Images in Congenital Cardiac Disease

Leaving antegrade pulmonary blood flow results in reversal of flow in the azygous vein and decompression of the cavopulmonary circulation: mechanism of cyanosis defined with cardiac magnetic resonance imaging

Shafkat Anwar, Kevin K. Whitehead, Matthew A. Harris

Department of Pediatrics, Division of Cardiology, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America

Abstract We report the case of a 2.5-year-old patient with single-ventricle physiology who underwent cardiac magnetic resonance study for cyanosis after Kawashima operation. Magnetic resonance imaging study showed a veno-venous collateral redirecting systemic venous flow, responsible for cyanosis.

Keywords: single ventricle; cardiac MRI; cyanosis; Kawashima

Received: 18 August 2013; Accepted: 20 October 2013; First published online: 18 December 2013

THE PATIENT IS A 2.5-YEAR-OLD BOY WITH DOUBLE-outlet right ventricle, moderate pulmonic stenosis, and interrupted inferior vena cava with azygos continuation to the superior vena cava. He had a Kawashima procedure consisting of a superior cavopulmonary connection. As a part of the procedure, antegrade pulmonary flow was

preserved to augment Kawashima flow and prevent the development of arteriovenous malformations by distributing hepatic venous blood to both lungs. He subsequently underwent a cardiac magnetic resonance study to investigate greater than expected cyanosis after Kawashima, oxygen saturation 83%.

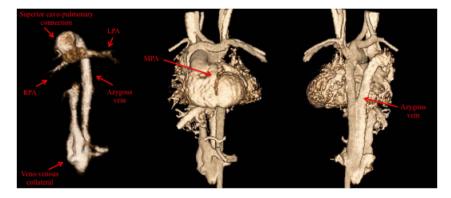


Figure 1. Three-dimensional volume rendering of veno-venous collateral from superior vena cava (SVC) draining inferiorly via azygous vein back to the beart. LPA = left pulmonary artery; MPA = main pulmonary artery; RPA = right pulmonary artery

Correspondence to: S. Anwar, Department of Pediatrics, Division of Cardiology, The Children's Hospital of Philadelphia, 34th Street and Civic Center Blvd. Room 8NW37, Philadelphia, Pennsylvania, United States of America. Tel: 202-486-6575; Fax: 314-454-2561; E-mail: Shafkat.md@gmail.com

Cine imaging and phase contrast velocity mapping showed antegrade main pulmonary artery flow providing nearly the entire flow to the branch pulmonary arteries. Time-resolved magnetic resonance angiogram (Supplementary video: S1) demonstrated superior vena cava flow decompressing caudally via the azygous vein to a large systemic to hepatic venovenous collateral system that ultimately returned to the heart via hepatic veins (Fig 1, Supplementary videos: S1 and S2). Flow heading caudally in the azygous vein at the diaphragm was equal to the flow measured in the superior vena cava. Presumably, antegrade main pulmonary artery flow led to redirection of Kawashima flow caudally with the development of a large veno-venous collateral that was responsible for the patient's cyanosis. This report underscores an important complication of leaving antegrade pulmonary flow in superior cavopulmonary connection patients, especially in the setting of a patent azygous system. The case also shows the utility of cardiac magnetic resonance imaging in delineating complex anatomy and physiology.

Acknowledgement

The authors wish to thank Dr. Mark A. Fogel, Director of Cardiac MRI, The Children's Hospital of Philadelphia.

Financial Support

This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest

None.

Supplementary material

To view supplementary material for this article, please visit http://dx.doi.org/10.1017/S104795111300214X

Reference

 Gérelli SS, Boulitrop CC, Van Steenberghe MM, et al. Bidirectional cavopulmonary shunt with additional pulmonary blood flow: a failed or successful strategy? Eur J Cardiothorac Surg 2012; 42: 513–519.