Pitfalls of the Fontan circulation during exercise; a modeling study

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Background - The Fontan palliation, used as surgical repair method for many life threatening complex congenital heart diseases, creates a univentricular serial circulation. Large variation of pathologies treated with this palliation makes that current insight in cardiovascular pathophysiology of exercise limitation in these patients is limited. We used a multi-scale computational model of the heart and circulation to explore the pathophysiology of exercise limitation in Fontan patients.

Methods - The model simulates beat-to-beat dynamics of the two cardiac cavities, the valves, and the systemic and pulmonary circulations. The univentricular circulation in rest and exercise was simulated. We evaluated the extreme situations of cardiac output (CO) increase by exclusive increase of either stroke volume (SV) or heart rate (HR).

Results and Discussion – Central venous pressure (CVP) rose independent of the fact whether CO increase was due to HR or SV increase, but CVP rose more with SV increase (Figure). The large end-diastolic volumes that accompanied SV increase, required higher ventricular filling pressure, hence, higher atrial pressure (AP). Because pulmonary resistance was not allowed to change, pulmonary pressure drop and, hence, CVP had to rise to enable the increase in pulmonary flow. Consequently, systemic pressure drop reduced, implying further decrease of either systemic resistance or flow. Limited ability to decrease systemic resistance or to cope with increased CVP may underlie reduced exercise capacity in Fontan patients.

Conclusions Our simulations suggest that limitation of exercise capacity in Fontan patients is primarily due to increase of CVP. The fact that CVP is more sensitively increased with SV than with HR may explain why bradycardia is not well tolerated.

Increase of cardiac output resulted in increase of atrial pressure (AP) and central venous pressure (CVP). The pulmonary pressure drop ($\Delta p_{\text{pulm}}$) rose to maintain pulmonary flow, resulting in a vastly decreased systemic pressure drop ($\Delta p_{\text{sys}}$).