Cardiology in the Young

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Palliative stenting of the venous duct in a premature neonate with obstructed infradiaphragmatic total anomalous pulmonary venous connection

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Brief Report

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Abstract

In infracardiac, infradiaphragmatic total anomalous pulmonary venous connection, all four pulmonary veins connect to a descending vertical vein that usually drains to the portal vein or one of its tributaries. Obstruction is common, and definitive treatment is surgical repair. We present a case of late-diagnosed infradiaphragmatic total anomalous pulmonary venous connection in a premature neonate who was too high risk for surgery and underwent palliative stenting of the venous duct. We demonstrate the feasibility of a transhepatic approach when umbilical access is no longer available.

Case presentation

A 31-week gestational age female neonate with respiratory distress syndrome required surfactant administration during resuscitation and positive pressure support during the first weeks of life. She subsequently tolerated wean to nasal cannula and was transferred from the neonatal ICU to the feeder grower unit where she was doing well and tolerating advancing feeds. At 3 weeks of age, she developed new desaturations to 88% requiring reinitiation of positive pressure ventilatory support and transfer back to the neonatal ICU. Echocardiogram demonstrated unobstructed infradiaphragmatic total anomalous pulmonary venous connection with a descending vertical vein draining to the portal vein (Fig 1). Given that she was clinically stable and weighed only 1775 g, consensus opinion following our case management conference was to defer surgical repair until a minimum of 36 weeks' gestation to allow for improved maturation and weight gain prior to undergoing cardiopulmonary bypass. In the interim, however, she developed *Klebsiella pneumoniae* pneumonia and worsening hypoxaemia which did not respond to 100% oxygen administration. She became progressively haemodynamically unstable and suffered bradycardic arrest for which she was successfully resuscitated. Echocardiogram demonstrated a newly narrowed venous duct and new severe pulmonary hypertension. Due to her critical status with guarded neurological prognosis, she was considered too high-risk for surgical repair or palliation with extracorporeal membrane oxygenation. Therefore, she was emergently brought to the cardiac catheterisation laboratory for stenting of the venous duct to relieve the obstruction.

Using ultrasound and fluoroscopic guidance, transhepatic access was obtained into the portal vein with a 5-Fr Prelude IDeal sheath (Merit Medical, South Jordan, UT). In order to expedite the intervention and to minimise the risk of losing sheath position, pressure measurements were not recorded. Angiogram through the sheath confirmed a severely stenotic venous duct measuring 1 mm (Fig 2a). The duct was crossed with a 0.014" Choice PT wire (Boston Scientific, Marlborough, MA) and a Maestro microcatheter (Merit Medical, South Jordan, UT) through a 4-Fr JR 2.5 catheter. With the microcatheter positioned in the superior caval vein, the Choice PT wire was exchanged for a 0.018" Steelcore wire (Abbott Laboratories, Chicago, IL). A pre-mounted 4 mm × 16 mm Formula 418 stent (Cook, Bloomington, IN) was implanted across the duct with balloon inflation to 12 ATM. Subsequent angiogram demonstrated excellent stent placement with vigorous flow into the hepatic venous system and the atria (Fig 2b). Coiling of the transhepatic tract was not possible due to the short distance between the liver capsule and the portal vein, so manual compression of the abdominal wall directly over the access site was performed. Post-procedure ultrasound demonstrated a small amount of blood in the abdomen, which remained stable on follow-up evaluation.

