Received: 1 January 2018 | Revised: 6 March 2018 | Accepted: 25 March 2018

DOI: 10.1002/ccd.27634

CASE REPORT

WILEY

A custom-made percutaneous flow-restrictor to manage a symptomatic congenital porto-systemic shunt in an infant

Mieke Roggen, MD | Bjorn Cools, MD | Geert Maleux, MD, PhD | Marc Gewillig, MD PhD FSCAI

Pediatric Cardiology, University Hospital Leuven, Leuven 3000, Belgium

Correspondence
Marc Gewillig, MD, FSCAI, Pediatric and Congenital Cardiology, UZ Leuven, Herestraat 49, 3000 Leuven, Belgium.
Email: marc.gewillig@uzleuven.be

Funding information
Eddy Merckx Research Grant, Belgian Foundation for Pediatric Cardiac Research FNRCP

Abstract

Portosystemic shunts allow splanchnic blood to bypass the liver unfiltered, and may cause serious pulmonary and cerebral dysfunction; closure is therefore recommended. In patients where the portal system is hypoplastic, closure by a staged approach with a flow reducer may be necessary. We report a new, reliable, short, and adjustable device that can be delivered through a small 8-Fr sheath.

KEYWORDS
Abernethy, flow regulator, hepatopulmonary syndrome, portal vein hypoplasia, venous duct dysfunction

1 | INTRODUCTION

Congenital porto-systemic shunts (PSSs) are rare and often difficult to diagnose. After birth a PSS allows the splanchnic blood to bypass the liver which then arrives unfiltered in the systemic circulation. This may result in a wide variety of symptoms including encephalopathy, mental retardation, cyanosis, clubbing, pulmonary hypertension, focal liver abnormalities, and cholestasis [1,2].

Treatment of this hepatic dysfunction consists of forcing the splanchnic blood through the portal venous system by occluding the PSS. However, when the portal system is too hypoplastic, a staged approach may be necessary. Such staged approach in small children requires a reliable flow reducer with a small landing-zone delivered through a small sheath. We report a custom-made device that was used successfully in an infant.

2 | CASE REPORT

A 14-month-old girl presented with progressive cyanosis. She had a prenatal diagnosis of abdominal situs inversus with azygos-continuation. She underwent surgery in the neonatal period for a duodenal web; saturations then were normal at all times. At the age of 14 months (9.3 kg) she presented with arterial desaturation of 81%. There was no respiratory distress and her chest X-ray was normal. Contrast echocardiography with agitated saline showed no early but some late bubbles in the left atrium, excluding a significant intracardiac right-to-left shunt. On CT-scan, no major arterio-venous malformations were seen in the lungs, but a large PSS between the main portal vein and right renal vein was visualized. Catheterization confirmed the above findings. The oxygen saturation in the pulmonary veins was low and injection in the pulmonary artery resulted in very fast opacification of the left atrium, which confirmed the diagnosis of diffuse micro arterio-venous malformations. Abdominal angiography confirmed a large shunt between the portal vein and the right renal vein. A portal vein to the main liver lobe (situated on the left) was visualized but it was small and hypoplastic (Supporting Information Clips 1 and 2). Pressure in the inferior caval vein and portal vein was 13 mm Hg. During temporary balloon occlusion of the PSS, the pressure in the portal vein increased to 26 mm Hg which we considered too high for single step closure. We therefore chose not to fully occlude the shunt but rather to implant a flow restrictor to progressively force the hypoplastic portal veins to develop. Informed, written consent was obtained from the parents.

Angiography and balloon interrogation had revealed a short 20 mm landing zone with a stretched diameter of 9 mm. After bench testing, the following custom-made flow restrictor was assembled with commercially available materials (all off-label use): a 6 mm Occlutech Muscular VSD Occluder (Occlutech, Jena, Germany; folded profile 6 Fr, center 6 mm, disks 13 mm, delivery cable 5.5 Fr) was perforated with a needle, wire, and a short 5-Fr sheath to allow a 5/16 Bentley covered stent (Bentley, Hechingen, Germany; covering ePTFE [expanded

