

Percutaneous treatment of a huge patent ductus venosus and severe portal vein hypoplasia using a Figulla Flex II atrial septal defect occluder in a 2-year-old infant: a case report

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Background

Intra- or extrahepatic porto-caval shunts (PCSs) can account for multiorgan dysfunction with pulmonary arterial hypertension and portosystemic encephalopathy as the most serious consequences of bypass of the hepatic circulation. The ductus venosus (DV) represents a rare foetal PCS and might be persistently patent in newborns after birth. Treatment strategies include surgical ligation and percutaneous device closure. The degree of portal vein hypoplasia limits therapy making liver transplantation the only option in some of them.

Case summary

In a newborn female patient a huge persistently patent DV, known already prenatally, resulted in severe secondary portal vein hypoplasia. She presented with hyperammonaemia, elevated liver enzymes, and pulmonary hypertension. With only diminutive portal venous branches and exceedingly high portal venous pressures during test-occlusion of the DV, shunt closure was not possible. At the age of 2 years more favourable portal venous pressures allowed transcatheter device closure with a nitinol atrial septal defect occlusion device. Pulmonary artery pressures and ammonia levels normalized after the procedure without any signs of portal hypertension.

Discussion

The case highlights the importance of meticulous imaging using balloon occlusion angiography of PCSs like the DV, to search for intrahepatic portal veins. Moreover, portal vein pressure during test-occlusion can identify patients amenable for surgical or endovascular shunt closure. Occlusion devices licensed for other indications like atrial septal defect closure can be used safely in huge PCS vessels in a one-step or staged procedure. Optimal timing of the intervention should be tailored to the patient's needs

Keywords

Portosystemic shunt • Figulla occluder • hepatopulmonary syndrome • Ductus venosus • Abernethy • Case report

ESC Curriculum 2.3 Cardiac magnetic resonance • 2.1 Imaging modalities

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Learning points

- Patients with persistently open ductus venosus should be investigated for portal venous hypertension, as portosystemic encephalopathy and pulmonary hypertension may occur.
- Imaging of the intrahepatic portal vein system by shunt occlusion angiography can detect even diminutive branches that allow porto-caval shunt (PCS) closure and by that may avoid liver transplantation.
- Percutaneous closure of PCSs should be tailored according to the patient's portal vein occlusion pressures, haemodynamics, and hepatic metabolism.

Introduction

The ductus venosus (DV) connects the umbilical vein to the inferior vena cava (IVC) during foetal life and is essential for the normal foetal circulation. Persistence of DV after birth acts as a porto-caval shunt (PCS) as first described by Abernethy, often associated with secondary portal vein hypoplasia.¹

The clinical signs of patent DV after birth are variable with incidentally detected elevated liver enzymes, to a symptomatic severely sick patient, with multiple organ involvement, namely portosystemic encephalopathy and pulmonary arterial hypertension (PAH).^{1,2}

The PCS may be closed percutaneously or surgically. Identification of the portal venous vessels by wedge angiography with occlusion of a PCS, and equally important, measurements of the portal venous pressure is crucial for decision-making concerning treatment.³ We present a rare anomaly, patent DV with severe hypoplasia of the portal veins, which was successfully closed percutaneously in a 2-year-old infant.

Timeline

Foetal life (31 weeks of gestation)	Detection of dilated ductus venosus (DV) and inferior vena cava, cardiomegaly
Newborn period	Jaundice, hyperammonemia, and pulmonary hypertension
3 months	Catheterization: no portal veins visible and pulmonary hypertension
7 months and 18 months	Catheterization: tiny portal veins visible, pulmonary hypertension, portal vein occlusion pressure 32 mmHg and 26 mm, respectively
23 months	Percutaneous closure of the DV with a Nitinol atrial septal defect occluder. Almost immediate normalization of pulmonary artery pressures and ammonia levels
3.5 years	Computed tomography scan: improved filling and size of intrahepatic portal veins with no residual porto-caval shunt
4.5 years	Clinically stable with normal pulmonary pressures and liver metabolism, normal neurological development

