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CASE REPORT: CLINICAL CASE SERIES

Novel Staged Device Occlusion of Portosystemic Shunt With Atrial Flow Regulator in IVC Stent Platform

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ABSTRACT

We present the cases of 2 children diagnosed with extrahepatic portosystemic shunts, a very rare vascular anomaly, on investigation of cardiac symptomatology. Poorly developed portal venous systems necessitated staged shunt occlusion. This was achieved using atrial flow regulator devices positioned in an inferior vena cava stent platform performed in the cardiac catheterization laboratory. (J Am Coll Cardiol Case Rep 2024;29:102340) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

CASE 1

A 9-year-old boy presented to the emergency department with episodes of exertional syncope. He was previously well with no background medical history and normal growth. Physical examination revealed a precordial heave and loud P2 with no hepatosplenomegaly.

Transthoracic echocardiography demonstrated severe pulmonary arterial hypertension with a tricuspid regurgitant gradient of 92 mm Hg and an end pulmonary regurgitant gradient of 33 mm Hg. Investigations for causes of secondary pulmonary arterial hypertension included an abdominal ultrasound, which revealed an extrahepatic portosystemic shunt (PSS). No splenomegaly or focal nodular hyperplasia were identified. Portal venous phase computed tomography demonstrated a 16-mm PSS between the inferior vena cava (IVC) and the posteromedial aspect of the main portal vein (MPV) at the level of the porta hepatis (**Figure 1**). Laboratory investigations showed an elevated random serum ammonia of 173 μmol/L (<40 μmol/L), a platelet count

LEARNING OBJECTIVES

- To identify a PSS in the differential diagnosis of pulmonary hypertension in childhood.
- To evaluate the indications for staged occlusion of a PSS.
- To apply staged PSS occlusion using an atrial flow regulator in an IVC stent platform complex into practice.

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ABBREVIATIONS AND ACRONYMS

AFR = atrial flow regulator

IVC = inferior vena cava

MPV = main portal vein

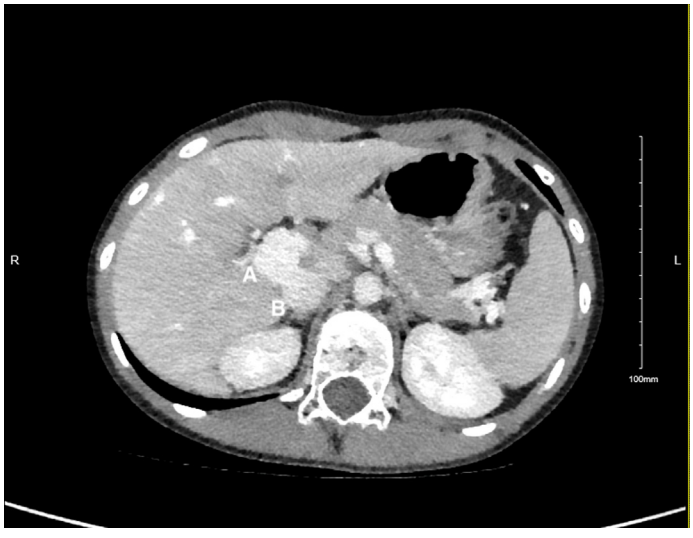
PSS = portosystemic shunt

of 120 × 10^9/L, an elevated prothrombin time of 15.8 seconds (normal range: 10.1-12.1 seconds), and normal liver function tests. Rifaximin and sildenafil were commenced. He was admitted for the insertion of an atrial flow regulator (AFR) to maintain cardiac output in pulmonary hypertensive crises and a staged occlusion of the PSS in the cardiac catheterization laboratory.

CASE 2

A 7-year-old girl was referred to cardiology services with palpitations. Her background medical history included intrauterine growth restriction, neonatal jaundice (resolved), failure to thrive, chronic spontaneous urticaria, and recurrent lower respiratory tract infections including empyema requiring surgical drainage. Clinical examination demonstrated frontal bossing, hypertelorism, and splenomegaly. Echocardiography revealed dilated and tortuous hepatic veins with left heart dilatation. Abdominal ultrasound identified a 14-mm tortuous extrahepatic PSS connecting the MPV to the IVC at the level of the porta hepatis, which was confirmed on computed tomography. Her liver was of normal size and echogenicity, and there was splenomegaly (11.1 cm, 3.1 SDs above mean). Her platelets were 135 × 10^9/L.

FIGURE 1 CT Scan of Portosystemic Shunt



Portal venous computed tomography demonstrating (A) a portosystemic shunt between the (B) intrahepatic inferior vena cava and the main portal vein.

