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Amplatzer Vascular Plug for Transcatheter Closure of Persistent Unligated Vertical Vein After Repair of Infracardiac Total Anomalous Pulmonary Venous Connection

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Repair of total anomalous pulmonary venous connection (TAPVC) involves anastomosing the pulmonary venous confluence with the left atrium and ligating the vertical vein. Sometimes, the vertical vein needs to be left open as a pop off with the idea that it will close over time. Infrequently an unligated vertical vein may remain patent after repair of infaracardiac TAPVC leading to hemodynamic instability. We report an infant in whom an unligated vertical vein remained patent after the repair of infracardiac TAPVC and caused hemodynamically significant left-to-right shunting. A successful transcatheter closure of persistent patent unligated vertical vein was performed using the Amplatzer Vascular Plug-I device. © 2012 Wiley Periodicals, Inc.

Key words: total anomalous pulmonary venous connection; vertical vein; Amplatzer vascular plug; infracardiac

INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is usually associated with a small noncompliant left atrium, especially in patients with obstructive TAPVC [1]. Typically the vertical vein is ligated at the time of repair to prevent hemodynamic consequences of a residual left to right shunt. Vertical vein ligation during the repair of obstructive TAPVC with smaller left sided chambers has been reported to increase mortality [2]. In a subset of such patients, an unligated vertical vein may improve survival by providing a temporary pop-off if postoperative pulmonary hypertensive crisis should occur [3]. On most occasions, the vertical vein spontaneously closes due to preferential flow to the left atrium as its compliance improves. However, there have been instances where the unligated vertical vein remains patent, which in turn leads to a significant left to right shunt requiring surgical ligation or device closure [2,4–6]. Transcatheter closure of unligated vertical vein in supracardiac TAPVC using PDA occluders has been reported [7,8]. However, device closure of an unligated descending vertical vein in infracardiac TAPVC has not been previously reported. We report an infant whose unligated descending vertical vein remained open following repair of infracardiac TAPVC. A transcatheter closure of persistent unligated vertical vein was performed using the Amplatzer Vascular Plug-I device and a Gianturco coil. In addition, we present a review of the literature on the fate of the unligated vertical vein following repair of TAPVC and subsequent interventions to close it.

CASE REPORT

A 5-day-old male infant presented to our hospital ER with seizure due to hyponatremia (serum sodium 112 mEq L^{-1}). He was born at full term via normal vaginal delivery and had an uneventful antenatal and

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Conflict of interest: Nothing to report.

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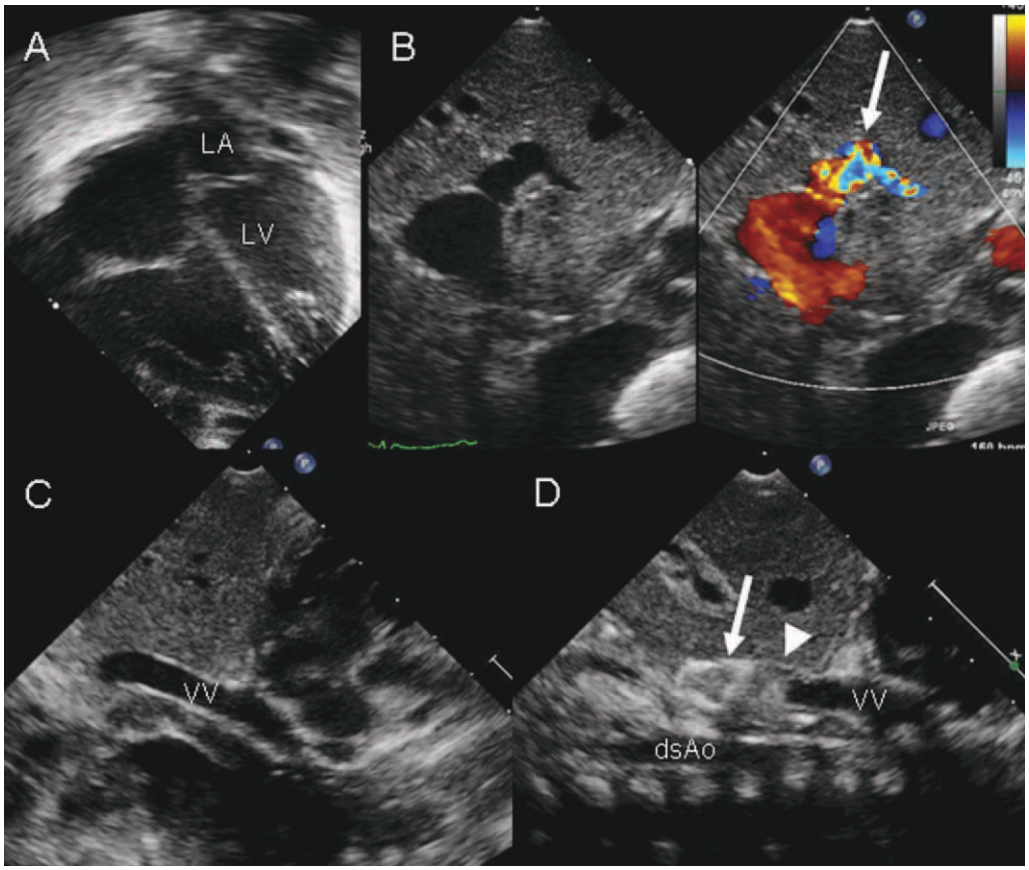