The patient tolerated the procedure well with acutely improved oxygenation. However, her pulmonary hypertension did not abate. Two weeks post-implantation, she returned to the cardiac catheterisation laboratory due to increasing congestion on chest X-ray with continued

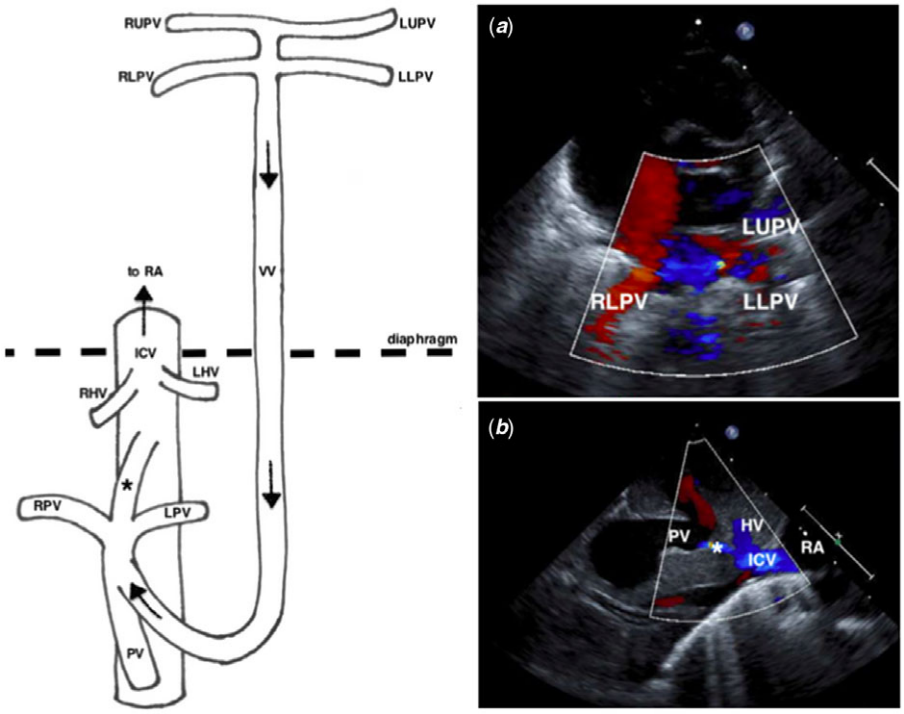

Figure 1. Left panel: schematic diagram of the patient's anatomy. The four pulmonary veins join a confluence that drains via a descending vertical vein (VV) to the portal vein (PV), venous duct (*), and inferior caval vein (ICV) to the right atrium (RA). Right panel (***a***) Parasternal short axis view demonstrating the pulmonary venous confluence posterior to the left atrium. Right panel (***b***) Subcostal view demonstrating an engorged portal venous system with patent venous duct (*) and laminar flow into the right atrium via the inferior caval vein. RUPV, right upper pulmonary vein; RLPV, right lower pulmonary vein; LUPV, left upper pulmonary vein; LLPV, left lower pulmonary vein; LPV, left portal vein; RPV, right portal vein; HV, hepatic vein; RHV, right hepatic vein; LHV, left hepatic vein.

