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Blood flow dynamics in the total cavopulmonary connection long-term after Fontan completion

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Citation
Rijnberg, F. M. (2023, December 20). *Blood flow dynamics in the total cavopulmonary connection long-term after Fontan completion*. Retrieved from https://hdl.handle.net/1887/3674148

Version:	Publisher's Version
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CHAPTER 7

7

Assessment of extracardiac conduit adequacy

CHAPTER 7A

Hemodynamic consequences of an undersized extracardiac conduit in an adult Fontan patient revealed by 4D flow MRI

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Circulation Cardiovascular Imaging 2021

Hemodynamic consequences of an undersized extracardiac conduit in an adult Fontan patient revealed by 4D flow MRI.

A 20 year old female patient born with a double inlet left ventricle with transposition of the great arteries previously underwent a bidirectional Glenn shunt at the age of 7 months. Final completion of the Fontan circulation (total cavopulmonary connection, TCPC) was performed at the age of 3 years and a body surface area of 0.6m^2, by connecting the subhepatic IVC and hepatic veins (HVs) with the pulmonary arteries (PAs) using a rigid 16mm extracardiac Goretex conduit. Seventeen years after Fontan completion (body surface area 1.8m^2), an MRI examination was performed as part of routine follow-up including 4D flow MRI. A 3D reconstruction of the TCPC revealed a small, flattened extracardiac conduit (**Figure 1A**, CAAS v5.2, MR Solutions, Pie Medical Imaging). Furthermore, an important dilatation of the distal SVC was observed. The cross-sectional area of the subhepatic IVC and conduit were 259mm^2 and 186mm^2, respectively. The flow rate at the subhepatic IVC and conduit were 1.7 and 2.5 L/min, respectively, indicating a contribution of 0.8 L/min (32% of total conduit flow) from the splanchnic circulation through the HVs. A 2.1-fold increase in mean velocity from the subhepatic IVC (11cm/s) towards the extracardiac conduit (23cm/s, **Figure 1B**) was present, indicative of an undersized extracardiac conduit (IVC-conduit velocity mismatch). While most of the conduit flow is directed towards the left PA, part of the accelerated conduit flow formed a swirling, vortical flow pattern intruding into the distal part of the SVC before reaching the right PA **(Figure 1B, Supplemental Video 1).** The entry of conduit flow into the SVC was associated with the area where the SVC was strongly dilated; the cross-sectional area of the dilated part of the SVC was a 2.4-fold higher compared to the level of the SVC just proximal to the dilatation (204mm^2 vs 481mm^2). Quantification of wall shear stress and viscous energy loss rate derived from 4D flow MRI revealed insights into the hemodynamic consequences of these altered flow patterns. Firstly, the increase in blood flow velocity from the subhepatic IVC and HVs towards the conduit extended into the LPA, leading to areas of elevated wall shear stress (WSS) in the conduit and LPA **(Figure 1C).** These areas with elevated WSS strongly correlated with areas of inefficient blood flow with an increased viscous energy loss rate **(Figure 1D, Supplemental Video 2).** The entry of accelerated conduit flow into the SVC did not result in an important increase in WSS, but rather resulted in the dilatation of the distal SVC. The venous tissue of the SVC is likely more susceptible to increased mechanical stresses caused by the accelerated conduit flow entering the SVC, resulting in adaptive vessel dilatation to keep WSS within a narrow, physiological range. The arterial wall of the LPA may be less susceptible to increased mechanical stresses caused by the accelerated conduit flow, and therefore increased levels of WSS and viscous energy loss are observed without adaptive dilatation of the pulmonary artery.

Single ventricle heart defects represent the most severe end of the spectrum of congenital heart disease, with the Fontan-circulation as a palliative approach. Currently most centers complete the Fontan circulation at the age of 3 to 5 years with the use of an extracardiac conduit[1], as was performed in this case. The main drawback of the extracardiac conduit Fontan technique is the lack of growth potential of the Goretex conduit. Therefore, ideally an oversized conduit is implanted to avoid future somatic overgrowth requiring conduit replacement. Currently, however, it is not known which conduit size is ideal for adult Fontan patients, despite the fact that optimal conduit sizing is of utmost importance to ensure efficient blood flow with minimal energy loss[2]. Regular echocardiographic qualitative assessment of the conduit during follow-up is often only able to identify patients with a distinct focal stenosis or thrombus. Blood flow obstruction due to an undersized conduit is often not recognized with echocardiography and may only become apparent during invasive angiography.[3] This case shows how 4D flow MRI reveals that the 16mm extracardiac conduit has become relatively undersized 17 years after Fontan completion, a timeframe in which body size tripled. The undersized conduit led to accelerated flow in the conduit, associated with increased viscous energy losses affecting the efficiency of blood flow towards the PAs. Because of the absence of a subpulmonary ventricle in Fontan patients and thus a relatively passive pulmonary blood flow, areas with increased energy loss are undesirable.[4] Furthermore, important downstream effects of the accelerated conduit flow were observed, with conduit flow entering the SVC leading to dilatation of the vessel. The competitive flow from the conduit into the SVC may pose increased afterload for SVC flow.

