

Catheter closure of a patent vertical vein after repair of an unusual total anomalous variant of pulmonary venous drainage

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ABSTRACT

Total anomalous pulmonary venous drainage (TAPVD) encompasses a wide spectrum of anatomical variants. The infradiaphragmatic type is almost always obstructive owing to the interposition of intrahepatic resistances; since almost invariable, the vertical vein inserts on the portal venous system. On correction of this variant, the vertical vein might not be ligated to avoid postoperative pulmonary hypertension. We hereby describe an unusual case of infradiaphragmatic TAPVD, with a vertical vein connected to ductus venosus. Since vertical vein was not ligated, it realized an unrestrictive pathway between the left atrium and the suprahepatic veins which resulted in persistent chylous peritoneal drainage. The patient successfully underwent catheter occlusion of the vertical vein which led to complete resolution of the clinical picture.

Keywords: Patent vertical vein, total anomalous pulmonary venous drainage, transcatheter structural heart intervention, vascular plug

INTRODUCTION

Infradiaphragmatic type is characterized by the confluence of pulmonary veins into a descending vertical vein that, most commonly, connects to the portal system.^[1] Infradiaphragmatic TAPVD is deemed to be always obstructed owing to the intrahepatic vascular resistances. Surgical repair consists of creating a wide communication between the pulmonary veins confluence and the left atrium.^[2] Leaving a patent vertical vein in selected cases may be beneficial in the immediate postoperative period. We describe a rare variant of TAPVD draining into the ductus venosus and the hemodynamic consequences of peculiar physiology that the patent vertical vein realized after repair.

A small series of three cases of significant pretricuspid left-to-right shunt after supradiaphragmatic TAPVD due to a patent vertical vein have been described.^[3] To our

knowledge, this is the first case of successful catheter closure of a vertical vein draining into the venous duct.

CASE REPORT

A 39-week-old gestation male baby was diagnosed at few hours of life, with infradiaphragmatic total anomalous pulmonary venous drainage (TAPVD) [Figure 1]. At admission, echocardiogram showed confluence of the right and left pulmonary veins to a descending vertical vein. Estimated pressure in the right ventricle measured through the tricuspid regurgitation jet was around 60 mmHg. He was operated on the following day because of respiratory distress. The anatomy was confirmed intraoperatively; in particular, the superior and inferior veins were found to drain at slightly different levels.

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A wide anastomosis between the common venous chamber and the left atrium was created, while the descending vertical vein was left patent as a possible decompressing route.^[4]

After 48 h, the baby remained ventilator dependent and developed worsening peritoneal drainage with a triglyceride level of 210 mg/dl and evidence of lymphocytes, supporting the diagnosis of chyloperitoneum. The chest X-ray showed increased cardiothoracic ratio and pulmonary plethora.

A chest computed tomographic (CT) scan demonstrated the confluence of the superior and inferior pulmonary veins connected with a widely open venous duct. An angiography confirmed the unrestrictive drainage of the vertical vein into the inferior vena cava (IVC) through the venous duct. Mean right atrium and pulmonary artery pressures were 14 and 30 mmHg, respectively [Figure 2].

The baby was elected to undergo transcatheter occlusion of the vertical vein. Right internal jugular vein access was achieved under ultrasound guidance. The jugular approach was electively chosen as it was the straightest route through the IVC, venous duct, and vertical vein. An AMPLATZER Vascular Plug II (AVPII™ 10 mm) and a second AVP™ 6 mm were advanced on a 0.038 J guide wire through a 6 F mammary catheter. The choice of the plug sizes was based on the CT and the angiographic images, taking into account the diameter of the target segment of the vein and avoiding to interfere with the pulmonary venous flow. Complete occlusion of the vertical vein was demonstrated by angiography [Figure 3a and b]. The procedure was uneventful and the abdominal chylous drainage settled over the following days. Chest X-ray showed improvement of cardiothoracic ratio and vascularity. At 1 and 6 months and 2 years of follow-up, the baby was asymptomatic and displayed physiological trend of growth. On echocardiographic examination, pulmonary venous flow to the left atrium was unobstructed and the pulmonary pressure was within normal range.