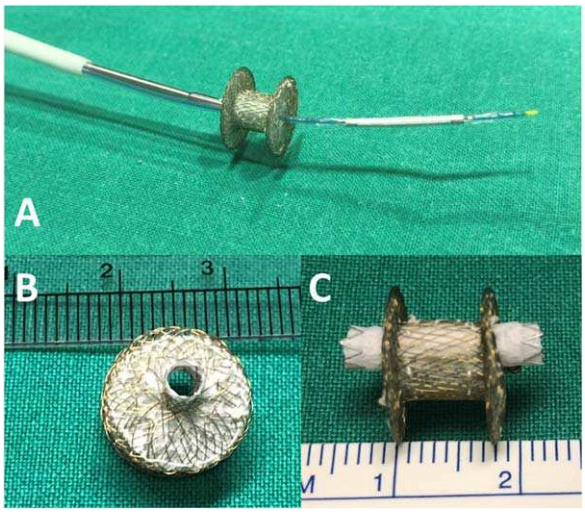

FIGURE 1 A, Flow restrictor assembled: muscular VSD Occluder, attached to cable, perforated with 5/16 Bentley covered stent on 0,014″ Iron Man, ready to be folded into 8-Fr sheath. B,C, Flow reducer: 5/16 mm Bentley Covered Stent deployed inside a 6 mm Occlutech muscular VSD Occluder. [Color figure can be viewed at wileyonlinelibrary.com]

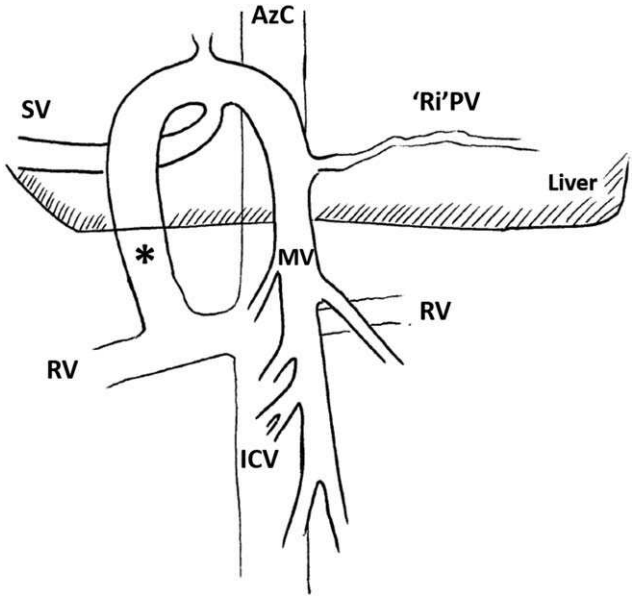

FIGURE 3 Schematic drawing. SV splenic vein, HV hepatic vein, "Ri" PV right portal vein (situated on the left), RV renal vein, IVC inferior caval vein, AzC azygos continuation, *PSS

polytetrafluoroethylene]; crimped stent profile 4 Fr, shaft just proximal of stent 3.2 Fr) on an 0.014″ coronary wire to be brought distal of the VSD device (Figure 1; Supporting Information Movie). This complex was pulled into an 8 Fr mounting sheath. Further bench testing showed that the Bentley stent could easily be pulled back into the pre-perforated device without encroaching of the stent on the rounded 0.067mm Nitinol wires, provided the stent was pulled into the device on a rather straight course.

Procedure: An 8 Fr delivery sheath was positioned in the shunt; the assembly was pushed from the 8 Fr mounting sheath into the 8 Fr delivery sheath. First, the VSD Occluder was deployed in the predefined landing zone; the small covered stent was then pulled back inside the device and deployed with an indeflator up to 10 atmospheres (Figures 2 and 3; Supporting Information Clips 5–7).

Control angiography showed good position of the device with a reduction of flow running through the shunt. The pressure in the portal system increased from 13 to 18 mm Hg, which was considered acceptable. She was started on acetylsalicylic acid 2 mg/kg/day and clopidogrel 0.2 mg/kg/day to prevent shunt occlusion.

At follow-up two months later, her saturations had risen to 91% in ambient air. Angiography showed a patent covered stent, without signs of thrombus or peel/intimal proliferation. Pressure in the portal system was 12 mm Hg. The dominant "right" portal system (situated in the left hypochondrial region in situs inversus) had well developed but no hepatopetal flow could be demonstrated in the smaller lobe (Supporting Information Clips 3 and 4). Portal pressure rose to 17 mm Hg after balloon test occlusion. The shunt was subsequently closed with a 7 mm Amplatzer Vascular Occluder (St. Jude Medical, Zaventem, Belgium).