Case presentation

A newborn, delivered at 37 weeks of gestation (birth weight 2080 g) had been diagnosed with cardiomegaly, splenomegaly, and dilation of the IVC, splenic vein, and DV already prenatally by ultrasound and magnetic resonance imaging (Supplementary material online, Figure S1). Due to respiratory distress, she was supported by non-invasive ventilatory support and oxygen supplementation. Echocardiography demonstrated a mildly dilated right ventricle, moderate PAH, and a small ventricular septal defect. Laboratory investigations revealed hyperammonemia (75 $\mu\text{mol/L}$), conjugated hyperbilirubinaemia (228 $\mu\text{mol/L}$), and elevation of liver enzymes (aspartate aminotransferase 1.1 $\mu\text{mol/L}$). Metabolic acidosis was ruled out, neonatal screening on inborn errors of metabolism was negative. The newborn was conscious and showed adequate neurology for age and no seizures.

The DV was detected persistently open, collecting blood from the extrahepatic part of the main portal vein. The liver was bypassed by the communication of the DV with the inferior portion of the IVC (Figure 1). No intrahepatic portal branches could be demonstrated by computed tomography (CT) angiography. The splenic and mesenteric veins appeared normal in number and offspring, but dilated together with splenomegaly. Three months after birth cardiac catheterization showed elevated pressures of 52/16/33 mmHg (systolic, diastolic, mean) in the main pulmonary artery. Selective blocked balloon angiography of DV confirmed severely hypoplastic portal venous branches (Figure 2). The mean pressure in the portal veins [portal venous pressure (PVP)] raised from 6 to 20 mmHg during test occlusion. After intense discussions within the team including a paediatric surgeon, the decision was made to leave the DV open at first, as the risk of acute portal hypertension after DV closure was considered too high. At the same time, ammonia levels were acceptable and the child had overall normal psychomotor development, so that there was no reason for urgent treatment. Furthermore, implants for transcatheter intervention would not have been available for the given size of the DV in the small infant. A gradual surgical ligation was discussed if the clinical status would deteriorate. At the age of 7 months and 18 months, PVP under DV occlusion raised again to 31 mmHg and 26 mmHg respectively. At all instances, the mean pulmonary artery pressure was elevated to 28–32 mmHg but well tolerated. Finally, at the age of 23 months, another catheterization revealed lower occlusion pressures in the portal vein circulation and small but existing portal veins were visualized, so the decision for permanent occlusion of the DV was made. Medical treatment to ameliorate the effects of liver bypass of the visceral blood included lactulose, Vitamin substitution (D, E, K and A and B-vitamins), as well as Digoxin and an aldosterone antagonist to support

