact of aortic coarctation on ventricular function we created groups according to history of intervention on aortic coarctation.

Patients und Methods:
56 HLHS patients (median age 5.4 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 (PV) conductance system. PV loops were recorded at baseline and during dobutamine infusion: A preload reduction manoeuvre was performed to analyze intrinsic RV function.

Results:

<table>
<thead>
<tr>
<th>Ea vs.</th>
<th>r</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>EF, %</td>
<td>-0.31</td>
<td>0.02</td>
</tr>
<tr>
<td>SW, mmHg x ml</td>
<td>-0.66</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Ees, mmHg/ml</td>
<td>0.29</td>
<td>0.04</td>
</tr>
<tr>
<td>Eed, mmHg/ml</td>
<td>0.60</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Ea correlated negatively with afterload dependent measure of systolic ventricular function (EF and SW).

Load independent systolic RV function (Ees) increased with increasing Ea.

End diastolic stiffness (Eed) of the systemic RV showed a negative correlation with Ea.

Ea of the reconstructed aorta was higher in palliated HLHS patients compared to normal values from the literature for the unoperated aorta in a biventricular circulation.

6.1±1.1 vs. 2.2±0.8 mmHg/ml; p<0.01

Chen CH et al., J Am Coll Cardiol, 1998

Conductance derived parameters:
- Ees: End systolic elastance, slope of the end systolic PV relationship (Ees: ESP vs. ESV)
- Eed: End diastolic stiffness, slope of the end diastolic PV relationship (Eed: EDP vs. EDV)
- Ea: Arterial elastance, end systolic pressure divided by stroke volume (Ea = ESP/SV)
- Ventriculo-arterial coupling: Ea / Ees

SW: stroke work, PV loop area

 Patients who needed an intervention for significant coarctation after aortic reconstruction showed higher Ea, Ees and Eed even four years after successful coarctation treatment.

Constitution:
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.