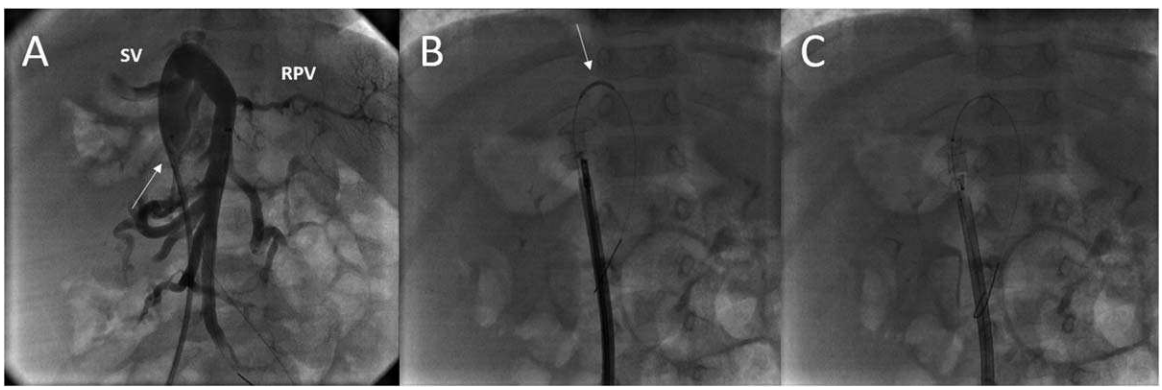

FIGURE 2 A, Venous portogram during balloon occlusion. Double access: 4-Fr sheath via right femoral vein into portal system for angiogram. From left femoral vein a 10 mm Tyshak balloon over 0.14 wire. Hypoplastic "right" portal vein (RPV), situated on the left site in patient with situs inversus abdominalis. Balloon interrogation shows 20 mm landing zone between splenic vein (SV) and right renal vein (white arrow). B, 6 mm mVSD Occluder deployed in situ but still attached to cable; covered stent (arrow) ready to be pulled back. C, Stent deployed inside device, which is released from cable; still 0.014″ wire through stent in portal vein

Follow-up demonstrated a further increase in saturation to 95% 2 months later, with normal portal flow in the dominant liver lobe.

3 | DISCUSSION

The normal intrahepatic portal system develops during fetal life as both the umbilical blood and the splanchnic blood are mainly drained through the liver, leaving the venous duct to drain only 30% directly to the heart [3]. If an abnormal connection between the portal and systemic venous system persists or develops (PSS), the "push" for the portal system to develop will be diminished, resulting in intrahepatic portal hypoplasia. Such PSS may not close after birth and can lead to abnormal portal drainage and insufficient hepatic filtering. This may result in significant pulmonary and cerebral dysfunction [1,2]. Treatment of the subsequent hepatic bypass and dysfunction consists of forcing the splanchnic blood through the portal venous system by occluding the PSS. For mild to moderate portal hypoplasia, a one-step occlusion of such shunt is usually well tolerated. However, when the portal system is too hypoplastic, a staged approach may be necessary to avoid portal hypertension, intestinal congestion and hypoperfusion. Flow reduction of the PSS can be obtained by surgery (banding) or by catheter techniques. Percutaneous treatment requires a reliable flow reducer with a small landing-zone preferably delivered through a small sheath; such flow restrictor is currently commercially not available. When these techniques fail to activate the portal system with persisting inadequate hepatic filtering, liver transplant may be the last option [4].

To determine whether a one- or two-step procedure is indicated, temporary balloon occlusion of the shunt with portal angiography and pressure measurement can be performed. Severe hypoplasia will be reflected by an acute rise of the portal pressure; however, no precise guidelines with proven cut-off values are reported, but suggestions ranging from 18 to 32 mm Hg can be found [4,5].

Many custom-made flow restrictors have been reported in various situations [4,6–11]. Most are bulky, complex, long, not easily adjustable, and require large or multiple sheaths for delivery. We developed a novel flow reducer which is easy to assemble, short, reliable, adjustable in size, and can be delivered through a single small sheath (8 Fr).

We chose a stent perforated occlusion device. Muscular VSD Occluders with two disks perpendicular to the flow have a high retention and occlusion power on a short distance (8 mm) and can fit in small and larger vessels. There is experience with stent perforating devices to allow controlled residual flow [10]. One method may consist of first deploying the occluder over a wire, then reperforate the device over that wire with a long thin perforating sheath, which then allows to position and deploy the stent within the device [10]. This technique requires a larger delivery sheath: bench testing showed that with the material as in our case at least a 10 Fr sheath would have been required as at one point the sheath must accept both the 5.5 Fr delivery cable and a 4 Fr perforating sheath (outer profile 5.5 Fr). Leaving the covered stent upstream during folding and deployment, as reported in this case, reduces the size of the delivery sheath from 10 to 8 Fr. Bench testing had shown that the folded VSD Occluder (profile 6 Fr) and the shaft (profile 3.2 Fr) of the premounted stent can easily fit in an 8 Fr sheath. Theoretically, the stent might slide off the balloon when pulling it into the device; bench testing and our clinical case demonstrated this to be unlikely. The new generation of premounted small covered stents has a favorable profile, allowing the stent to be pulled over a wire from distally into the occluder, provided this path is predilated and rather straight. If during this maneuver the stent would hook on the nitinol wire of the occluder device and would slide off the balloon (what did not happen during multiple tests), we expect that the whole complex can be refolded into the delivery sheath. Bench testing showed that the flow opening can easily be adjusted: the stent can be trimmed by adding other (smaller) stents [11], and can be increased by simple balloon dilation of the covered stent up to 7 mm.