Fig. 1. Echocardiography before and after device closure of the descending unligated vertical vein (A) Apical four chamber view at initial presentation, showing the hypoplatic left atrium (LA) and left ventricle (LV). (B) Subcostal short-axis view an initial presentation, showing the confluence of the descending vertical vein (VV) and portal vein. Doppler interrogation of portal vein showed increased flow velocity suggesting obstruction (arrow). (C) Subcostal long-axis views before transcatheter occlusion of the vertical vein, showing the widely patent vertical vein. (D) The same view after occlusion of the vertical vein, showing that the Amplatzer vascular plug (arrow) and adjunct Gianturco coil (arrow head) occludes the vertical vein. The proximal vertical vein remains patent just after the transcatheter closure. dsAo, descending aorta. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

perinatal course. His birth weight was 3.5 kg. He was initially discharged home at 2 days of age. At presentation, he was tachypneic with a respiratory rate of 80 breaths per minute, heart rate of 168 beats per minute, oxygen saturation of 87% on 1 L min^{-1} oxygen via nasal cannula and blood pressure of 67/51 mm Hg in the right arm. Chest radiography demonstrated pulmonary edema and cardiomegaly. Echocardiography demonstrated an infracardiac type obstructed TAPVC. The pulmonary venous confluence was present behind the left atrium and coursed inferiorly via the descending vertical vein to portal vein (Fig. 1). Doppler interrogation of the vertical vein showed increased velocity (mean of 9.2 mm Hg) suggesting severe obstruction. There was a moderate size right to left shunt at the atrial level with moderate dilatation of right atrium and right ventricle. The left sided chambers were hypoplastic with the left atrial dimension of 7.5 mm (*z*-score -3.2), left ventricular internal diameter in diastole of 10.1 mm (*z*-score -5.2), and mitral valve annulus of 8.4 mm (*z*-score -4.0). Emergent surgery was performed to repair the TAPVC and relieve obstruction.

Intraoperative inspection showed a medium-sized horizontal vein and a larger vertical vein. Using a right atriotomy, a circumferential incision was made in the back of the left atrium. The corresponding horizontal vein was incised and anastomosed with the left atrium. The atrial septal defect was closed using autologous pericardium. Transesophageal echocardiography (TEE) showed nonrestrictive flow through the pulmonary veins, no mitral valve regurgitation and normal left ventricular function. A snare was placed around the descending vertical vein. Temporary occlusion of the vertical vein resulted in severe hypoxemia along with severe mitral regurgitation; release of the snare improved oxygenation with only a mild mitral

