# Novel use of covered stents to treat profound cyanosis in a hepatic vein exclusion Fontan

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## ABSTRACT

**Fontan completion in patients with complex cardiac anatomy, and specifically heterotaxy syndrome, can present unique physiologic considerations. For example, existing venous connections may be "unmasked" after a cavopulmonary anastomosis operation. We present the case of a child with heterotaxy, dextrocardia, single-ventricle physiology, and anomalous hepatic venous drainage that resulted in profound shunting and cyanosis. We addressed the problem utilizing a novel strategy with a "fenestrated" covered stent.**

**Keywords:** Congenital heart disease, covered stents, Fontan

## INTRODUCTION

The first reported Fontan procedure was performed on April 25, 1968, and involved direct anastomosis of the superior vena cava (SVC) to the right pulmonary artery (RPA) and right atrial appendage to the left PA (via the proximal RPA) in a 12-year-old child with tricuspid atresia.[1] The procedure has been modified significantly since being utilized on patients with increasingly complex cardiac and vascular anatomy.[2] Important aspects of patient and vascular pathophysiology have been identified as techniques and outcomes have improved. One known issue that can arise after Fontan completion relates to the presence of partially anomalous hepatic venous drainage. In this situation, one or more hepatic veins connect to both the inferior vena cava (IVC) (i.e., systemic venous circulation) and either atria (i.e., pulmonary venous circulation). This is a potential problem because the hepatic venous vasculature has the ability to remodel and form large interlobar connections in a short period. After Fontan completion, the hepatic veins in connection with the systemic venous circulation are subjected to significantly increased resistance while the hepatic veins connecting to the atria remain at lower resistance. This serves as an impetus for the development of these interlobar hepatic vein collaterals, creating a pathway with decompression of the systemic veins directly to the atrium with potentially massive right-to-left shunting.

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## CASE REPORT

We present the case of a 4-year-old patient with heterotaxy, dextrocardia, mitral atresia, pulmonary atresia, d-malposed great vessels, supracardiac totally anomalous pulmonary venous return (TAPVR), and late-diagnosed partially anomalous hepatic venous connections. The child underwent TAPVR repair and central shunt placement in the 1st week of life followed by a superior cavopulmonary anastomosis (i.e., bidirectional Glenn) at 5 months of age. The patient did well in the interstage and post-Glenn periods. Pre-Fontan catheterization at 3 years of age demonstrated normal PVRi of 1.8 WUi with an unobstructed pulmonary venous confluence, no significant valvar dysfunction, and normal ventricular function. The Glenn circuit had no focal angiographic obstructions or pressure gradients, though Glenn



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pressures were 16 mmHg and the PAs were mildly hypoplastic diffusely. Multiple hepatic veins were visualized to connect to the IVC.

Given the dextrocardia and borderline PA anatomy/pressure, an extra-cardiac Fontan (18 mm) was performed. The patient had a tenuous postoperative course secondary to conduit obstruction related to her dextrocardia and resultant tension on the SVC anastomosis. The child required extracorporeal membrane oxygenation with stent placement in the conduit on postoperative day 3. The patient had a steady though protracted recovery, but developed slowly progressive and profound hypoxemia and polycythemia ($SpO_2$ 65% and hemoglobin 22.4 g/dL) beginning approximately 1 month after Fontan. Subsequent catheterization diagnosed massive right-to-left shunting via hepato-hepatic venous collaterals–allowing systemic venous blood to bypass the PAs and flow directly into the atrium–with little-to-no effective antegrade pulmonary blood flow [Figures 1 and 2]. At this time, we retrospectively reviewed a prior computed tomography scan that was interpreted as having normal hepatic venous drainage. The scan demonstrated at least one hepatic vein, very small at the time, that anomalously connected directly to the atria.

We first attempted percutaneous occlusion of the anomalous hepatic using a vascular plug, a technique previously described.[3] The procedure was acutely successful with an increase in $SpO_2$ to ~85%. A second, smaller, and unrecognized anomalous hepatic vein was present, which slowly enlarged resulting in fairly rapid regression to the same degree of preocclusion cyanosis [Figure 2]. After multidisciplinary discussion, we opted to attempt a novel percutaneous therapeutic strategy involving modified covered stent placement.