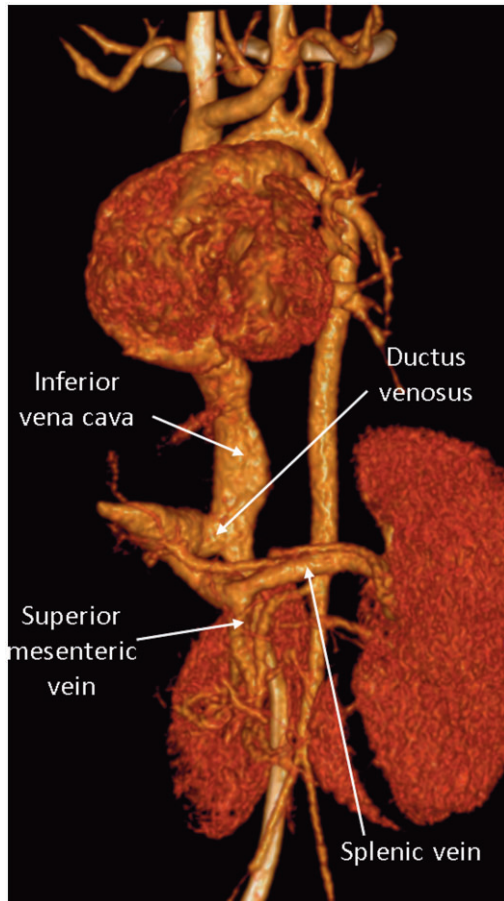


Figure 1 Computed tomography angiography of ductus venosus, three-dimensional reconstruction with delineation of the mesenteric vein and portocaval communication with the inferior vena cava.

the hypertensive right ventricle. Fortunately, the girl thrived well at the 95th percentile of weight, she was a little pale but had proper circulation and blood pressure and no signs of right heart failure, especially no dyspnoea. As ammonia levels remained only slightly elevated under a moderate protein reduced diet, together with the medication mentioned above, detoxification processes in the liver were assumed to some extent to allow normal neurologic development.

As height (88 cm) and weight (14.9 kg) as well as the morphology of the DV seemed appropriate, an endovascular closure was planned.

The procedure was performed under sedation with antibiotic prophylaxis and heparinization. Angiographies in the IVC and DV revealed a diameter of the DV of 13.4 mm and a length of 21.2 mm. Via jugular venous access, the DV was engaged and test-occluded by a Tyshak™ 20 × 12 mm balloon catheter. The PVP was measured 5 mmHg before and 20 mmHg after balloon occlusion. The intrahepatic portal branches were delineated and showed improved size and filling (Figure 3) at that time. Via a 7F long sheath, placed in the DV and according to the measured dimensions, a Figulla Flex II atrial septal defect occluder with a size of 6 mm (central waist) was selected for shunt closure. The retention disks measure 16.5 mm (left atrial disk) and 12.5 mm (right atrial disk) and were calculated to fit within the DV without protrusion into the IVC. The device was implanted in the planned position without obstruction of the IVC and without residual flow across the device 10 min after implantation (Figure 4). The whole procedure was well tolerated by the patient, aspirin was initiated for 6 months for anticoagulation.

After DV closure, ammonia levels normalized. 1.5 years after the intervention, ultrasound and CT scan of portal system confirmed complete occlusion of the DV; an improved filling of the portal venous branches and increase of the vessel diameter were documented (Figure 5).