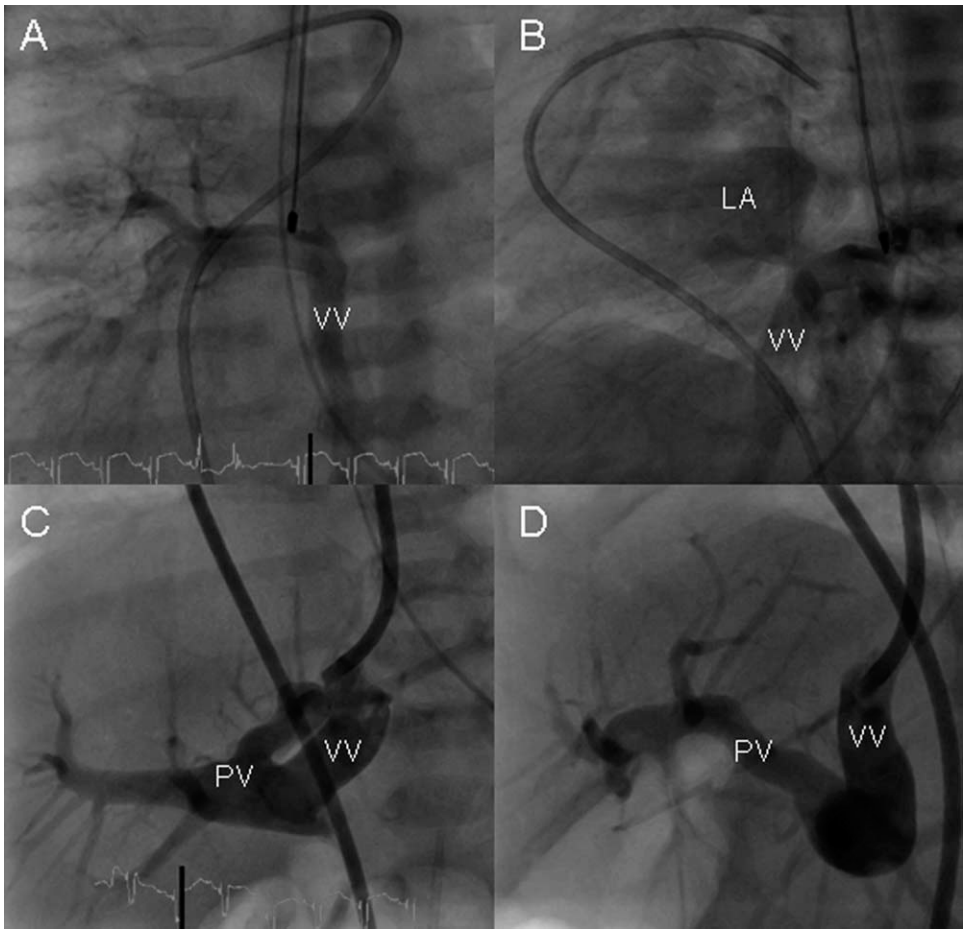

Fig. 2. Cardiac catheterization after the repair of infracardiac TAPVR with unligated vertical vein. The 4-Fr Berman angio catheter was positioned in the right pulmonary artery, where the angiography was performed: (A) anteroposterior and (B) lateral view, showing right pulmonary venous flow drains into the left atrium (LA) and widely patent vertical vein (VV) in the levo phase. The 4-Fr JR-2 catheter was advanced into the distal vertical vein, where the angiography was performed: (C) anteroposterior and (D) lateral view, showing the widely patent vertical vein drained into the portal vein (PV). The distal vertical vein measured 8.2 mm (anteroposterior) and 7.3 mm (lateral) in diameter.

regurgitation. It was surmised that the patient's deterioration was caused by a poorly compliant left sided chamber. Therefore, it was decided to leave the vertical vein open. Postoperative transthoracic echocardiography showed mild tricuspid regurgitation with estimated right ventricular systolic pressures (RVSP) of 34 mm Hg. Postoperatively, nitric oxide was used for pulmonary hypertension for 6 days. Hemodynamically he remained stable and successfully extubated on postoperative day 6. Because of feeding intolerance, he remained in the hospital. On postoperative day 16, the infant developed respiratory distress and increased pulmonary edema on chest radiography despite aggressive diuretics and required reintubation. Blood and respiratory cultures were negative. Echocardiogram demonstrated a widely patent descending vertical vein. The mean pressure gradient across the confluence/left atrial anastomosis was 5 mm Hg. Right atrium and right ventricle were moderately dilated with estimated RVSP of 47 mm Hg. A very small left to right shunt was noted through the interatrial communication.

