

Transcatheter occlusion of a hepatic vein-to-left atrium fistula: Should we close venovenous collateral vessels following Fontan operation?

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ABSTRACT

Right-to-left shunt due to abnormal systemic venous drainage to the left heart is among the causes of hypoxemia following Fontan operation. There are conflicting data regarding the closure of the venovenous collaterals (VVCs) post-Fontan, showing decreased survival in older patients. In a child with visceral heterotaxy, we describe a rare fistula draining a right-sided hepatic vein into hepatic venous plexus and a right-sided pulmonary venous atrium. The patient presented with severe hypoxemia post-Fontan and underwent fistula occlusion with AMPLATZER™ Vascular Plug II, successfully improving hemodynamic status with resolution of the hypoxemia. Younger patients with cyanosis due to VVCs may benefit from percutaneous occlusion post-Fontan.

Keywords: AMPLATZER™ Vascular Plug II, Fontan, left atrium-to-hepatic vein fistula, transcatheter closure of fistula

INTRODUCTION

Fontan procedure is a palliative surgery done as a final stage treatment in patients with single-ventricle physiology.^[1] This surgery directs blood from both venae cavae into the pulmonary arteries.^[2] This procedure can be performed with or without “fenestration,” which is a residual right-to-left shunt intentionally created to decompress the relatively high pulmonary artery pressure (PAP).^[3] Some centers advocate that fenestration might improve the surgical results in high-risk patients.

Patients with the Fontan circulation usually have an arterial oxygen saturation of around 90%, even in the absence of fenestration. The coronary sinus drains into the left atrium (LA), causing a modest arterial

desaturation of around 2%. Other causes of hypoxemia post-Fontan include a residual atrial septal defect or fenestration, intrapulmonary arteriovenous fistulae, and abnormal systemic venous drainage to the heart or the pulmonary veins. Closure of the venovenous collaterals (VVCs) may be necessary to decrease thromboembolism and hypoxemia.

However, Poterucha *et al.*^[4] reported that the embolization of the VVCs in older patients post-Fontan is associated with decreased survival, as these patients may benefit from the “natural” fenestration that VVCs provide. Few reported cases show favorable outcomes after occlusion of intrahepatic VVCs in patients presenting with profound cyanosis post-Fontan.^[5-10]

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We report a rare case of a fistula between a right hepatic vein (HV) and a right-sided pulmonary venous atrium in a child with profound hypoxemia post-Fontan.

CASE REPORT

We report a 7-year-old boy born to first-degree consanguineous parents, diagnosed with an abdominal situs ambiguous, unbalanced atrioventricular canal (single-ventricle physiology), severe pulmonary stenosis with confluent pulmonary arteries, and normally related great vessels. The patient underwent Blalock-Taussig shunt insertion during infancy, followed by bidirectional Glenn at 18 months of age.

Cardiac catheterizations pre-Fontan revealed atrial situs inversus with the pulmonary veins draining into a posterior right-sided atrium (anatomical LA) and an inferior vena cava (IVC) on the left of the spine draining into the left-sided atrium. The mean PAP was 13 mmHg. There was a small connection between the right HV and the LA.

We performed total cavopulmonary circulation without fenestration at the age of 3 years with no significant complications. Immediately postoperatively, the patient had an arterial oxygen saturation (SaO₂) of 97%. However, 1-month postoperative, the SaO₂ declined to 80% and reached the mid-70s a year later. Transthoracic echocardiography revealed normal systolic function and no atrioventricular valvular regurgitation.

Cardiac catheterization post-Fontan revealed a fistula between a right HV and a right-sided pulmonary venous atrium. The HV was connected to the IVC and a right-sided hepatic venous plexus [Figure 1 and Video 1]. The mean LA pressure was 9–10 mmHg. Occlusion of the fistula with a 12-mm TYSHAK® balloon dilatation catheter resulted in an increase in the mean PAP from 14 mmHg to 16 mmHg with a rise in the SaO₂ to the

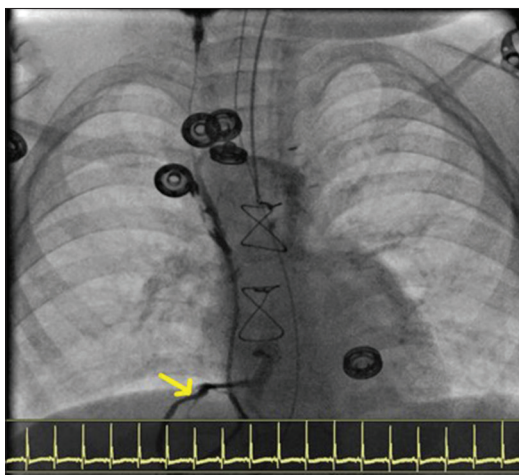


Figure 1: Angiograms in AP projection. Small collateral (yellow arrow) seen pre-Fontan

high 90s with appropriate systemic blood pressure. Accordingly, we decided to close the fistula.

A 5F Judkins catheter was advanced from the IVC to the LA and into the LA through the abnormal venous fistula. We exchanged the Judkins catheter for a 10F TorqVue LP long sheath. An AMPLATZER™ Vascular Plug II (AVP II) (14 mm × 10 mm) was delivered successfully and occluded the HV-LA fistula, with no obstruction across the Fontan tube [Figures 2,3 and Videos 2,3]. SaO₂ increased to 100% with excellent hemodynamics.

Three years following fistula's occlusion, the patient was doing well and reporting good exercise tolerance. His SaO₂ was 99%, and the previously noted fistula was completely occluded.