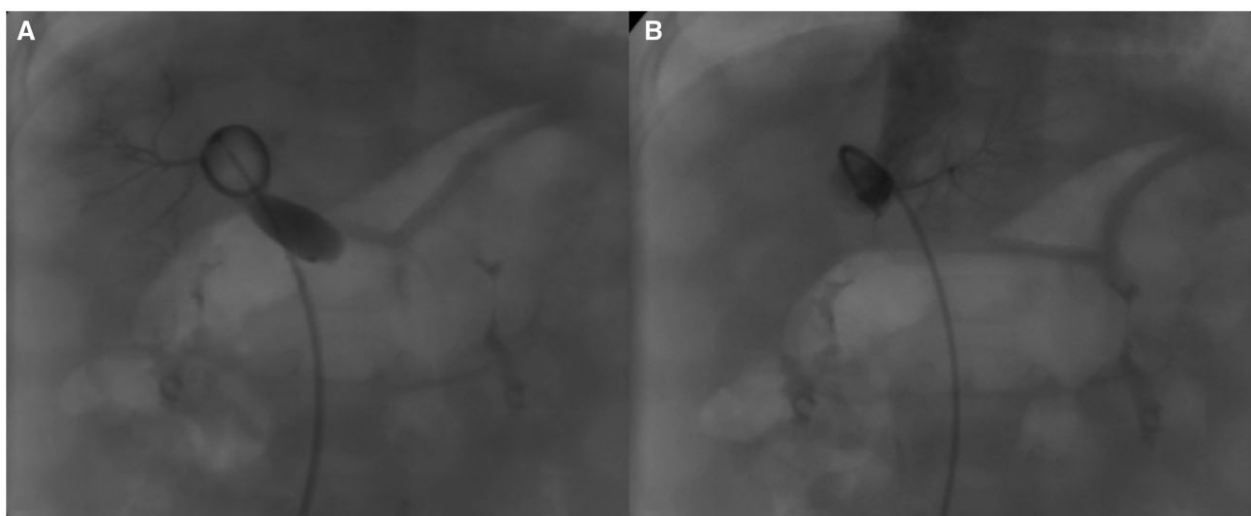


Figure 2 Wedge angiography of hypoplastic right (A) and left portal branches (B) at 3 months of age.

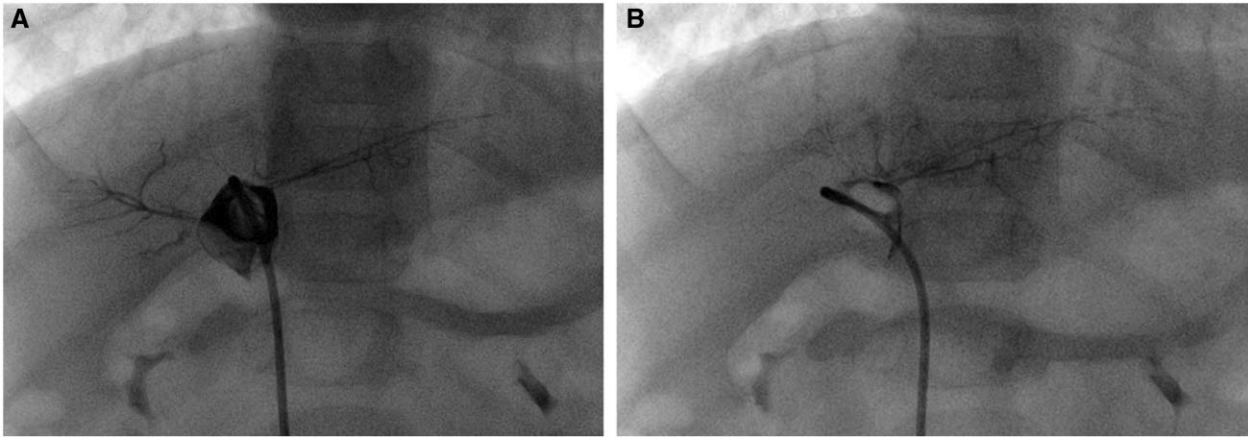


Figure 3 Wedge angiography at the time of transcatheter closure of the ductus venosus (23 months of age), visualizing the still small but grown intrahepatic portal vasculature of the right (A) and left (B) branches.

