

Assessment of intraatrial lateral tunnel anatomy and venous blood flow in children with hypoplastic left heart syndrome in Fontan circulation

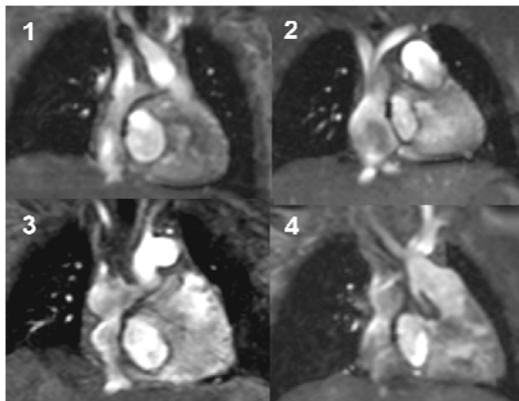
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Introduction: In patients (pts) with hypoplastic left heart syndrome (HLHS) post completion of the total cavopulmonary connection (TCPC) with an intraatrial lateral tunnel, deviations of the tunnel from an ideal tubular shape are common. However, little is known about frequency and potential adverse effects of such shape deviations. We sought to analyze tunnel anatomy, dimensions and blood flow with magnetic resonance imaging (MRI).

Methods: Sixty-one HLHS pts (mean age 6.4 ± 2.7 years) underwent 3.0-T MRI with gradient-echo cine sequences, 2D- and flow-sensitive 3D-phase-contrast MRI. We analyzed anatomy, diameters, cross-sectional areas and volumes of the tunnel. Tunnel blood flow was measured at the level below the connection of the inferior vena cava (IVC) with the pulmonary arteries.

Results: 23 pts had a tubular-shaped tunnel (Figure 1). In 28 pts bulging and/or narrowing at different locations of the tunnel was present (Figure 2-4). In 10 pts a classification was not possible because of artifacts from implanted devices. Cross-sectional areas, tunnel volumes, the mean blood flow and the mean and maximal flow velocity were not significant different between pts with a tubular tunnel and pts with shape deviations of the tunnel. We found a relation between the normalized tunnel volume and age ($r=0.44$; $p=0.002$), body surface area ($r=0.42$; $p=0.005$) and time after TCPC ($r=0.42$; $p=0.001$). The mean tunnel blood flow correlated with age ($r=0.75$; $p=0.001$) and body surface area ($r=0.83$; $p<1.0e-4$). Flow-sensitive 3D-phase-contrast MRI showed retrograde flow at the junction between the IVC and the tunnel and a non-linear tunnel blood flow (e.g. reflux, vortices) below the fenestration.



Conclusion: 1) The volume and the mean blood flow of the intraatrial lateral tunnel correlated with age and body surface area of HLHS pts in Fontan circulation, suggesting that the conduit capacity of the tunnel adjusts to body growth, unlike an extracardiac tube. 2) Flow-sensitive 3D phase-contrast MRI showed a non-linear blood flow in the lower part of the tunnel. Follow-up MRIs are needed to detect long term effects of irregular tunnel shapes on flow dynamics.