MRI catheter stress haemodynamics in hypoplastic left heart syndrome after Fontan completion

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Introduction Effort intolerance and signs of a failing circulation are common following the Fontan procedure. We studied the haemodynamics of the Fontan circulation using MRI catheterisation at baseline and maximal pharmacological stress.

Methods Prospective study of children with hypoplastic left heart syndrome (HLHS) post Fontan referred for MRI catheter due to signs or symptoms of a failing Fontan, poor cardiopulmonary exercise test performance or abnormalities on routine MRI. Quantification of volumetrics and flow was from MRI with simultaneous invasive catheter pressure measurements. Measurements were made during baseline conditions (Stage 1), dobutamine at 10mcg/kg/min (Stage 2) and 20mcg/kg/min (Stage 3). Control data was from 10 healthy adults with biventricular hearts without invasive measurements.

Results Eleven patients recruited; median age 9.47years (3.54-11.1yrs). All reported exercise intolerance. Two had plastic bronchitis, of whom 1 also had protein losing enteropathy. One patient only received 10mcg/kg/min dobutamine due to an elevated blood pressure response. There were no reported adverse events.

Median heart rate (HR) increased from 80bpm to 162bpm (p <0.05) in HLHS, against 67bpm to 119bpm (p<0.05) in controls. In HLHS, indexed cardiac output (iCO) was 2.8L/min/m², increasing by 53% from baseline to 4.3L/min/m² (p<0.05) at stage 2 matching controls (Figure 1). However there was no significant further increase at stage 3 (p=0.21), with iCO of 4.4L/min/m² despite increased HR and ejection fraction (EF). There was a significant fall in indexed end diastolic volume (iEDV) during stage 2 and 3 not seen in controls (Figure 2). The maximal fall in indexed end systolic volume (iESV) from baseline was similar in both groups at 50% in HLHS and 54% in controls.

Mean baseline transpulmonary gradient in HLHS was 3.5mmHg±1.12 with a pulmonary vascular resistance (PVR) of 1.63wu.m²±0.55. Mean resting right ventricular end diastolic pressure (EDP) in HLHS was 6.5mmHg±2.8.

Conclusion There is good systolic reserve in response to dobutamine stress. The rise in cardiac output is blunted at higher dose dobutamine in the HLHS Fontan. As the resting ventricular EDP is normal, we hypothesize that this is due to an inability to appropriately increase blood flow through the pulmonary circulation and hence ventricular preload despite low resting PVR.