PROCEDURE

The procedure was the same for both cases. After bilateral ultrasound-guided femoral vascular access, 100 IU/kg heparin was administered. Angiography demonstrated large shunts measuring up to 16 mm with a poorly developed intrahepatic portal venous system, when the PSS was balloon occluded (Figure 2).

An Amplatz Super Stiff Guidewire (Boston Scientific) was externalized between the right femoral and the right internal jugular veins. The veno-venous rail was used for the delivery of IVC stents across the MPV to IVC junction (uncovered AndraStent XXL [AndraMed GmbH] 30 and 43 mm mounted on 24- and 26-mm balloon-in-balloon platforms, respectively). Using the left femoral vein, the PSS was accessed, and stent struts were sequentially dilated to accommodate a 10-F Oscor long sheath (OSCOR Inc), the tip of which was flexed, facilitating delivery of a 4-mm AFR device (Occlutech 65AFR04M) in both cases (Figures 3 to 5).

In case 1, the baseline mean portal venous pressure was 4 mm Hg, increasing to 11 mm Hg with test occlusion. In case 2, the baseline mean portal venous pressures increased from 11 to 24 mm Hg. The final partial staged occlusion portal venous pressures were 8 mm Hg and 9 mm Hg, respectively. Patient 1 received low molecular weight heparin initially and subsequently aspirin. Patient 2 received aspirin as a single agent. No thrombotic events occurred.

In case 1, hemodynamic assessment revealed a pulmonary artery pressure of 82/33 mm Hg and a pulmonary vascular resistance index of 9 WU/m^2. A 6-mm Occlutech AFR was deployed across the atrial septum to mitigate against pulmonary hypertensive syncopal events. This patient returned 3 months later to the catheterization laboratory. Test occlusion of the fenestrated AFR-stent complex with a balloon wedge catheter demonstrated a rise in the mean portal venous pressure from 11 mm Hg to 13 mm Hg. Digital subtraction angiography demonstrated small but adequate intrahepatic portal venous branches. Therefore, the AFR was occluded using an Amplatzer

Duct Occluder II device (4-mm waist, 4.25-mm length) (Figure 6). Pulmonary resistance remained elevated with a pulmonary vascular resistance index of 7.79 WU/m^2, which decreased to 3.09 WU/m^2 on reversibility testing. No further exertional syncope has occurred. The tricuspid regurgitation gradient remains 88 mm Hg by echocardiography. The patient remains on 2 antipulmonary hypertensive agents. Patient 2 at the time of this writing is awaiting final staged closure of the PSS. The initial procedure was uncomplicated.

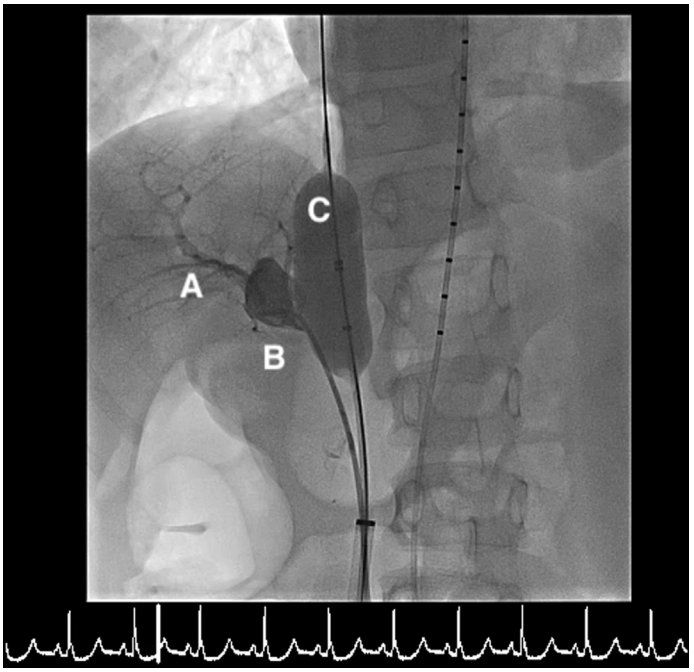
DISCUSSION

PSSs represent a group of rare vascular anomalies with an incidence of 1 in 50,000. PSSs may be intrahepatic or extrahepatic. Extrahepatic shunts are classified as either type 1, in which the portal vein does not supply any branches to the liver and may connect directly to the IVC or be absent entirely, or type 2 (as in our patients' cases), in which the MPV and the IVC are connected in a side-to-side fashion via a shunt. There may be significant hypoplasia of the intrahepatic portal venous system.[1]

