Determinants and Clinical Significance of Flow via the Fenestration in the Fontan Pathway: A Multimodality Study

The Hospital for Sick Children, Toronto, Canada

Introduction:
The use of a fenestration in the Fontan pathway remains controversial, partly because its hemodynamic effects and clinical consequences are insufficiently understood. The objective of this study was to quantify the magnitude of fenestration flow and to characterize its hemodynamic determinants approximately 1 year after surgery.

Methods:
Twenty three patients with a fenestrated extracardiac conduit prospectively underwent investigation by cardiac magnetic resonance (CMR), echocardiography, and invasive manometry under the same general anesthetic 12±4 months after Fontan surgery. Fenestration flow was determined using phase contrast CMR either by subtracting flow in the Fontan pathway above the fenestration from Fontan flow below the fenestration or by direct measurement (12 patients, Figure).

Results:
Measured and calculated fenestration flows showed an excellent agreement (r=0.92, p<0.0001, Figure). Fenestration flow constituted a mean of 31±12% (range 8-50%) of ventricular preload. It was associated with a lower Qp/Qs (r=-0.64, p=0.001) and oxygen saturation (r=-0.74, p<0.0001). Fenestration flow volume was correlated with pulmonary vascular resistance (r=0.45, p-0.04) and markers of ventricular diastolic function (early diastolic strain rate r=0.57, p=0.008 and ventricular untwist rate r=0.54, p=0.02).
In 14 patients (61%) all of the net inferior vena cava flow and part of the superior vena cava flow were diverted into the systemic atrium and did not reach the lungs. The magnitude of contribution of fenestration flow to ventricular preload was the most important predictor of ability to close the fenestration.

Conclusions:
Fenestration flow can be measured accurately with CMR. In most patients the fenestration delivers most or all of the inferior vena cava flow to the systemic circulation, and in two thirds of patients some of the superior vena cava flow is also diverted through the fenestration. Therefore, the pulmonary blood flow increases only minimally and sometimes even decreases after the fenestrated Fontan operation. The amount of fenestration flow is related both to the pulmonary vascular resistance and systemic ventricular diastolic ventricular function, and may be used to predict hemodynamic suitability for fenestration closure.