Cardiac catheterization was performed to assess hemodyanmics with the patient being mechanically ventilated. Using a 4-Fr sheath, a 4-Fr Berman wedge catheter was advanced from the right femoral vein to the branch pulmonary arteries. Right pulmonary artery injection of contrast demonstrated that the anastomosis of pulmonary vein confluence to left atrium was widely patent but the majority of flow coursed inferiorly to the liver through the large vertical vein (Fig. 2). A 4-Fr JB glide catheter was advanced over a 0.035-in. glide wire through the interatrial communication into the left atrium. The mean right and left atrial pressures were 6 and 9 mm Hg, respectively. The mean pressures in the

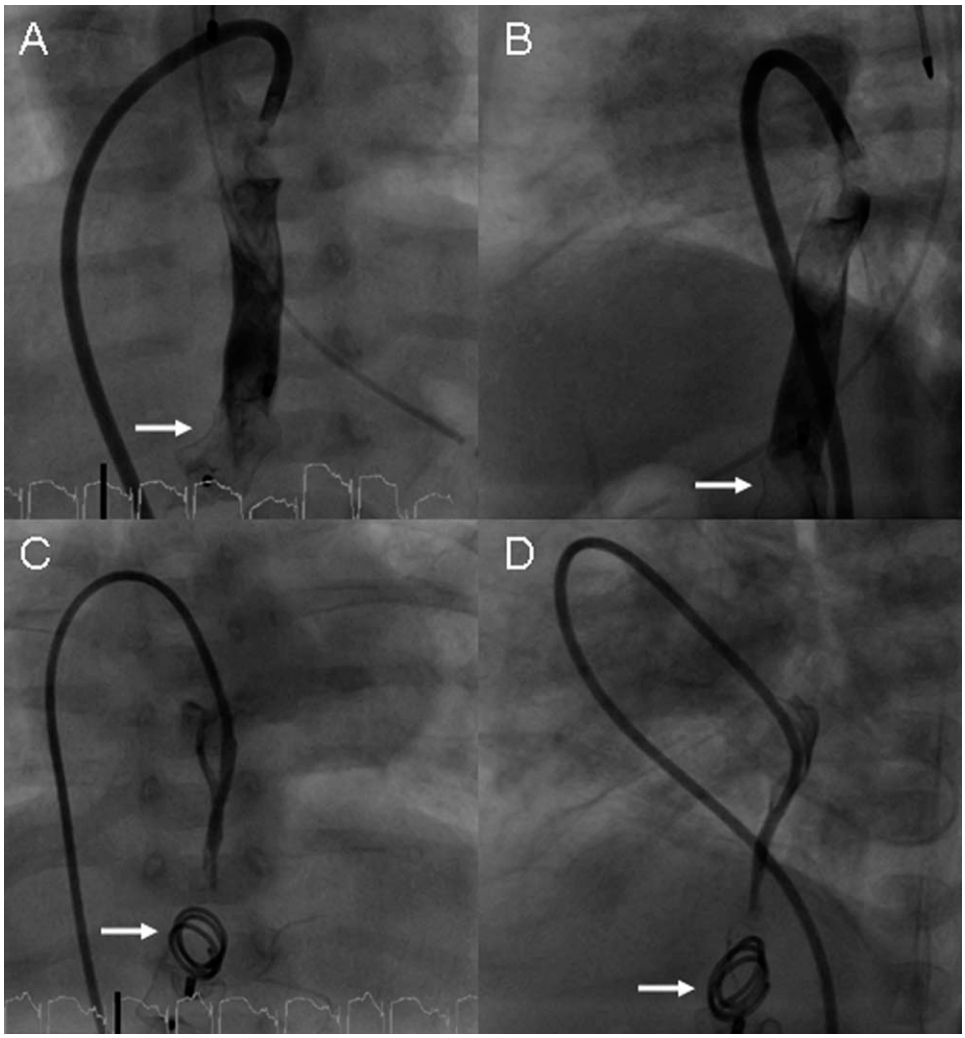

Fig. 3. Cardiac catheterization at transcatheter closure of vertical vein (A, B) and 1 month later (C, D). The angiography in the vertical vein after the deployment of Amplatzer vascular plug: (A) anteroposterior and (B) lateral view, showing the Amplatzer vascular plug (arrow) remains in a stable position and nicely occludes the entire diameter of vertical vein. The angiography in the vertical vein 1 month after the device occlusion: (C) anteroposterior and (D) lateral view, showing the proximal vertical vein was patent but small with the distal part being completely occluded by the Gianturco coil (arrow) and Amplatzer vascular plug.

