Fontan circulation shows deranged relative intra-cardiac blood flow components but normal kinetic energy profile

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Introduction:
Ventricular function assessment is an important part of the assessment of the Fontan circulation. Recent advances in 4D flow cardiovascular magnetic resonance imaging now allow detailed assessment of left ventricular intra-cardiac flow patterns and kinetic energy (KE). A previous study has shown abnormal flow parameters in adults with dilated cardiomyopathy and low normal ejection fraction. We hypothesised that flow changes may already be apparent in the ventricle in Fontan patients prior to a decline in ejection fraction.

Methods:
We prospectively enrolled 16 participants (8 Fontan patients and 8 age and sex matched controls) who underwent 3T CMR assessment. For analysis the ventricular volume was divided into 4 functional components; direct flow (DF), delayed ejection flow (DEF), retained inflow (RI) and residual volume (RV). For each component the volume was calculated and expressed as percentage of end diastolic volume and the kinetic energy was calculated over the cardiac cycle and measured at end diastole.

Results:
The mean age was 22 years (range 14-35). Mean ejection fraction was 57% in the Fontan patients and 67% in the healthy volunteers. 6 Fontan patients had an atrio-pulmonary Fontan connection and the remaining 6 had a total cavopulmonary connection.
Compared to healthy volunteers, Fontan patients had significantly reduced direct flow (28% vs 40%; p=0.018) and increased residual volume (38% vs 28%; p=0.019). This was less apparent in the 2 patients with a single ventricle and intact septum.
The kinetic energy was comparable in both groups (KE direct flow in Fontan 2.5x10^{-4} vs healthy volunteers 2.7 x10^{-4} mJ; p=0.84).

Conclusions:
This study shows that ventricular 4D flow assessment is feasible in complex congenital heart disease. Fontan patients show less efficient blood flow patterns than healthy volunteers. Unlike adults with cardiomyopathies, the Fontan patients had normal kinetic energy values. This may suggest that ventricles in the Fontan circulation are different from adults with ventricular dysfunction; the anatomical ventricular geometry may impact on the efficiency of blood flow but less on kinetic energy profiles. This new technique may allow novel insights into the pathophysiology of ventricular dysfunction in the Fontan circulation and may provide novel imaging biomarkers.