Clinical manifestations include hepatic encephalopathy, liver nodules, hepatopulmonary syndrome, and portopulmonary hypertension.[2] Portopulmonary hypertension is present in 13% to 66% with varying degrees of severity, with the risk likely increasing with age and shunt size.[3,4] The pathophysiology is multifactorial. Pulmonary histology demonstrates small arterial microthrombi suggesting recurrent microemboli from the mesentery. Potent vasoconstrictive substances typically metabolized in the liver pass directly to the lungs.[3] Closure of the shunt may not reverse pulmonary hypertension but appears to prevent progression.[4] Portopulmonary hypertension carries a mortality rate of up to 50% in untreated shunts.[3]

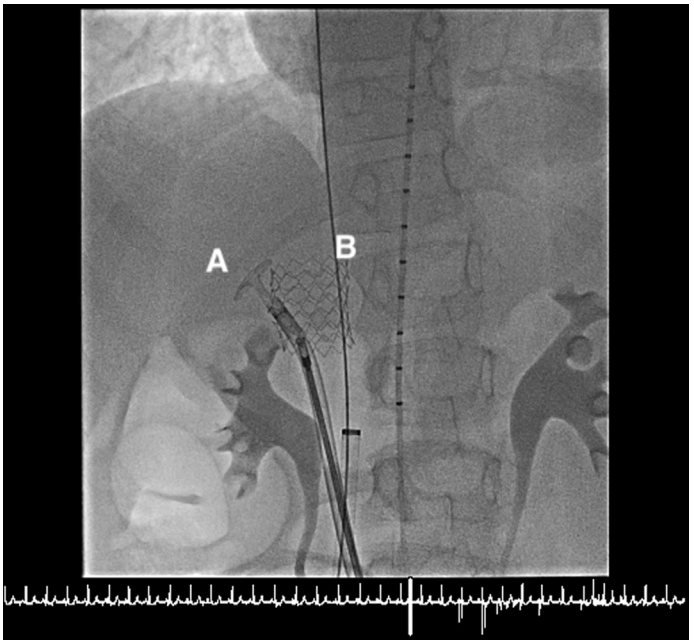
Closure is recommended for shunts persisting past the first year of life.[2] Before occlusion, angiography of the intrahepatic portal veins and balloon test occlusion of the shunt ideally with direct measurement of portal venous pressure are undertaken. Hepatic venous wedge pressure can alternatively be used as a surrogate to portal venous pressure. If the intrahepatic portal veins appear adequate and the portal venous pressure remains acceptable (<18 mm Hg), 1-stage shunt occlusion may be performed.[5-7] Although portal venous pressure in case 1 was below the threshold, it more than doubled with test balloon occlusion, and angiography of the hepatic portal veins demonstrated hypoplasia. Thus, staged occlusion to allow growth of the portal system was

FIGURE 2 Balloon Test Occlusion IVC



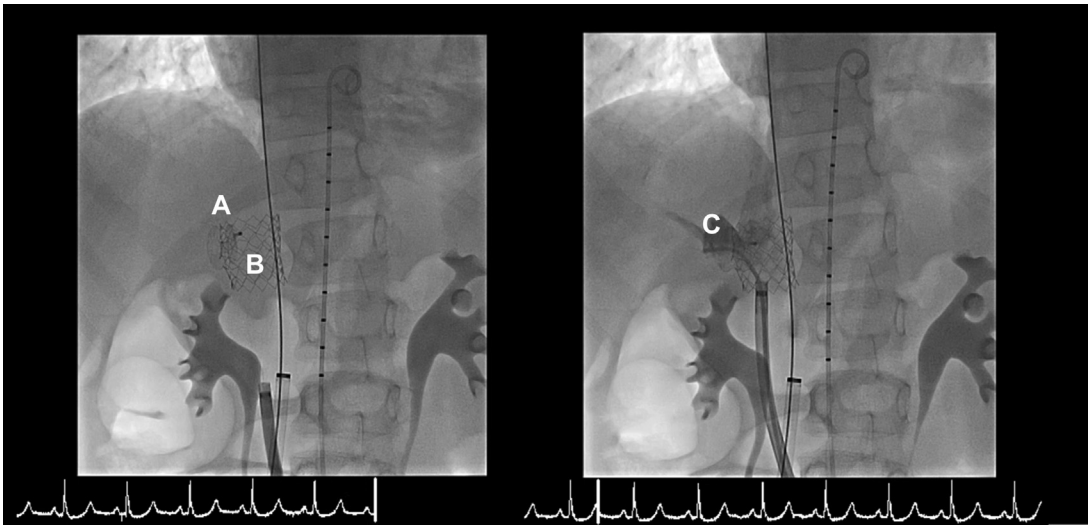
Contrast seen in a poorly developed (A) intrahepatic portal venous system, (B) portosystemic shunt, and (C) inferior vena cava (IVC) balloon occlusion at the level of the shunt.