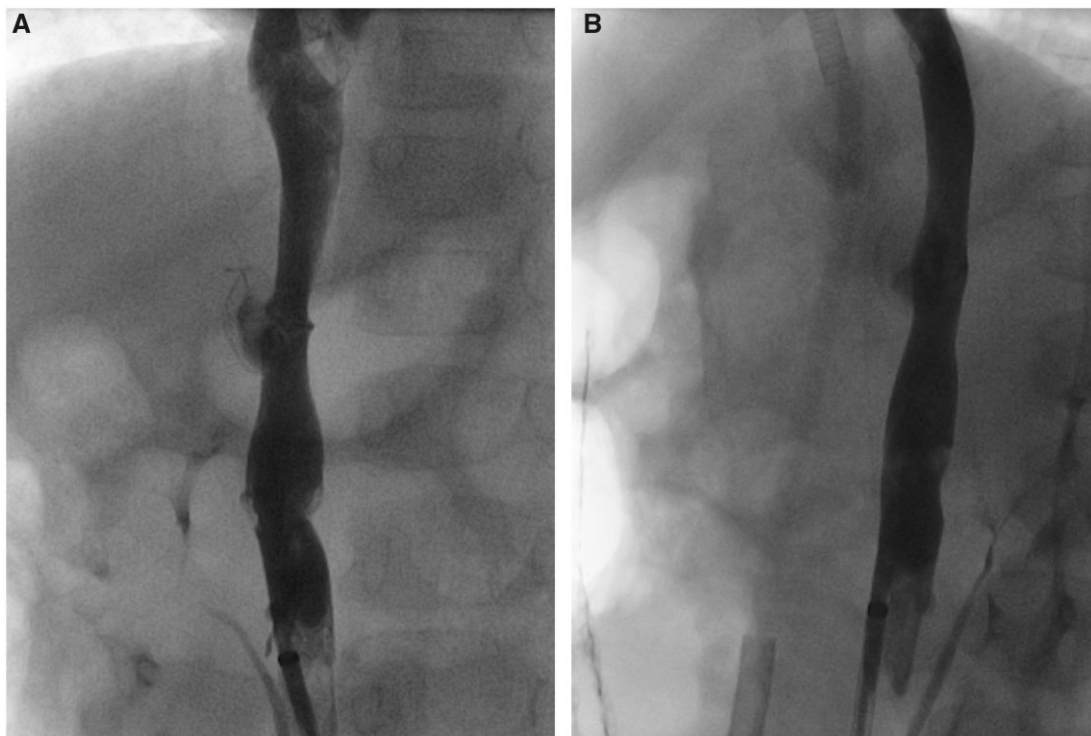


Figure 4 Angiography in the inferior vena cava after placement of an atrial septal defect occluder in the ductus venosus in posterior–anterior (A) and lateral view (B), demonstrating complete occlusion of the ductus venosus and unobstructed venous flow to the right atrium.

Discussion

The DV is dedicated to the normal foetal circulation while persistence after birth presents as an extrahepatic PCS and may lead to severe complications. The majority of symptoms develop in early infancy, but there are patients who remain unrecognized for longer periods.

Neonates may present with congenital cholestatic jaundice during the first days of life associated with hyperammonaemia as in our case. For older patients, symptoms of hepatopulmonary syndrome with triad of liver disease, arterial hypoxaemia, and pulmonary vascular disease, are typical.²

Several classifications were described for PCS.^{1,4} The most commonly used anatomical classification divides PCS into type I with

