

## 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

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**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

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**T**HE 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 heart. LPA = left pulmonary artery; MPA = main pulmonary artery; RPA = right pulmonary artery

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,<sup>1</sup> 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.

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### Conflicts of Interest

None.

### Supplementary material

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

### Reference

1. 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.