At the next catheterization, we modified an 8-zig, 22-mm long covered Cheatham-Platinum® (CP) stent (B. Braun, Bethlehem, PA, USA) by cutting a small hole (1–2 mm) in the expanded polytetrafluoroethylene lining one-third from the stent end. During catheterization, an 0.018" wire was advanced into the largest hepatic vein connected to the IVC. The back end of this wire was placed through the hole we cut in the CP stent, and the stent was then mounted on an 18 mm Balloon-in-Balloon® catheter (PFM Medical, Carlsbad, CA, USA). The balloon-stent assembly was loaded onto a delivery wire in the SVC, and then stent was deployed. Sterling balloons® (Boston Scientific, Marlborough, MA, USA)–4 and 6 mm–were next used to dilate the side hole, creating a functional "fenestration" [Figures 3 and 4]. The procedure was well tolerated, with $SpO_2$ rising to 85% in room air immediately, and symptoms improving over coming months. Follow-up echocardiography demonstrated a 4 mmHg fenestration gradient and normal hepatic venous Doppler signals [Figure 4]. Hepatic elastography will be followed serially.

## DISCUSSION

The postoperative physiology that developed in our patient has been described in the past.[4] Dr. LeCompte of surgical fame theorized that excluding the hepatic veins from the Fontan circulation may limit postoperative effusions and reduce mortality.[4] However, he abandoned

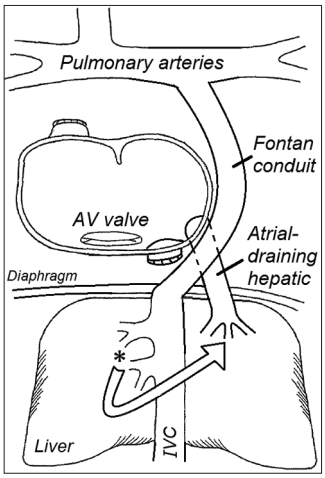


**Figure 1: Schematic representation of the relevant anatomy and physiology. Interlobar hepatic venous collaterals have developed between the hepatics connected to the inferior vena cava (*) and the atrial-draining hepatic vein. Given the lower resistance in the atrium, the result is massive right-to-left shunting of inferior vena cava blood directly into the atria via these hepatic venous collaterals (broad arrow)**

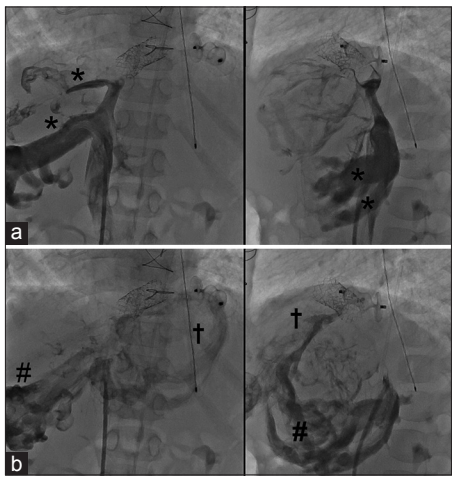


**Figure 2: Power injection in anteroposterior (left) and lateral (right) projections within the inferior vena cava limb of the Fontan pathway. Note the dextrocardia and mid-line liver. (a) The normally connecting hepatic veins (*) backfill early after contrast injection into the inferior vena cava. (b) A complex meshwork of interlobar hepatic collaterals (#) connects the inferior vena cava-hepatics to an anomalous hepatic that drains directly into the atrium (†). No contrast is seen within the inferior vena cava stent since all inferior vena cava flow is directed right-to-left into the atrium via the hepatic venous connections. Also seen is the vascular plug device in a previously occluded anomalous atrial-draining hepatic vein**