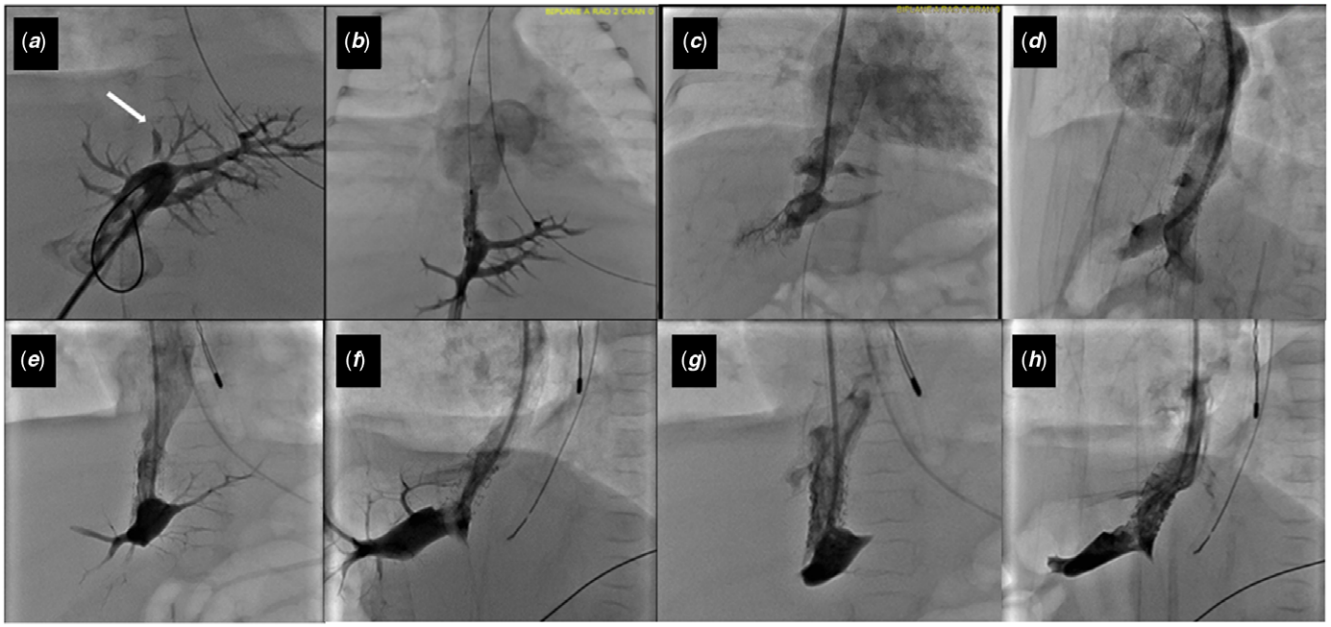

Figure 2. (***a***) Portal vein angiogram demonstrating a severely stenotic venous duct (white arrow). (***b***) Following stent implantation, there is good angiographic flow through the stent, inferior caval vein, and across the atrial septum. (***c***, ***d***) Second intervention, AP and LAT projections respectively. Following balloon angioplasty, there is good angiographic flow across the stent. (***e***, ***f***) Third intervention, AP and LAT projections respectively. There is severe in-stent stenosis present. (***g***, ***h***) Third intervention, AP and LAT projections respectively. Following angioplasty and re-stenting, there is improved angiographic flow through the stent.

systemic right ventricular pressures on echocardiogram. Access was via the right internal jugular vein. Pulmonary artery pressure was systemic at 63/25/42 mmHg, mean pulmonary capillary wedge pressure was 10 mmHg, pulmonary vascular resistance was 4.9 WU × m^2, mean portal venous pressure was 10 mmHg, and mean stent gradient was 5 mmHg. Following balloon angioplasty with a

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6mm Viatrac balloon (Abbott Laboratories, Chicago, IL), there was a residual gradient of 3 mmHg across the stent with unobstructed flow by angiography (Fig 2c–d). Four weeks post-implantation, she was referred for haemodynamic assessment to determine her candidacy for surgical repair. Access was again via the right internal jugular vein. Right ventricular pressure was 133% systemic, mean pulmonary artery pressure was 55 mmHg, mean pulmonary capillary wedge was 15 mmHg, and pulmonary vascular resistance was 6.8 WU × m^2, with mean right atrial pressure of 8 mmHg. Angiography revealed significant in-stent stenosis requiring angioplasty and eventual re-stenting with a 6 mm × 12 mm Formula 418 stent (Fig 2e–h). This resulted in no residual gradient, and angiography revealed unobstructed flow through the stented ductus. However, due to her small size, refractory pulmonary hypertension, and severe haemodynamic instability, she was not deemed a surgical candidate. Following the second reintervention, the parents opted to transition her care to comfort measures only.

