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*Brief Report*

# Transcatheter creation of an atriopulmonary communication in the Hemi-Fontan or Glenn circulation

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**Abstract** There have been several modifications of the Fontan procedure, which has been used for palliation of the functionally univentricular circulation since its conception in the early 1970s. The development of intrahepatic veno-venous fistulas, pulmonary arteriovenous fistulas, and communications from the hepatic vein to the pulmonary venous atrium, are some of the complications encountered by patients with cavo-pulmonary communications. We describe transcatheter creation of an atriopulmonary communication, with simultaneous implantation of a stent, as a method of reducing or preventing pulmonary arteriovenous fistulas and associated right-to-left shunting in this setting. The intervention has been undertaken in two patients following a Hemi-Fontan procedure, itself created subsequent to a Glenn anastomosis. The combination of these procedures creates communicating atriopulmonary and cavopulmonary circulations.

Keywords: Atriopulmonary communication; functionally univentricular heart; pulmonary arteriovenous malformations

Since the first report of the Fontan operation, the procedure has undergone numerous modifications as it has been used in the management of patients with functionally univentricular physiology.[1] Palliative procedures in such patients may include a total cavopulmonary communication, a unidirectional cavopulmonary connection combined with an adjustable atrial septal defect, a bidirectional Glenn anastomosis with fenestration of the baffle, or a modified Fontan operation with exclusion of a hepatic vein as a form of fenestration.[2–5] Regardless of the precise modification, these patients share certain potential complications. Leaks and obstructions related to the baffle are well recognized. In addition, the development of intrahepatic veno-venous fistulas, pulmonary arteriovenous fistulas, and communications between the hepatic veins and the pulmonary venous atrium, are further potential problems.[6–8] As treatment of these problems, placement of endovascular stents within the obstructed baffle, and transcatheter occlusion of communications from the hepatic vein to the pulmonary venous atrium, have previously been described.[8–10] We have now used a catheter to create a new atriopulmonary communication, simultaneously implanting a stent, as a means of reducing or avoiding the development of intrapulmonary arteriovenous fistulas and the associated right-to-left shunting.

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## Case reports

Our first patient, a girl, was diagnosed at birth with tricuspid atresia, discordant ventriculo-arterial connections, pulmonary stenosis, and a ventricular septal defect. Subsequent palliative procedures included a modified Blalock-Taussig shunt as a neonate, a classic right Glenn anastomosis at 6 years of age, and finally a baffle placed from the inferior caval vein to the left pulmonary artery to complete her modified Fontan circulation at 7 years of age. She was referred with a 2-year history of progressive cyanosis, with an oxygen saturation of 80% in room air, secondary to the development of multiple right-sided pulmonary arterio-venous fistulas. Cardiac catheterisation was

undertaken with the intent of occluding her fistulas with coils.

A prograde right heart catheterisation was performed through a 7 French sheath in the right femoral vein and the right internal jugular vein. Angiography demonstrated close proximity of the Glenn and Fontan circulations. The 7 French sheath in the groin was exchanged for a 7 French transseptal sheath, through which a perforation was made from the right atrium to the right pulmonary artery. Both the transseptal sheath and needle passed easily into the right pulmonary artery from the right atrium. The newly created channel was dilated twice with a 10 mm XXL balloon inflated to 17 atmospheres. The 7 French sheath in the groin was then exchanged for a 11 French transseptal sheath. A Palmaz P108 stent (Johnson and Johnson, Piscataway, New Jersey) was mounted on a 15 mm × 3 cm balloon and successfully implanted within the channel. Two P308 stents were additionally implanted in a stenotic segment of the left pulmonary artery, with significant improvement in flow of blood to the left lung. All sheaths and catheters were removed at the end of the case with no complications.

