Rare venous connection causing severe hypoxia after Fontan operation

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Abstract

We describe a rare case of cyanosis following the Fontan operation with right-to-left shunting at the venous level, that is, an azygos venous-to-pulmonary venous connection. Few cases with partial anomalous pulmonary venous connection to azygos vein have been reported; however, there have been no reports describing the connection from azygos vein to pulmonary vein, which results in desaturation after Fontan operation. Multidetector-row computed tomography (MDCT) was a useful tool to depict this vascular malformation. © 2008 Published by European Association for Cardio-Thoracic Surgery. All rights reserved.

Keywords: Fontan operation; Cyanosis; Collateral vessel

1. Introduction

Persistent desaturation after a Fontan operation usually results from right-to-left shunting at the site of the atrial baffle or within the lung through pulmonary arteriovenous malformations. Recently, we treated a rare case of right-to-left shunting at the venous level, with an azygos venous to pulmonary venous connection. This is the first report showing the occurrence of desaturation through this abnormal venous connection after a Fontan operation.

2. Case report

An 11-year-old girl with right atrial isomerism after total cavopulmonary connection (TCPC) has continued hypoxia. She was born with transposition of the great arteries (situs inversus, l-loop, l-malposition [I,L,L]), dextrocardia, pulmonary stenosis, mitral atresia, straddling tricuspid valve, ventricular septal defect, atrial septal defect, bilateral superior vena cava (SVC), and a total anomalous pulmonary venous connection (TAPVC) draining into the left superior vena cava at its junction with the atrium.

She underwent a hemi-Fontan operation with a TAPVC repair at the age of two years. The vertical vein was ligated and common pulmonary vein chamber was directly anastomosed to the atrium. At four years, the modified Fontan operation (intracardiac total cavopulmonary connection) was completed. In the postoperative period, she continued to be cyanosed; her systemic oxygen saturation was 80–85%.

To identify a residual systemic vein draining into the atrium, she underwent cardiac catheterization at eight years; however, an angioagram of the bilateral SVC and inferior vena cava (IVC) at the level of the renal vein demonstrated no residual right-to-left shunt.

Multidetector-row CT (MDCT; Aquillian 16, Toshiba Corporation, Medical system company, Tokyo, Japan) angio-graphy was performed at the age of 11 years. Contrast enhancement was achieved through a 22-gauge catheter into a saphenous vein. MDCT demonstrated that a tortuous enlarged left lumbar vein and an ascending lumbar vein were connected to the azygos vein, which returned blood to the left pulmonary vein (Fig. 1). Furthermore, on MDCT, the left pulmonary vein was loop-shaped. A femoral venous angiogram confirmed that it flowed upward to the left lumbar veins and an ascending lumbar vein draining into an azygos vein, which returned blood to the left pulmonary vein (Video 1).

After surgical ligation of the azygos vein, systemic saturation increased to the high 90s, and she continues to do well one year after her operation.

3. Discussion

The indications of Fontan operation have been extended to complex anomalies including visceral heterotaxy [1]. Because there are various types of systemic venous connec-tions in patients with visceral heterotaxy [2, 3], unantici-pated abnormal systemic venous channels may pose additional problems after the operation. We reported a rare case of cyanosis following the Fontan operation with right-to-left shunting through an azygos vein connected to pulmonary vein. This is a hazardous anomaly in a Fontan candidate and should be considered in those with unexpected postoperative desaturation.

The incidence of partial anomalous pulmonary venous connection (PAPVC) is 0.4–0.7% of all autopsy cases, PAPVC is not a rare disease, but the location of the anomalous
Multidetector-row CT (MDCT) angiography was performed to identify a residual systemic vein draining into the atrium. Contrast was injected through a saphenous vein. MDCT showed that a tortuous enlarged left lumbar vein and an ascending lumbar vein were connected to the azygos vein, which returned blood to the left pulmonary vein. After surgical ligation of the azygos vein, systemic saturation increased to the high 90s.

Video 1. A conventional angiography confirmed that contrast medium from the femoral vein flowed upwardly to the left lumbar vein and azygos vein, which was connected to the left pulmonary vein.

MDCT is a non-invasive diagnostic tool that gives useful anatomical information about extracardiac vascular malformations [5, 6]. In addition, it may obviate the need for angiography, and be an important modality in the diagnosis and management of congenital heart disease.

References