FIGURE 3 AFR Deployed



(A) The first disc of the atrial flow regulator is deployed within the portosystemic shunt (B) as the sheath is withdrawn through the inferior vena cava stent.

FIGURE 4 Final Fluoroscopy and Angiography

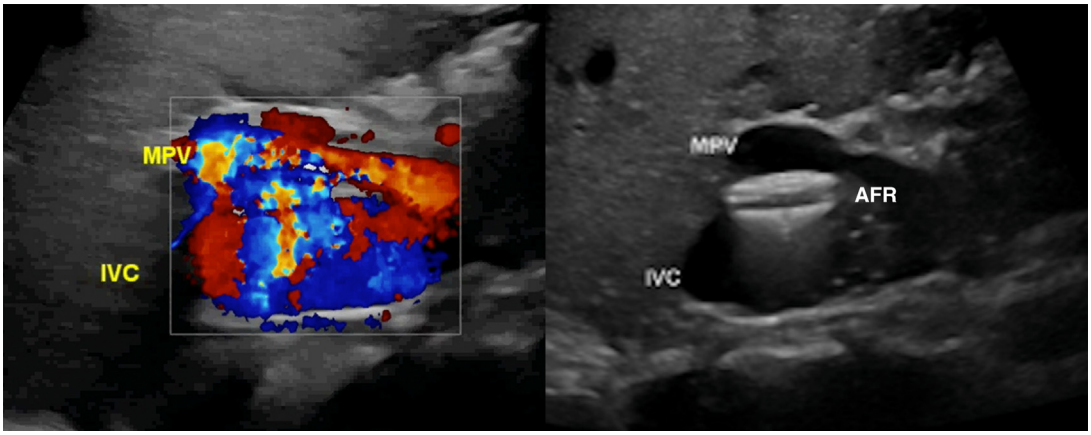


(A) The atrial flow regulator in (B) the inferior vena cava (IVC) stent platform. (C) Contrast seen to fill the portosystemic shunt with contrast seen in the IVC.

performed. Case 1 fulfilled both occlusion criteria on the follow-up assessment.

Various methods of staged endovascular intervention have been described including tailor-made flow-restricting stents, ductal or ventricular septal defect occlusion devices, and occlusion of the primary defect with iatrogenic creation of a transjugular intrahepatic PSS.[5-7] Surgical management is recommended if interventional techniques fail or if the shunt is unsuitable, with some centers advocating for surgical closure of all side-to-side shunts (such as described here) because of the challenges of closing large, short shunts endovascularly.[8] Splanchnic thrombosis complicates shunt closure in 8% to 27%,

FIGURE 5 Ultrasound of the IVC Stent AFR Complex



AFR = atrial flow regulator; IVC = inferior vena cava; MPV = main portal vein.

with most authors recommending prophylaxis frequently with heparin.[4,7,9] No consensus exists on choice or duration. Aspirin is routinely prescribed post-AFR insertion. Both patients were followed with serial ultrasonography.

We describe a novel approach in which an uncovered IVC stent provides a supportive platform, allowing an AFR device to be deployed across and partially occlude a type 2 extrahepatic PSS. This approach was performed collaboratively between cardiac interventionalists and interventional radiology.

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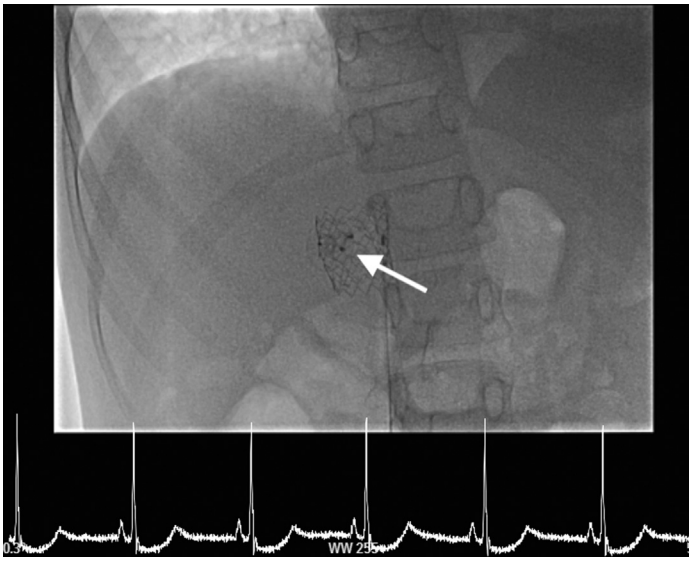
FIGURE 6 Post-Final Occlusion of AFR Fenestration



Amplatzer Duct Occluder II (arrow), occluding the atrial flow regulator lumen.

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