Repeat cardiac catheterisation was performed one year later. The stent in the atriopulmonary communication was redilated to a diameter of 17 mm. The stents in the left pulmonary artery were redilated to a diameter of 19 mm. There was a significant reduction in the pulmonary arterio-venous fistulas. She was discharged with systemic saturation of 95% in room air on follow-up 6 weeks later.

Our second patient was a boy diagnosed at birth with tricuspid atresia. The ventriculo-arterial connections were concordant, and there was some restriction of flow of blood to the lungs at the level of the ventricular septal defect. At 5 days of age, therefore, a modified Blalock-Taussig shunt was constructed surgically, followed by a right-sided Glenn shunt at the age of 5 years. The palliation was completed by a Hemi-Fontan procedure, anastomosing the right atrium to the left pulmonary artery, at 7 years of age. Four years later, he developed exercise intolerance and cardiac catheterisation was performed. A Palmaz 308 stent was implanted in the left pulmonary artery, abolishing a pressure gradient of 4 mmHg and significantly improving the flow of blood to the left lung. A leak across the baffle was closed using a device. At 15 years of age, repeat catheterisation was performed to re-evaluate his hemodynamics. The mean pressure in the anastomosis between the superior caval vein and the right pulmonary artery was 16 mmHg, and mean right atrial pressure was 24 mmHg. Angiography in anastomosis revealed a mid-segmental narrowing of the stent, which measured 11.7 mm at its narrowest dimension. A second Palmaz 308 stent was implanted, increasing the diameter of the stenotic area to 14 mm, and significantly improving flow to all segments of the left lung. As the right atrial pressure was significantly elevated, and with the risk of developing pulmonary arteriovenous malformations in the right lung, it was elected to connect the right atrium and the right pulmonary artery. A 7 French transseptal sheath and dilator were positioned in the superior and posterior aspect of the right atrium. Simultaneous angiography in the cavo-pulmonary atriopulmonary anastomoses demonstrated these structures to be within 3 mm of each other (Fig. 1a). A transseptal needle was advanced into the cavopulmonary anastomosis using a pigtail catheter as a guide (Fig. 1b). The transseptal sheath could not be passed into the right pulmonary artery. A platinum-plus wire was passed through the transseptal sheath and dilator, and the distal end was snared from the right internal jugular vein through the neck. The transseptal sheath could not be advanced because of dense scar tissue, despite advancing a small balloon of 6 mm diameter and dilating it with pressures up to 22 atmospheres. A second transseptal puncture was therefore made high and posterior to the first, and this made it possible to advance the sheath into the right pulmonary artery.

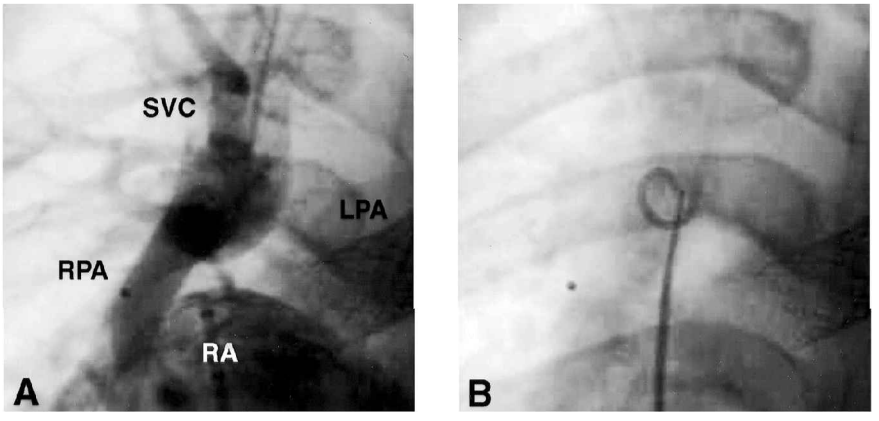


**Figure 1.**
*Simultaneous angiography in the Glenn and Hemi-Fontan circuits (A) demonstrates a minimal distance between the two circulations. Use of a transseptal needle (B) establishes access between the right atrium and the cavopulmonary anastomosis.*