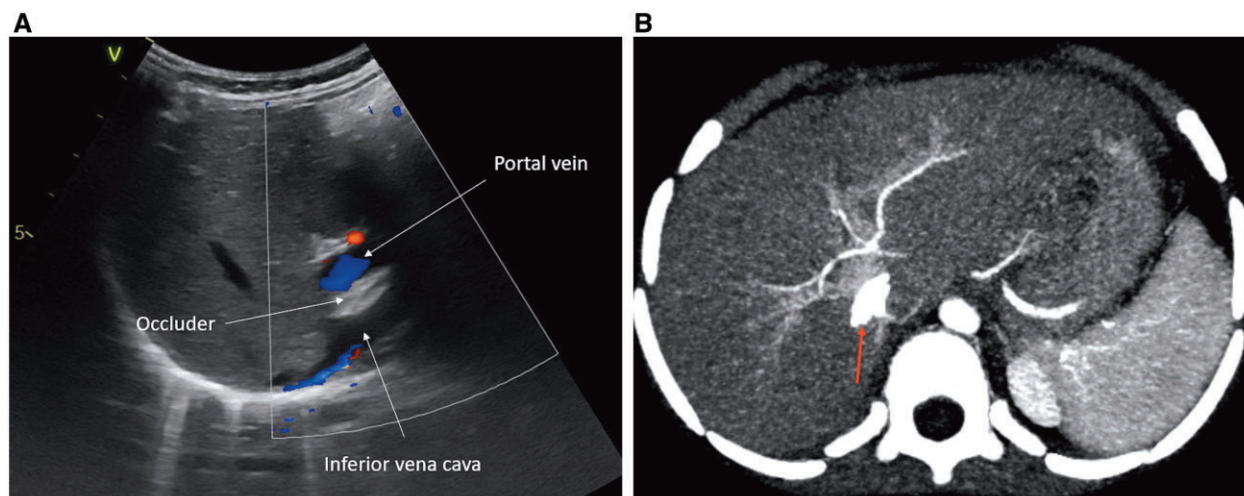


Figure 5 Postprocedural imaging after ductus venosus occlusion (1.5 years after the intervention). (A) Ultrasonography of the liver after ductus venosus closure. Flow in the inferior vena cava is without obstruction, and no residual or additional portocaval shunting is visible. (B) Computed tomography angiography of the liver showing improved filling of the intrahepatic portal veins as well as the location of the atrial septal defect occluder device in the ductus venosus.

absence of portal branches and *type II* with maintained intrahepatic portal perfusion. Usually, liver transplantation has been the only option in PCS I, while PCS II patients are eventually eligible for shunt closure. However, there was a paradigm shift since Kanazawa *et al.*³ could demonstrate by shunt occlusion angiography that also in PCS I an intrahepatic portal system (IHPS) can be detected. They classified PCS into three types depending on the severity of the portal branches hypoplasia.

Accordingly, before closure, the identification of the portal venous system by balloon occlusion shunt angiography and measuring wedge PVP is crucial and can avoid liver biopsy. In severe hypoplasia of the IHPS and exorbitant wedge pressures, a step-wise closure should be planned, to avoid acute life-threatening portal hypertension.

Exact numbers of PVP during shunt occlusion that should not be exceeded are scarce and vary between 20 and 32 mmHg.⁵ In our patient, portal vein occlusion pressures came down within the first 2 years of life and reached 20 mmHg at the time of the intervention. Collateral flow via other PCS during test occlusion of the DV was ruled out and hence falsely low PVP was obviated.

Stepwise closure of the PCS can be achieved either surgically by ligation^{6,7} or by transcatheter intervention with placement of coils, plugs or custom-made flow reducing devices^{8,9} to induce portal vein growth and by such avoid acute portal hypertension. Consequently, more patients might receive treatment by staged shunt closure if a primary single-step approach does not seem advisable.

Our patient represents Abernethy malformation type II with moderately hypoplastic IHPS, which would match with a type 2 of the Kanazawa classification. She could be treated safely in a one-step complete closure procedure by placement of a FigullaTM-atrial septal defect occluder in an off-label location in the DV. Due to the huge dimensions, no other vascular plug would have been appropriate. Post-procedure increased portal vein flow and complete shunt

occlusion were observed. Follow-up of meanwhile 2.5 years revealed no signs of portal venous hypertension, normalized right heart dimensions and pulmonary artery pressure, as well as ammonia blood levels.

Conclusions

The persistence of a DV after birth is a very rare entity of a PCS with variable clinical presentation. Balloon shunt occlusion angiography is mandatory and highly sensitive to detect IHPS and extends the number of patients eligible for treatment. Endovascular DV closure can be performed safely in a staged or single-step procedure depending on portal vein pressure as a minimally invasive approach. The exact timing of a procedure remains difficult, amelioration of PVP by time can occur and patients reach candidacy for treatment. Implants designed for atrial septal defect closure are a valuable tool in huge PCS.

Lead author biography



Univ. Prof. Dr Ina Michel-Behnke is head of the Division of Pediatric Cardiology/Pediatric Heart Center at Medical University Vienna since 2008. She completed a postgraduate research fellowship at the Max-Planck-Institute for Biochemistry in Munich. Her special clinical and scientific interest within paediatric cardiology is interventions and heart failure. From 1998 to 2007, she

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Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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