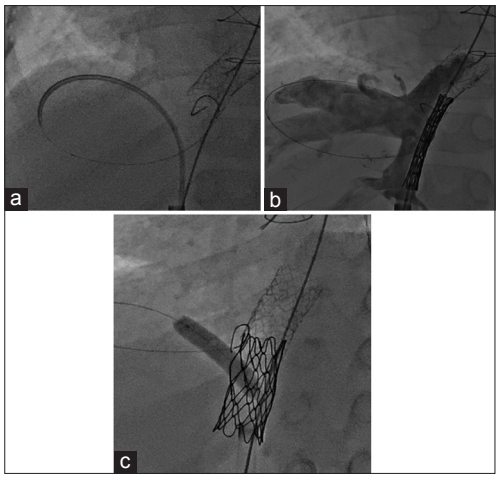


**Figure 3: Fenestrated covered stent delivery. (a) A 4 Fr catheter is positioned in the hepatic vein via the inferior vena cava and an exchange length 0.014" wire is advanced as distally as possible. (b) The 0.014" wire has been inserted through the covered stent (outside the body) which was then hand-mounted and advanced into position on a wire coursing from the inferior vena cava to superior vena cava. (c) The covered stent has been deployed and a 6 mm balloon was advanced over the 0.014" wire and into the hepatic vein, dilating the fenestration**

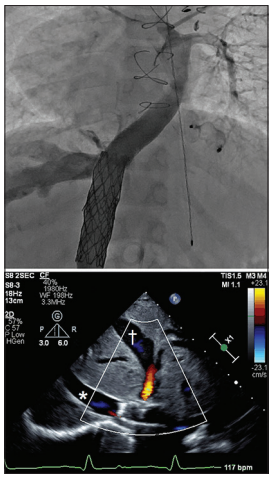

**Figure 4: Final inferior vena cava angiogram (top) and echocardiogram (bottom) after fenestrated covered stent placement. Top: Contrast now flows through the inferior vena cava stent and into the branch pulmonary arteries. The fenestration is patent with filling of the hepatic vein, but only the inferior vena cava-connecting hepatic vein has opacified. The interlobar connections and anomalous atrial-draining hepatic vein have not filled secondary to flow restriction through the controlled fenestration. Bottom: The inferior vena cava and stent (*) are widely patent with red fenestration flow visible into the hepatic vein (†)**

this technique because many patients developed progressive and profound cyanosis. Jacobs and Norwood also utilized this technique in at least 72 of 200 Fontan operations performed during the 1990s. The early patients suffered occlusion of the excluded hepatic veins, but after modifying their technique, those patients had notably less postoperative effusions.[5] However, the majority developed progressive and severe cyanosis.[6-9] A similar fate has been suffered by patients such as the one we present, who also had partially anomalous hepatic venous connections.[10,11]

Techniques have been described to deal with anomalous hepatic venous drainage resulting in cyanosis and shunting. In addition to the report above by Guerin, open surgical ligation techniques have been described using ultrasound guidance; interestingly, a similar report described a case in which a second additional anomalous hepatic was not identified like our patient.[12] The fundamental treatment must involve occlusion of all anomalous hepatics that drain into the heart or all hepatic veins arising from the systemic circulation as in our case.

In an attempt to curb protein-losing enteropathy (PLE), Brizard *et al.* have recently employed a similar rationale as LeCompte relating to post-Fontan effusions. That team has performed an operation re-directing the three major hepatic veins from the systemic venous circulation directly to the atria while placing a covered stent in the intra-hepatic IVC to exclude smaller hepatic veins.[13] Their rationale is that the higher hepatic vein pressures associated with a traditional Fontan are transmitted to the portal venous system, which overcomes regulatory mechanisms resulting in PLE. They have reported two patients in whom this technique was employed, and both had resolution of PLE with follow-up of 7 and 19 months. Kogon *et al.* have employed another unique operative technique in single-ventricle patients. That team performed Fontan completion by dividing the IVC below the liver and connecting the infrahepatic IVC to the pulmonary circulation using an extended extracardiac conduit. The native IVC to atrial connection was left intact, so that all hepatic veins drained directly into the atrium.[14] In essence, the team recapitulates the "Kawashima" circulation, a subset of patients with low incidence of PLE. More data are required to determine the safety and efficacy of these techniques, but the initial results are promising.