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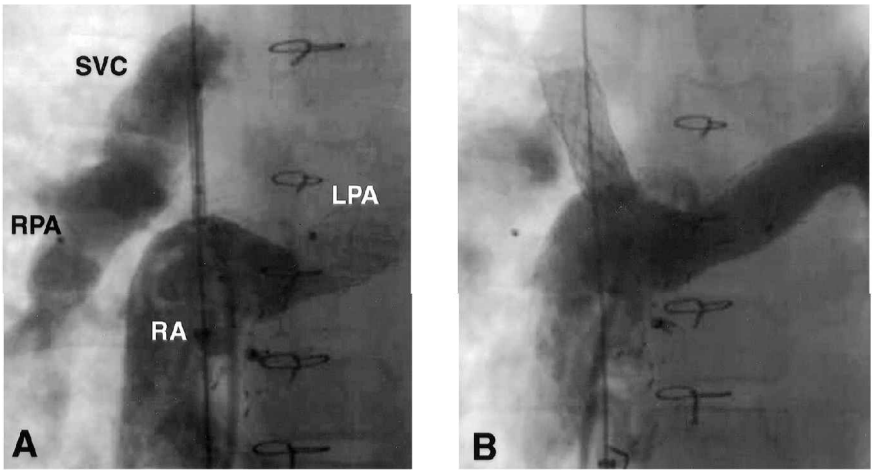


**Figure 2.**
*A stent (A) has been placed in the communication created between the right atrium and the cavo-pulmonary artery. Angiography (B) shows unrestricted flow to the right pulmonary artery from the Hemi-Fontan circuit.*

A 13.4 mm Park septostomy blade catheter was placed through the sheath, and two pull-backs were made from the cavopulmonary anastomosis to the right atrium. Subsequently, a Palmaz 308 stent was mounted on a 10 mm XXL balloon, and passed through a 10 French sheath to the site of the anastomosis (Fig. 2a). The balloon was thrice dilated, each time to 12 atmospheres. After the procedure, the mean pressure was 16 mmHg, with no gradient across the left pulmonary artery. Angiography demonstrated unobstructed flow in the left pulmonary artery, with a significant improvement in flow to that lung. There was unobstructed flow from the right atrium to both pulmonary arteries (Fig. 2b).

## Discussion

Several complications remain unique to the Fontan procedure, despite several modifications over the last three decades. Intrahepatic veno-venous fistulas, fistulas from the hepatic veins to the left atrium, pulmonary arteriovenous fistulas, as well as baffle leakage, may all give rise to right-to-left shunting and increased cyanosis following cavo-pulmonary anastomoses. Various interventional maneuvers have been described for amelioration of these problems.[9–10]

Patients who undergo a classic Glenn anastomosis combined with an atriopulmonary Fontan procedure may also develop pulmonary arteriovenous fistulas with consequent right to left shunting and desaturation.[8] Whether their development is purely secondary to elevated venous pressure, absence of hepatic derived growth factor, or a combination of both, remains speculative. The concept of creating a transcatheter communication between the Glenn and Fontan circulations, is not novel with completion of the Fontan circulation by catheterization previously reported by Konertz et al.[5] The transcatheter creation of an atriopulmonary communication using transseptal needle puncture, with simultaneous implantation of a stent, completes the cavopulmonary circuit and enables decompression of elevated venous pressure and supply of hepatic derived venous blood flow to the lung on that side, which may reduce or prevent development of pulmonary arteriovenous fistulas. The improvement in systemic oxygen saturation may significantly improve quality of life for these patients. Patients, however, must be carefully chosen, with minimal distances between their atriopulmonary and cavopulmonary anastomoses, and the procedure obviously should only be performed in centers with adequate technical experience. It should also be noted that, as in one of our patients, certain patients may have sufficient scar tissue to require use of a blade so as to create a suitable communication.

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