This case raises important concern on the hemodynamic adequacy of the 16mm conduit used in the Fontan circulation and highlights how 4D flow MRI derived blood flow and hemodynamic parameters provide intuitive information about the hemodynamic performance of the TCPC during follow-up. We recommend regular evaluation of the TCPC with 4D flow MRI during somatic growth to allow for early identification of patients with inadequately sized conduits leading to adverse TCPC hemodynamics. As such, this case illustrates the additional role of 4D flow MRI as an in vivo, non-invasive screening tool that can identify patients that may require further invasive hemodynamic evaluation and possible intervention.

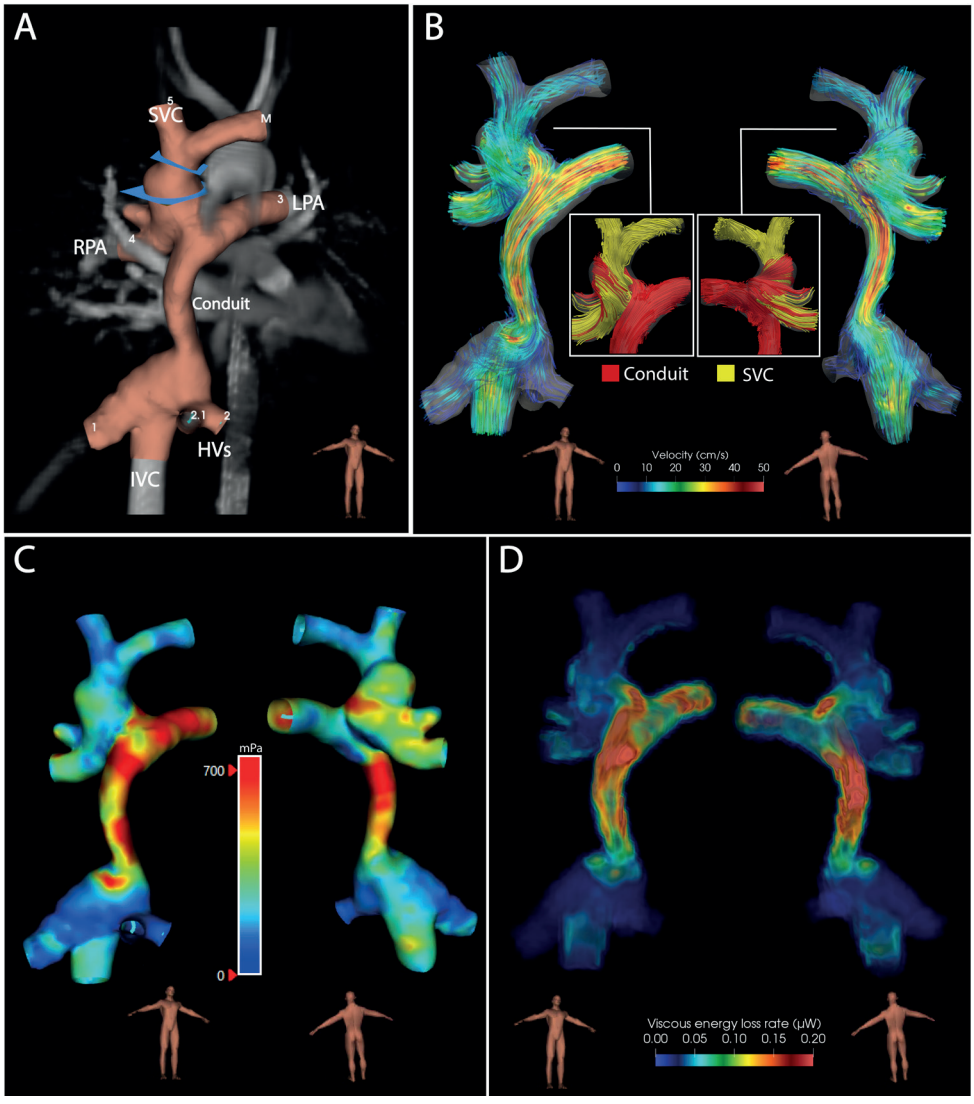

Figure legend. The 3D reconstruction of the TCPC is presented, including the subhepatic IVC and HVs **(A)**. The blue planes indicate the two levels where the cross-sectional area of the SVC is measured; at the dilatation and just proximal to the dilatation. Velocity color-coded streamlines illustrate acceleration of blood flow from the level of the subhepatic IVC towards the conduit extending into the LPA. This accelerated conduit flow partially enters the distal SVC into a swirling flow pattern, correlating with the area of SVC dilatation **(B).** The accelerated blood flow resulted in areas of elevated WSS in the extracardiac conduit and anterior LPA **(C)**, illustrating the relative undersized extracardiac conduit in this patient. No elevated WSS was observed in the dilated SVC. The areas of increased WSS strongly associated with areas of increased viscous energy loss rate indicative of decreased flow efficiency **(D).**

HVs; hepatic veins, IVC/SVC; inferior/superior vena cava, RPA/LPA; right/left pulmonary artery. mPa; millipascal, µW; microwatt

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