vertical vein were 13 mm Hg. The RVSP was half the systemic pressure at 36 mm Hg. Cardiac index was 2.5 L min^{-1} m^{-2} with a Q_p/Q_s of 1.7:1. The pulmonary vascular and systemic venous resistances were 1.7 and 19.6 indexed Wood units respectively. The JR-2 catheter was successfully advanced into the confluence using a 0.035-in. glide wire. Angiography at the vertical vein showed that the entire liver was perfused by the vertical vein and emptied into the hepatic inferior vena cava (Fig. 2). In the anteroposterior view, the distal vertical vein measured 8.2 mm and the pulmonary venous confluence measured 9.5 mm. While the descending vertical vein was temporarily occluded by an inflated 8 mm Tyshak II balloon catheter for 25 min, simultaneous TEE showed neither change in ventricular function nor change in the mitral valve regurgitation. Using a 6-Fr sheath, a 6-Fr right coronary artery guide catheter was advanced over a 0.035-in. Rosen wire into the vertical vein. The vertical vein was occluded by a 10 mm Amplatzer Vascular Plug-I and 8 mm 0.038-in. Gianturco coil. Postocclusion angiography in the descending vertical vein and confluence showed minimal residual flow into the liver (Fig. 3). Postocclusion hemodynamic study showed no significant changes in cardiac index (2.6 L min^{-1} m^{-2}) with the RVSP remaining half systemic. The infant was successfully extubated 2 days later and discharged home 3 days after the device placement.

TABLE I. The Fate and Intervention of the Unligated Vertical Vein (VV) After the Repair of Total Anomalous Pulmonary Venous Connection (TAPVC)

References	No of pt	Obstructed TAPVC before surgery	Follow-up period	Postoperative persistent unligated VV		Closure of persistentVV	
				n	Detail	*n*	Surgery/catheter intervention
Supracardiac							
Capsi J 2001	3	3	mean 38 months	0/3	Gradually decreased blood flow in VV and unobstructed anastomosis	0/3	
Ishino K01997	1	NA	23 days	1/1	Unligated VV was closed surgically 12 days after surgery	1/1	Surgery
Kumar RNS 2001	3	NA	3–8 months	2/3	Two patients had patent VV	2/3	Surgery
Cope JT 1997	2	NA	NA	0/2	No flow in VV without no signs of left-to-right shunt	0/2	
Narula N 2007	1	NA	5 years	1/1	Q_p/Q_s of 1.5 with persistent unligated VV	1/1	Amplatzer PDA device
Hausdorf G 1992	1	NA	6 months	1/1	Persistent unligated VV	1/1	Rashkind-PDA-Occluder
Cheung 2005	10	NA	median 4.7 years	5/10	4 pts had $Q_p/Q_s > 1.5$ with unobstructed anastomosis	3/10	Surgery (1 awaiting surgery)
Chowdhury UK 2007	23	15	mean 33.3 months	11/23	12 pts- no flow in VV; 11 pts- right heart failure due to a left-to-right shunt	11/23	Surgery
Shah MJ 2000	2	0	7–12 months	2/2	Persistent unligated VV	2/2	Surgery
Total	46	64% (18/28)		50% (23/46)		46% (21/46)	
Infaracardiac							
Caspi J 2001	18	18	mean 38 months	0/18	Gradually decreased blood flow in VV and unobstructed anastomosis	0/18	
Kumar RNS 2001	1	NA	24 months	1/1	Patent VV portal vein and unobstructed anastomosis	0/1	Awaiting surgery
Cope JT 1997	3	NA	1–106 months	0/3	No flow in VV without no signs of left-to-right shunt	0/3	
Chowdhury UK 2007	2	2	mean 33.3 months	0/2	No flow in VV	0/2	
Jegier W 1967	1	NA	2 years	0/1	No flow in VV	0/1	
Total	25	100% (20/20)		4% (1/25)		0% (0/25)	
Mixed							
Capsi J 2001	1	1	mean 38 months	0/1	Gradually decreased blood flow in VV and unobstructed anastomosis	0/1	
Cope JT 1997	2	NA	NA	1/2	one died of anastomostic stricture, suprasystemic PAP, and widely patent VV	1/2	Surgery
Total	3	100% (1/1)		33% (1/3)		33% (1/3)	

