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Brief Report

Major intrahepatic veno-venous fistula after modified Fontan operation treated by transcatheter implantation of Amplatzer septal occluder

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Abstract Patients with complex congenital cardiac malformations who have been converted to the Fontan circulation with partial exclusion of the hepatic veins may develop progressive cyanosis because of formation of intrahepatic veno-venous malformations. We describe transcatheter closure of a major intrahepatic fistula in such a setting using an Amplatzer septal occluder delivered by the left jugular venous approach in a 5 year old boy.

Keywords: Fontan circulation, intrahepatic fistula, transcatheter closure, interventional catheterization.

Isomerism of the right atrial appendages, in the setting of a right-sided heart, is commonly associated with complex cyanotic congenital cardiac disease. Most of the patients are not suitable for biventricular repair. In such situations, construction of a total cavo-pulmonary anastomosis with a left-sided lateral tunnel offers good palliation. Additionally, either a fenestration, or partial exclusion of the hepatic veins is often performed, to reduce the venous pressure in the splanchnic vascular bed, minimizing protein loosing enteropathy at the expense of systemic arterial desaturation. The pressure gradient between the high pressure systemic venous tunnel, and the low pressure pulmonary venous atrium, which reflects the transpulmonary gradient, may lead to development of an intrahepatic veno-venous malformation. This, in turn, causes increasing right-to-left shunting, with progressive cyanosis. This problem has been treated surgically by ligation of the hepatic vein or its reinclusion in the Fontan circulation. So far, such large intrahepatic fistulas were not considered suitable for transcatheter closure. We report a patient in whom such uncontrolled shunting was treated by implantation of an Amplatzer septal occluder, with good early result.

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Case report

A 5-year-old boy was admitted with a right-sided heart, isomerism of the right atrial appendages, a midline liver, a common atrioventricular valve, totally anomalous pulmonary venous drainage to the left-sided systemic atrium, discordant ventriculo-arterial connections, and pulmonary stenosis. He was initially palliated by construction of a modified Blalock-Taussig shunt on the right side at the age of 17 months. Two years later, spontaneous closure of the shunt was diagnosed when arterial saturation dropped to 56% and an urgent bidirectional Glenn operation was performed. The postoperative course was uneventful, and he was discharged on the 8th postoperative day with a saturation of 83%. The second stage of the Fontan procedure, consisting of construction of an intraatrial left lateral tunnel with the use of a dacron patch, with a small baffle fenestration, was performed after 7 months. Diagnostic catheterization prior to this operation consisted of angiography, together with measurement of pressures in

the pulmonary arteries and the Glenn anastomosis, and had been performed through a left jugular venous approach. Thus, the arrangement of the hepatic veins was not elucidated. The postoperative course was complicated by pleural effusion, which was relieved by puncture. The boy was discharged 2 weeks after surgery with a saturation of 89 %. During the first year of follow-up, he progressively became desaturated to 65–75%, and was readmitted to our center for reevaluation. Angiographic visualization of the lateral tunnel revealed poor filling of the pulmonary arteries, rapid retrograde filling of the left sided inferior caval vein, and a small leak across the baffle. No stenosis or thrombosis of the systemic venous bed or pulmonary arteries was observed. There was no evidence of pulmonary arteriovenous malformations. Injection into the inferior caval vein below the diaphragm demonstrated two hepatic veins. The left one was of normal size, while the right one was significantly dilated. Selective injection to the right hepatic vein, which had been included in the Fontan circulation, showed a large vessel 20 mm in diameter, which divided into multiple sinusoidal collaterals to an equally large hepatic vein, unrecognized prior to surgery, which drained into the pulmonary venous atrium (Fig.1). A steal phenomenon through the liver was evident. The mean systemic venous pressure was 15 mmHg. The boy was scheduled for reoperation by construction of a new baffle to divert the right hepatic venous drainage to the left-sided lateral tunnel. Prior to the planned operation, saturations dropped still further, and he developed heart failure. Emergency transcatheter occlusion of the malformation was carried out, using a 28 mm Amplatzer septal occluder. The procedure was started through the left femoral venous approach. At first, the proximal part of the included right hepatic vein was occluded for 15 minutes using a large occlusion balloon (OBW 27mm-Meditech). This resulted in a rise of the saturation from 50 to 77%. The catheter was then advanced to the distal part of the "included" right hepatic vein, and a 0.035 Amplatz extrastiff exchange (Meditech) wire was advanced, permitting introduction of a 9 French delivery sheath (AGA Med.). Thereafter, the septal occluder was introduced through the sheath. Considerable resistance was encountered when trying to advance the device through the curved part of the sheath, resulting in kinking of the sheath. The procedure was repeated with the same equipment deployed from the left jugular vein. The device was then delivered by withdrawing the sheath. Control angiograms were performed before (Fig. 2) and immediately after release of the device. They revealed a minor residual leak through veno-venous malformations. At the end of the procedure, the saturation rose to 80%, and the central venous pressure remained unchanged. After one year of follow-up, the child is in a good general condition, with the saturations ranging from 80 to 85%. Echocardiography has confirmed complete occlusion of the fistula.