After two months this device remained patent in this low gradient-low flow setting without thrombus or intimal proliferation under antiplatelet therapy. It was easily closed with a small vascular occluder.

4 | CONCLUSION

We report on a custom-made vascular flow restrictor that can be used in small patients with a PSS. It is easily assembled from commercially available devices, can be delivered through an 8-Fr sheath, allows a short landing zone, is adjustable and gives reliable flow for at least several weeks.

CONFLICT OF INTEREST

none declared.

ORCID

Mieke Roggen MD http://orcid.org/0000-0001-9659-8710
Marc Gewillig MD PhD FSCAI http://orcid.org/0000-0002-4595-5922

REFERENCES

[1] Bernard O, Franchi-Abella S, Branchereau S, Pariente D, Gauthier F, Jacquemin E. Congenital portosystemic shunts in children: Recognition, evaluation, and management. Semin Liver Dis 2013;32:273–287.

[2] Witters P, Maleux G, George C, Delcroix M, Hoffman I, Gewillig M, Verslype C, Monbaliu D, Aerts R, Pirenne J, Van Steenbergen W, Nevens F, Fevery J, Cassiman D. Congenital veno-venous malformations of the liver: Widely variable clinical presentations. J Gastroenterol Hepatol 2008;23:e390–e394.

[3] Kiserud T, Rasmussen S, Skulstad S. Blood flow and the degree of shunting through the ductus venosus in the human fetus. Am J Obstet Gynecol 2000;182:147–153.

[4] Bruckheimer E, Dagan T, Atar E, Schwartz M, Kachko L, Superina R, Amir G, Shapiro R, Birk E. Staged transcatheter treatment of portal hypoplasia and congenital portosystemic shunts in children. Cardiovasc Intervent Radiol 2013;36:1580–1585.

[5] Franchi-Abella S, Branchereau S, Lambert V, Fabre M, Steimberg C, Losay J, Riou J-Y, Pariente D, Gauthier F, Jacquemin E, Bernard O. Complications of congenital portosystemic shunts in children:

Therapeutic options and outcomes. J Pediatr Gastroenterol Nutr 2010;51:322–330.

[6] Maleux G, Verslype C, Heye S, Wilms G, Marchal G, Nevens F. Endovascular shunt reduction in the management of transjugular portosystemic shunt-induced hepatic encephalopathy: Preliminary experience with reduction stents and stent-grafts. Am J Roentgenol 2007;188:659–664.

[7] Maleux G, Heye S, Verslype C, Nevens F. Management of transjugular intrahepatic portosystemic shunt-induced refractory hepatic encephalopathy with the parallel technique: Results of a clinical follow-up study. J Vasc Interv Radiol 2007;18:986–992.

[8] Blue B, Lo G, Kim E, Patel R, Nowakowski F, Lookstein R, Fischman A. Transjugular intrahepatic portosystemic shunt flow reduction with adjustable polytetrafluoroethylene-covered balloon-expandable stents using the "sheath control" technique. Cardiovasc Intervent Radiol 2016;39:935–939.

[9] Abdelkarim A, Levi DS, Tran B, Ghobrial J, Aboulhosn J. Fenestrated transcatheter ASD closure in adults with diastolic dysfunction and/or pulmonary hypertension: Case series and review of the literature. Congenit Heart Dis 2016;11:663–671.

[10] Gorenflo M, Gewillig M. A flow restrictor implanted percutaneously across a loose pulmonary artery band. Catheter Cardiovasc Interv 2011;77:696–699.

[11] Demir F, Cools B, Gewillig M. Flow reduction of a neonatal stented arterial duct by covered coronary stent. Catheter Cardiovasc Inverv 2015;86:E73–E75.

SUPPORTING INFORMATION

Additional Supporting Information may be found online in the supporting information tab for this article.

How to cite this article: Roggen M, Cools B, Maleux G, Gewillig M. A custom-made percutaneous flow-restrictor to manage a symptomatic congenital porto-systemic shunt in an infant. *Catheter Cardiovasc Interv*. 2018;00:1–4. https://doi.org/10.1002/ccd.27634