Discussion

Total anomalous pulmonary venous connection is an anomaly in which the pulmonary veins do not connect to the left atrium and the pulmonary venous blood returns to a systemic vein or directly to the right atrium. It is classified into four subtypes: supracardiac, cardiac, infracardiac/infradiaphragmatic, and mixed. The definitive treatment for total anomalous pulmonary venous connection is surgical repair. Patients who develop obstruction are often critically ill and typically require emergent surgical intervention. In patients who have comorbidities that make surgery prohibitively high-risk, palliative interventions are sometimes necessary. Stenting of obstructed vertical veins and venous ducts has been described as a palliative measure to promote clinical stabilisation prior to undergoing surgical repair.[1,2] Umbilical venous access is the most frequently reported approach for stenting the closing venous duct.[3–5] Our patient was a late diagnosis making umbilical venous access a non-viable option. Jugular venous access was considered; however, given that there was near complete obstruction of the superior end of the venous duct by procedural echocardiogram, we felt that crossing such a narrow target would be exceedingly more challenging from this approach and would significantly increase the risk of traumatic wire injury. As such, this is the first reported use of direct transhepatic access for palliative venous duct stenting in a patient with obstructed infradiaphragmatic total anomalous pulmonary venous connection.

Given this patient's prematurity and weight, several modifications were made to the procedure. The lab was maintained at the temperature and protocol which we follow for all transcatheter interventions performed on low-weight premature infants. This included warmed heparinised saline and low-volume flushes, with careful attention to minimising procedural blood loss. We elected to use the 5-Fr Merit Prelude IDeal sheath as it tracks easily over 0.014" access wires, has a smooth transition from dilator to sheath, and has an outer diameter that is equivalent to most 4 Fr sheaths. This allowed us to use a pre-mounted Formula stent – which requires a 5-Fr sheath – through the equivalent of a 4-Fr outer diameter, reducing the risk of bleeding after sheath removal. The Formula stent offered the lowest profile at a diameter that we thought would be necessary both for adequate portal vein decompression and for significant re-dilation should it be required in the future. A lower profile coronary stent could have been used, but the potential for re-dilation would be limited. Although coil embolisation of the tract could not be performed in this patient, haemostasis was achieved with manual pressure and ultrasound was negative for significant haemorrhage. Serial ultrasounds may be necessary in the immediate post-procedure period as bleeding is a well-reported complication of transhepatic access.[6,7]

Early diagnosis is important in the management of infradiaphragmatic total anomalous pulmonary venous connection due to the high incidence of obstruction. In our patient, there was low suspicion for CHD in the early postnatal period due to the presence of other comorbidities and diagnosis was delayed. Earlier diagnosis may have allowed for palliative transcatheter stenting in a more stable patient via a more standard interventional approach (umbilical vein or internal jugular vein). Although this salvage transhepatic approach proved to be effective for procedural success, our patient ultimately had a poor outcome due to the refractory nature of her pulmonary hypertension. Despite adequate decompression of the portal venous system with the stent and the documented lack of obstruction at the vertical vein, the pulmonary hypertension persisted. We hypothesise that the persistence of the pulmonary hypertension was due to relative prematurity, ongoing sequelae of the Klebsiella pneumonia, and late diagnosis of obstructed veins. The refractory pulmonary hypertension and clinical instability precluded her from definitive surgical intervention. These points underscore the importance of early recognition and diagnosis of obstructed total veins and either palliative interventions or definitive surgical repair before the sequelae of persistent obstruction sets in. Patients with treatment-responsive pulmonary hypertension can be managed effectively in the post-operative period and may have improved clinical trajectories despite the high risk conferred by low birth weight and prematurity.

Conclusion

We demonstrate that transhepatic venous duct stenting is feasible for the palliative treatment of obstructed infradiaphragmatic total anomalous pulmonary venous connection when the restriction is at the venous duct and umbilical venous access is not an available option. It results in immediate decompression and acutely improved oxygenation for affected patients. This should be considered for patients who have other high-risk comorbidities until they are clinically more suitable for complete surgical repair. Earlier intervention should be considered to improve clinical outcomes and allow for relief of obstruction before severe, refractory pulmonary hypertension develops.

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Conflicts of interest. None.

Ethical standards. Not applicable.

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