Discussion

In an attempt to reduce the incidence of effusions, hepatic congestion, and other adverse complica-

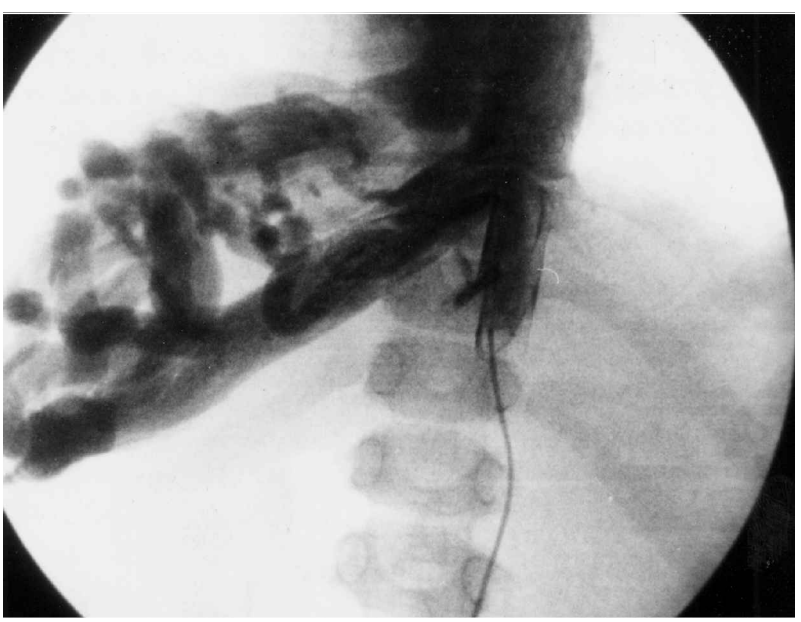

Figure 1.
Selective injection to the right hepatic vein shows a large vessel (20 mm in diameter) dividing into multiple sinusoidal collaterals communicating with an equally large hepatic vein draining into the pulmonary venous atrium.