DISCUSSION

The development of decompressing VVCs is common following Fontan operations. This phenomenon is thought to be a biological reaction to the increased systemic venous pressure changes associated with this type of circulation. Often, these vessels are of supradiaphragmatic origin.^[5]

The severe hypoxemia encountered by our patient was due to an infradiaphragmatic right-to-left shunt draining the systemic venous circulation into the LA. Cardiac catheterization pre-Fontan revealed a small hepatic venous to LA connection that increased markedly in size postprocedure. Hepatic venous malformations are seen in 8% of heterotaxy patients undergoing Fontan procedures.^[9] The physiology of the Fontan operation might stimulate the enlargement of rudimentary embryologic connections from the systemic veins to the pulmonary veins or the pulmonary venous atrium, resulting in decompression of VVCs and causing desaturation.

Given the risk of progression of any VVC post-Fontan, all abnormal collaterals should be studied and closed pre-Fontan. Cyanotic patients post-Fontan should undergo diagnostic catheterization and balloon occlusion of the VVCs, unless contraindicated.

VVCs are found in 20%–45% of patients with Fontan circulation.^[3] High-risk Fontan patients, such as older patients with mildly decreased SaO₂ (88%–90%) and increased central venous pressures, like those enrolled in Poterucha *et al.* may benefit from these collaterals, and thus, VVC-closure in such patients may decrease survival rate.^[4] Poterucha *et al.* observed decreased 5-year survival in patients who had VVC embolization after Fontan.^[4] However, their study included older patients with a mean age of 26 years who presented with mild arterial oxygen desaturation at 88%–90%. In contrast, our patient had severe hypoxemia and low mean PAP of 14 mmHg. The patient improved immediately after the closure of the veno-atrial fistula, with an increase in his

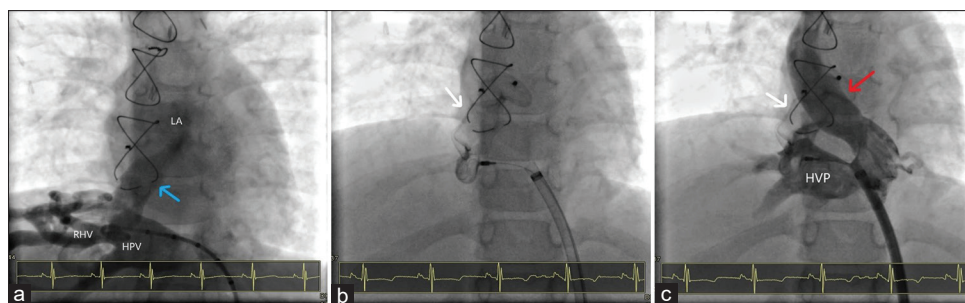


Figure 2: Angiograms in AP projection: (a) RVH, dividing into multiple sinusoidal channels in the liver (HVP), and draining into the right-sided pulmonary venous atrium (LA) through a fistula (blue arrow), (b) AMPLATZER™ Vascular Plug Type II (white arrow) placed successfully in the fistula, attached to the delivery wire, (c) AMPLATZER™ Vascular Plug Type II (white arrow) closing the fistula with no obstruction of the Fontan circulation (red arrow). RVH: Right hepatic vein, AP: Anteroposterior, HVP: Hepatic venous plexus, LA: Left atrium

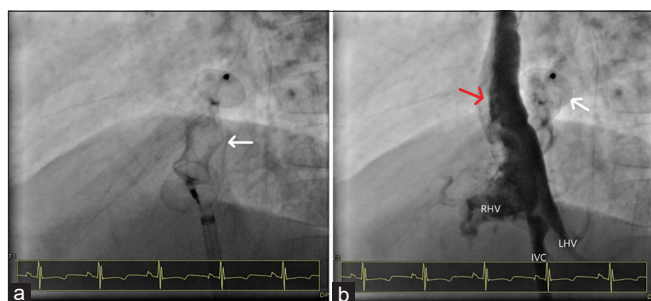


Figure 3: Angiograms in lateral projection: (a) AMPLATZER™ Vascular Plug Type II (white arrow) placed successfully in the fistula, attached to a delivery wire, (b) AMPLATZER™ Vascular Plug Type II (white arrow) closing the fistula with no obstruction of the Fontan circulation (red arrow)

SaO₂ from 75% to 100% with a slight increase in PAP from 14 mmHg to 16 mmHg and excellent hemodynamics. A 3-year follow-up revealed favorable results.

The relatively rare infradiaphragmatic VVCs presenting with severe cyanosis may be treated differently from those VVCs in older patients with mild cyanosis. The few reported cases that involved occlusion of these intrahepatic VVCs were associated with favorable outcomes.^[5-10]

Veno-venous collaterals closure is still debatable since high-risk patients (patients with elevated central venous pressure or heterotaxies) might benefit from the natural fenestrations created by the veno-venous collaterals.

CONCLUSION

Closure of a fistula causing right-to-left shunt in patients post-Fontan procedure should be considered cautiously.

Approach to VVCs in older patients after the Fontan operation may differ from that in younger children with severe hypoxemia and subdiaphragmatic VVCs. Patients with significant cyanosis due to intrahepatic to atrial connection may benefit from occlusion of these connections.

Authors' contributions

MA and ZB diagnosed and followed the patient in the clinic. FB and FS performed the procedure. MK wrote the manuscript with the supervision of FB. All authors approved the final manuscript.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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