

# Conversion from Total Cavopulmonary Shunt to Fontan Circulation: Improved Cyanosis with an 11-Year Interval

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**We experienced a conversion case from total cavopulmonary shunt (TCPS) to the Fontan circulation with an 11-year interval in the left isomerism and an interruption of the inferior caval vein. Marked cyanosis (pulse oximetry measuring 74%) progressed 8 years after TCPS because of the prominent development of bilateral pulmonary arteriovenous fistula (PAVF), which was diminished after the Fontan conversion, and recent pulse oximetry measured 95% 22 months after the conversion. We believe that this report is a case with the longest interval from TCPS to Fontan in which PAVF was diminished and marked cyanosis was improved. (Ann Thorac Cardiovasc Surg 2008; 14: 29–31)**

**Key words:** total cavopulmonary shunt, Fontan conversion, pulmonary arteriovenous fistula, left isomerism, hepatic vein

## Introduction

Residual cyanosis can frequently progress after total cavopulmonary shunt (TCPS) because of the development of pulmonary arteriovenous fistula (PAVF) in patients with left isomerism and interruption of the inferior caval vein,<sup>1</sup> and there have been some reports that a redirection of hepatic venous drainage could improve cyanosis efficiently with a 30–80 month interval.<sup>2</sup> We experienced a conversion case from TCPS to the Fontan circulation in left isomerism with a 130-month interval in which PAVF was diminished and cyanosis was improved.

## Case Report

In February 2005, we performed an extracardiac Fontan

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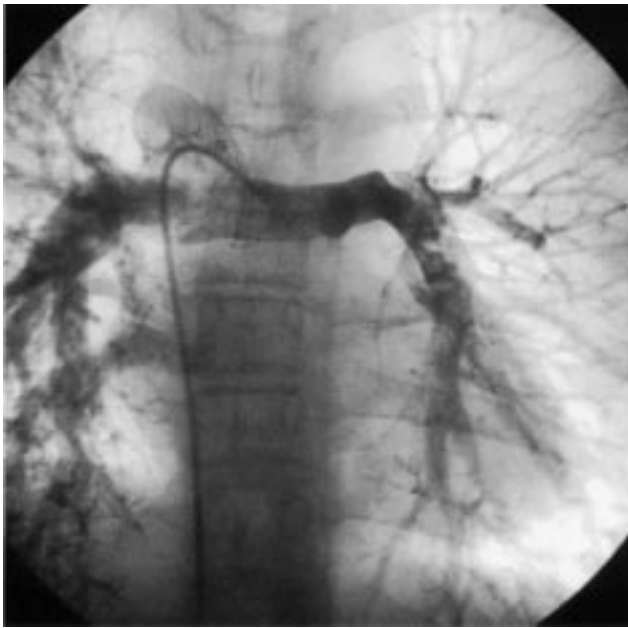
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operation on a 12-year-old girl (50 kg body weight) with the following diagnosis: polysplenia, hypoplastic left heart syndrome, azygos connection of the inferior vena cava, and persistent left superior vena cava. At 1 month, the Norwood procedure was performed, and at 1 year of age the patient underwent TCPS. A subsequent serial follow-up revealed an asymptomatic child developing normally with pulse oximetry measuring around 90%. No further operation was planned to include hepatic venous flow to the pulmonary circulation because the child had progressed normally without symptoms.

However, marked cyanosis (pulse oximetry measuring 74%) had progressed by the age of 9 years. Catheterization prior to an establishment of the Fontan circulation showed prominent bilateral PAVF (especially PAVF in the right lung is typical) at 11 years of age (Fig. 1), and 9 mm Hg of mean pulmonary arterial pressure, 262 on the Nakata Index,<sup>3</sup> and 0.61 and 9 mmHg of ejection fraction and end diastolic pressure of the right ventricle, respectively. Prior to the conversion, major abnormal communication was not identified between the azygos vein and either the hepatic or the portal vein.

An operation was performed at 12 years of age with

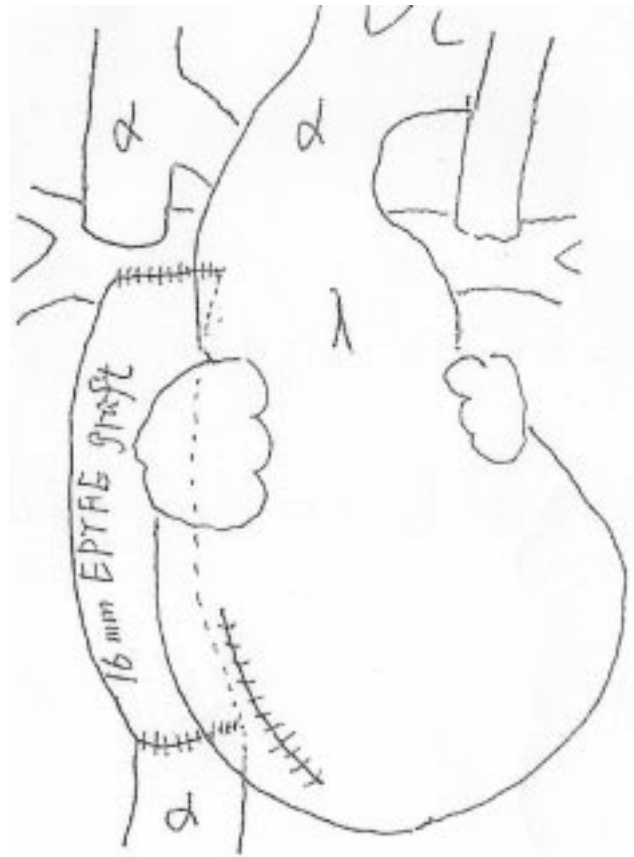


**Fig. 1.** An angiography of the right superior vena cava anastomosis to the pulmonary arteries shows marked pulmonary arteriovenous fistula before the conversion (especially PAVF in the right lung is typical).