DISCUSSION

TAPVD is complicated by pulmonary venous obstruction in up to 50% of cases. Early clinical distress in patients with infracardiac TAPVD invariably prompts immediate surgery as it is deemed to reflect pulmonary venous obstruction.^[5-7]

In this clinical context, pulmonary hypertension is a well-recognized complication ensuing in the immediate postoperative period which portends a high mortality. One of the technical issues still debated is whether to leave the vertical vein open or not. It has been advocated that an open vertical vein may serve as a pop-off route in case pulmonary hypertensive

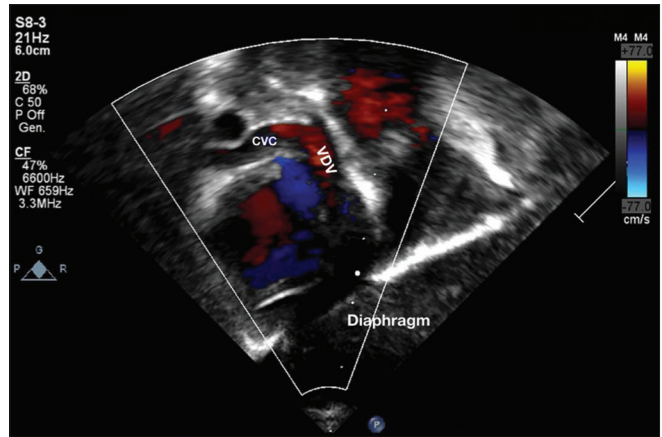


Figure 1: Echocardiographic sagittal subcostal view showing the common venous chamber and vertical vein descending toward the diaphragm

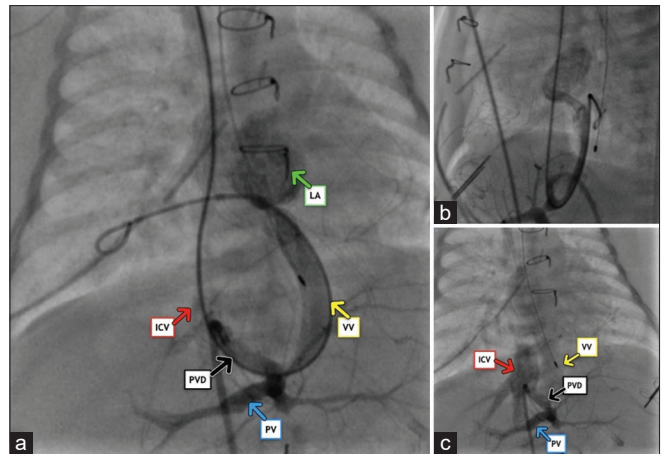


Figure 2: (a) Anteroposterior angiographic image delineating the anatomy. (b) Lateral projection. (c) Angiographic image selecting the connection between the portal venous system and the IVC through the PVD. LA: Left atrium; VV: Vertical vein; PVD: Patent venous duct; PV: Portal vein; ICV: Inferior vena cava

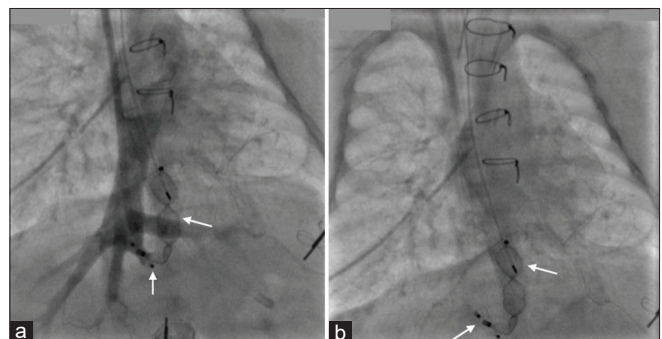


Figure 3: (a) Angiography demonstrating exclusion of the vertical vein (white arrows). (b) Chest X-ray showing final deployment of the two plugs

crises and as an atrial unloading reservoir. In selected cases, this strategy is associated with a lower postoperative mortality.^[4] Due to the particular anatomy characterized by a small left atrium and the

connection of the pulmonary vein at different level, the vertical vein was electively left open, to minimize the risk of postcapillary pulmonary hypertension after the repair.^[3]

The postoperative clinical condition was interpreted as the consequence of the unrestrictive communication between the vertical vein and the IVC, causing persistent significant left-to-right shunt and impaired splanchnic drainage due to increased systemic venous hypertension. Since we did not find any other explanation for the poor clinical conditions, it was felt that immediate interventional treatment was the only therapeutic option. Complete transcatheter occlusion of the vertical vein was feasible and resulted in complete resolution of the chylous drainage, confirming its causative pathophysiological role in the postoperative course.

CONCLUSIONS

This report highlights that the presence of a not ligated vertical vein to patent ductus venosus, following a successful repair of infracardiac total anomalous pulmonary venous drainage (TAPVD), may cause chyloperitoneum. Persistence of heart failure after surgery should prompt recognition of this physiology and catheter closure of the patent vertical vein.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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