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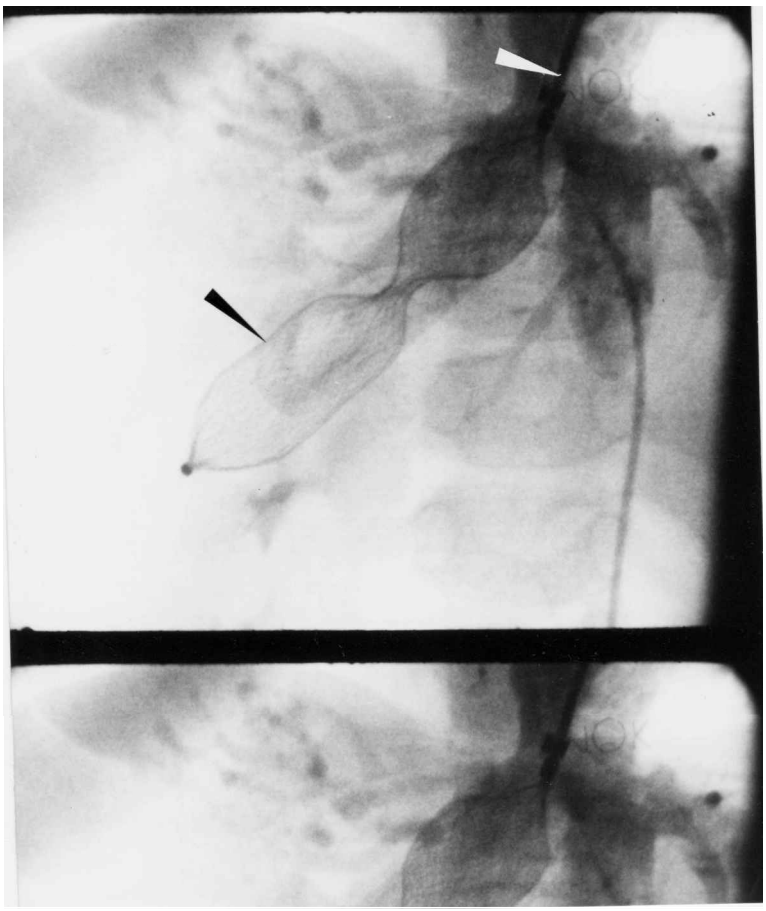

Figure 2.
Amplatzer septal occluder (28 mm) still connected to the delivery cable (white arrow) deployed as an "hour-glass" shaped plug (black arrow) in the huge and long proximal part of the fistula. An angiogram in the inferior caval vein just below the diaphragm shows a small residual leak.

tions of the Fontan operation, Jakobs and Norwood[1] introduced the concept of partial exclusion of the hepatic vein as an equivalent to fenestration and a "controlled" right-to-left shunt. This concept is especially attractive in more complicated cases such as those with right-sided hearts and right isomerism, in which either the Kawashima operation[2] or a total cavopulmonary connection using a left sided lateral tunnel[3] are alternative options. After this operation, systemic venous pressure exceeds that in the portal system and the "excluded" hepatic vein. The difference in pressure is dependent on the transpulmonary capillary gradient, which is normally in the range of 5 to 10 mmHg. Thus, there exists a risk for the development of fistulas and collateral vessels from the systemic to the portal and hepatic circuits. Precise diagnosis of such infradiaphragmatic veno-venous malformations requires angiography in the inferior caval vein. Such complications leading to progressive cyanosis have previously been reported.[4] This problem has usually been treated surgically by banding,[3] or re-inclusion of the "excluded" hepatic vein to the systemic venous tunnel.[5,6] Transcatheter closure is another option. The Rashkind umbrella,[7] detachable silicone balloons, and stainless steel coils[4] have been used, but only to close smaller fistulas. In our patient, the size of the abnormal vessel was too large for such techniques because of the risk of embolization, while the general condition of the patient made reoperation exceedingly risky. The implantation of a large Amplatzer atrial septal occluder[8] resulted in cessation of uncontrolled shunting, significant improvement of oxygen saturation, and stabilization of a critically ill patient. Despite this, a right intrahepatic shunt still remained. That is why implantation of a device into the "excluded" hepatic vein is a better option. Such treatment was reported by Tofeig at al,[9] who closed a 10 mm residual hepatic venous channel using a a 10 mm Amplatzer atrial septal occluder. Their technique of implantation of the Amplatzer device was different from ours. Closure of the "excluded" left hepatic vein was performed having crossed the entire length of the intra-hepatic fistula through a femoral venous approach. We decided to close the "included" right hepatic vein, because this vessel was long enough to accept the large plug, in contrast to the shorter "excluded" right hepatic vein. It might also have proved very difficult to cross tortuous

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intrahepatic branches with a long and stiff device. The left jugular venous approach in our case was technically easy, in contrast to the femoral approach. The latter proved unsuitable because of the sharp angle between the inferior caval vein and the "included" right hepatic vein. Another option would have been to close the excluded right hepatic vein through a left jugular venous approach and the fenestration in the baffle. This procedure would have been more hazardous in our case because the "excluded" hepatic vein was large but relatively short, giving a potential risk of embolization.

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