the heart beating under a cardiopulmonary bypass. A 16-mm expanded polytetrafluoroethylene (EPTFE) conduit was used for the redirection of the hepatic vein interposing between the hepatic venous orifices and the pulmonary artery (Fig. 2). No fenestration was made in the conduit, and the immediate postoperative central venous pressure was 15 mmHg. The postoperative course was uneventful, and the pulse oximetry measured 86% at discharge. Cardiac catheterization 6 months later revealed significant improvement in mixed venous saturation to 75%, systemic arterial saturation to 94%, and 14 mmHg of mean pulmonary artery pressure. A repeat pulmonary angiogram and contrast echocardiogram demonstrated diminished PAVF in both lungs (Fig. 3). The patient is doing well 22 months after this operation, and recent pulse oximetry measured 95%.

## Discussion

Although TCPS is a surgical option for polysplenic patients with interrupted inferior vena cava and azygos or hemiazygos continuation, residual cyanosis can frequently progress after TCPS because of the development of PAVF.<sup>1)</sup> Recent findings may support speculation that the presence of polysplenia and the exclusion of the hepatic

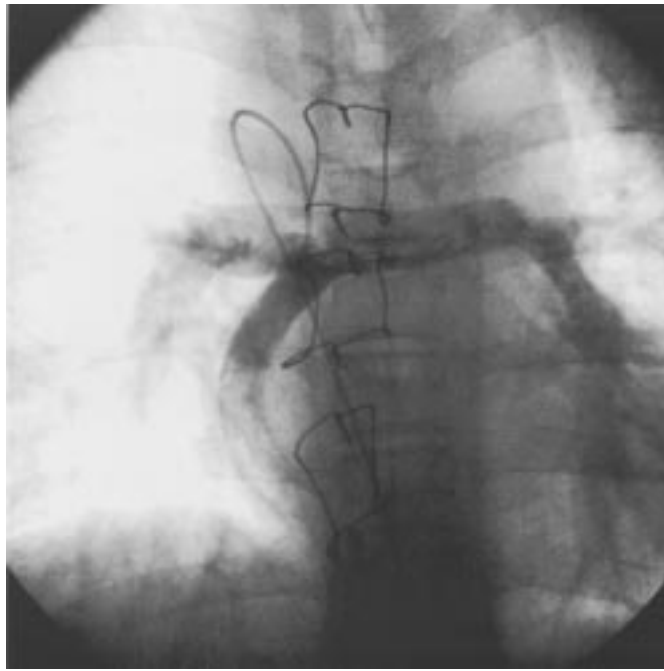


**Fig. 2.** Scheme of the operation.

A 16-mm expanded polytetrafluoroethylene (EPTFE) conduit was used for the redirection of the hepatic vein interposing between the hepatic venous orifices and pulmonary artery.

venous blood from the pulmonary circulation are two of the major risk factors.<sup>4)</sup> A conversion from TCPS to the Fontan circulation (redirection of hepatic venous drainage to the pulmonary circulation) could improve cyanosis efficiently, and Uemura et al. reported that the interval between the initial TCPS and the conversion was 30–80 months.<sup>2)</sup> This report is a case with the longest interval (130 months) from TCPS to Fontan, in which PAVF was diminished and cyanosis was improved. Although the precise mechanism by which hepatic venous flow prevents the development of PAVF is still not completely understood, the results of this report is consistent with the hepatic factor hypothesis, according to which the development of PAVF is facilitated when an unidentified factor produced or metabolized in the liver does not reach the pulmonary circulation before traversing another capillary bed.<sup>5)</sup>

An anastomosis of the hepatic veins to the azygos vein



**Fig. 3.** Angiography of the extracardiac conduit to the pulmonary arteries shows the diminished pulmonary arteriovenous fistula after the conversion.

has been described as an alternative approach for the treatment of PAVF after TCPS, and it allows for a balanced distribution of hepatic venous blood to the pulmonary arteries,<sup>6)</sup> but this approach may require circulatory arrest and may become a potential drawback if anastomosed directly. Nevertheless, redirecting hepatic venous drainage to perfuse both lungs in a balanced fashion appropriately is important at the conversion. PAVF could progress markedly in the unilateral lung even after the conversion in patients, in whom the hepatic veins had been exclusively diverted to the contralateral lung.

In conclusion, a conversion from TCPS to the Fontan circulation could diminish PAVF and improve cyanosis efficiently by appropriately redirecting hepatic venous drainage to perfuse both lungs in a balanced fashion with even an 11-year interval.

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