The surgical connection of the vena cava to the pulmonary arteries creates a circulation in series and defines the Fontan operation for univentricular hearts. The central venous pressure is elevated (≈15 mmHg) and drives the blood into the pulmonary vessels.

This elevated venous pressure overpowers the postsinusoidal regulatory mechanism normally responsible for maintaining portal pressure at ≈10 mmHg. The elevated venous pressure is transmitted to the sinusoids, portal vein, and gut capillaries. Protein-losing enteropathy (PLE) is a complication of this hemodynamic state, where proteins leach into the gut from submucosal lymphangiectasia. PLE occurs in ≈5% of patients undergoing the Fontan procedure and is often fatal within 5 years of onset.

We performed a novel surgical procedure to restore a gradient similar to the physiological one between the portal vein and the inferior vena cava (IVC) in 2 Fontan patients with PLE. Hospital Human Ethics Committee’s approval and informed consent were obtained.

Under cardiopulmonary bypass, the 3 major hepatic veins (right, middle, and left) were separated and connected to the common atrium. An extracardiac conduit without fenestration, placed in the hepatic portion of the IVC, completed the Fontan circuit. The other hepatic veins, including the vein draining the caudate lobe and multiple venules that normally drain into the IVC, were occluded by a custom-made covered stent placed in the retrohepatic IVC deployed percutaneously immediately after surgery (Figure).

The venous drainage of the territories occluded with the covered stent rerouted itself toward the 3 major hepatic veins, with no resulting hepatic dysfunction. The portal flow (now routed directly to the common atrium) thus became a right-to-left shunt equivalent to ≈20% of systemic venous flow.

Our first patient received a fenestrated extracardiac Fontan at age 4. PLE was diagnosed 18 months postoperatively. The Fontan pressure was 15 mmHg with a patent fenestration. Endoscopy and small-bowel biopsy revealed duodenal lymphangiectasis. Despite treatment with high-dose budesonide, diuretics, and albumin transfusions, the albumin level was constantly <1.4 g/dL, and α1-antitrypsin clearance (a measure of net gastrointestinal protein loss) was 450 mL/d before surgery.

Postoperatively, his liver transaminases did not increase. Warfarin was instituted (target international normalized ratio, 2–3). There was no lessening of PLE initially; parenteral nutrition and fractionated heparin were added. Seven weeks postoperatively, signs of PLE subsided. The α1-antitrypsin clearance was normal (<16 mL/24 h) and serum albumin was 4.0 g/dL. The patient resumed normal activities and diet with a resting oxygen saturation >82%. Catheterization showed no communication between the Fontan circuit and the liver. The Fontan pressure was 12 mmHg and hepatic venous pressure was 8 mmHg. Endoscopy and small-bowel biopsy showed resolution of the lymphangiectasis.
Our second patient was a teenager with hypoplastic left heart syndrome who had undergone a fenestrated Fontan at age 4. Her PLE was diagnosed at age 7 and worsened over the next 3 years despite medical therapy. She had good hemodynamics with a patent fenestration on cardiac catheterization. Her $\alpha_1$-antitrypsin clearance before surgery was 720 mL/d, and liver ultrasound suggested cirrhosis.

The same surgical technique was used. Her liver biopsy did not confirm cirrhosis. Her course was smoother than our first patient’s. The $\alpha_1$-antitrypsin clearance declined continuously but did not normalize over the first 7 months postoperatively, with a value of 181 mL/d. Her albumin level took 5 months to reach 3.2 g/dL. Her resting oxygen saturation is 81%.

Several authors have rerouted either the 3 superior hepatic veins\(^2\) or only the left hepatic vein\(^3\) to try improving Fontan physiology. Within days, a shunt developed inside the liver from the high venous pressure compartment (the parenchyma draining at Fontan venous pressure) to the low-pressure compartment (the hepatic veins connected directly to the common atrium). Unsustainable cyanosis ensued, leading to reintervention and takedown of the modification or death.

We previously demonstrated in animals that we could avoid intrahepatic shunting by isolating the whole retrohepatic IVC and connecting it to the low-pressure pulmonary venous atrium.\(^4\) This technique was recently used by another team in adult Fontan conversion.\(^5\)

Our 2 patients with PLE are the first to be successfully treated surgically without Fontan takedown or transplantation. Other alternatives, either creation or enlargement of a fenestration or medical treatment, typically provide only temporary and partial relief of symptoms and may cause extreme oxygen desaturation. The novel approach we describe is straightforward to perform and restores physiological portal vein hemodynamics and pressure while achieving a single-pressure compartment within the liver. No intrahepatic shunting appeared during the 19 and 7 months follow-up with stable oxygen saturations.

In summary, we report midterm results from a novel combined surgical and percutaneous approach to the treatment of PLE in which we isolate the entire hepatic venous flow from systemic blood flow in the Fontan circuit. The preliminary outcomes are favorable and should be reproducible in PLE patients providing the common atrial pressure is low.
Surgical Treatment of PLE in Fontan

DISCLOSURES
Dr Brizard is a member of the advisory board to Admedus, Australia.

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FOOTNOTES
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REFERENCES
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