One specific concern in our patient is that the lobes of liver that were draining through normally connected hepatic veins now decompress through the interlobar hepatic collaterals [Figure 1]. These connections have presumably higher resistance, so those segments of liver remain under somewhat higher–albeit an unknown degree–pressure compared to the atrium. However, we know the resistance through this collateral network is lower than the Fontan circulation given the rapid and progressing cyanosis the child developed prior to this intervention. Nevertheless, we plan to monitor hepatic function and elastography at regular intervals to investigate this effect on the liver. In addition, the lack of "hepatic factor" flow to the lungs is a concern for the

development of microvascular pulmonary arteriovenous malformations. We will monitor for this condition as an additional operation to incorporate hepatic venous blood into the Fontan circuit may be indicated.

## CONCLUSION

Patients with heterotaxy and single-ventricle physiology can develop unique circulatory pathology that evolves after operations for single-ventricle palliation. Covered CP stents can be modified to serve unique needs including creation of fenestrations for decompression of the Fontan circulation and maintaining access to "jailed" vessels.

### 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.

## REFERENCES

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971;26:240-8.
2. Ohye RG, Schranz D, D'Udekem Y. Current therapy for hypoplastic left heart syndrome and related single ventricle lesions. Circulation 2016;134:1265-79.
3. Guérin P, Losay J, Baron O. Transcatheter occlusion of an intrahepatic venovenous fistula after modified Fontan circulation by implantation of an amplatzer atrial septal occluder. Catheter Cardiovasc Interv 2005;64:117-20.
4. Lecompte Y. Subtotal cavopulmonary connection. J Thorac Cardiovasc Surg 1992;104:1500.
5. Jacobs ML, Norwood WI Jr. Fontan operation: Influence of modifications on morbidity and mortality. Ann Thorac Surg 1994;58:945-51.
6. Schneider DJ, Banerjee A, Mendelsohn AM, Norwood WI Jr. Hepatic venous malformation after modified Fontan procedure with partial hepatic vein exclusion. Ann Thorac Surg 1997;63:1177-9.
7. Reed MK, Leonard SR, Zellers TM, Nikaidoh H. Major intrahepatic venovenous fistulas after a modified Fontan operation. Ann Thorac Surg 1996;61:713-5.
8. Erickson LC, Lopez A, Vlahakes GJ, King ME, Doody DP, Lang P, *et al.* Massive intrahepatic shunting seen as severe cyanosis after the Fontan procedure in heterotaxy syndrome. Am Heart J 1996;131:608-11.
9. Rao IM, Swanson JS, Hovaguimian H, McIrvin DM, King DH, Furnary AP, *et al.* Intrahepatic steal after Fontan operation with partial hepatic exclusion. J Thorac Cardiovasc Surg 1995;109:180-1.
10. Fernandez-Martorell P, Sklansky MS, Lucas VW, Kashani IA, Cocalis MW, Jamieson SW, *et al.* Accessory hepatic vein to pulmonary venous atrium as a cause of cyanosis after the Fontan operation. Am J Cardiol 1996;77:1386-7.
11. Yoshimura N, Yamaguchi M, Oshima Y, Tei T, Ogawa K. Intrahepatic venovenous shunting to an accessory hepatic vein after Fontan type operation. Ann Thorac Surg 1999;67:1494-6.
12. Ikeda A, Hiramatsu Y, Horigome H, Hori T, Noma M, Sakakibara Y, *et al.* A pitfall in ligation of intrahepatic shunting after Fontan type operation. Asian Cardiovasc Thorac Ann 2006;14:e6-8.
13. Brizard CP, Lane GK, Alex G, Cheung MM. Original surgical procedure for the treatment of protein-losing enteropathy in Fontan patients: Report of two midterm successes. Circulation 2016;134:625-7.
14. Kogon B, McConnell M, Book W. Fontan conversion with hepatic vein exclusion: A means for hepatic preservation in single ventricle heart disease. Cardiol Young 2016;26:582-5.