PAP, pulmonary artery pressure; PDA, patent ductus arteriosus VV; vertical vein.

At his 2-week follow-up visit, echocardiography revealed an increased mean pressure gradient across the confluence into the left atrium (11 mm Hg). A small left to right shunt was seen at the atrial level. The right ventricular function was severely decreased with an increase in estimated RVSP to 44 mm Hg. Cardiac catheterization showed suprasystemic right ventricular systolic pressure of 79 mm Hg with femoral arterial systolic pressure of 63 mm Hg. The gradient across the stenotic confluence between the left atrium and pulmonary veins was mean of 20 mm Hg, suggesting severe stenosis. A 6 mm $\times$ 2 cm Tyshak II balloon was inflated on five occasions to a maximum of 6 atmospheres at the stenotic confluence. Postangioplasty, the right ventricular pressure remained nearly systemic with no improvement in the gradient across the stenotic confluence. The angiography in the proximal vertical vein showed total occlusion of distal descending vertical vein (Fig. 3). After the catheterization, he failed extubation and required surgical correction of the obstructed pulmonary venous return with sutureless pericardial technique. At the latest follow-up at 18 months of age, he remains asymptomatic with echocardiography showing nonrestrictive pulmonary venous flow into the LA, no flow in the descending vertical vein, normal biventricular size and function, and normal right sided pressures.

DISCUSSION

TAPVC constitutes 1–1.5% of children with congenital heart disease and is classified based on the site of drainage into the systemic circulation: supracardiac 45%, infracardiac 25%, cardiac 25%, and mixed 5% [9]. Although recent surgical outcomes of isolated TAPVC have improved, with an operative mortality as low as 10%, postoperative pulmonary artery hypertension can be problematic especially obstructed TAPVC [9]. Traditionally, the vertical vein is ligated at the time of TAPVC repair to obviate residual left-to-right shunting [10]. However, an unligated vertical vein may improve survival with a reduction of pulmonary artery pressures due to the vertical vein by acting as a temporary pop-off [3]. A patent vertical vein can unload the small, noncompliant, left-sided cardiac chambers by functioning as a temporary reservoir for pulmonary venous blood after TAPVC repair [2]. Shah et al. reported the beneficial effect of a patent vertical vein to decompress the noncompliant left atrium in two patients with supracardiac TAPVC [11]. Caspi et al. reported that a snare was placed around the vertical vein during surgery in patients with obstructive TAPVC and manipulated according to the patient's hemodynamics in the immediate postoperative period [5]. Patients with preoperative obstructed TAPVC had varying degree of hemodynamic deterioration postoperatively which improved by opening the vertical vein. The vertical vein spontaneously closed subsequently in all their patients with no hemodynamic complications [5]. Although the persistent unligated vertical vein in the repaired TAPVC infant is rare, patency of the descending vertical vein should be followed by serial echocardiography and intervened if indicated.

We performed a review of literature on the fate of the unligated vertical vein after the repair of TAPVC and subsequent interventions to close it (Table I). A total of 74 cases in the literature underwent the repair of TAPVC without ligation of the vertical vein (Table I) [2–8,11–13]. Forty six patients had supracardiac type, 25 had infracardiac type, and 2 had mixed type TAPVC. Obstruction was seen in 64% of patients with supracardiac TAPVC and in all the patients (100%) with infracardiac and mixed TAPVC. In supracardiac TAPVC, 50% of patients had persistent unligated vertical vein and 46% subsequently underwent transcatheter (2 patients) or surgical closure of vertical vein (19 patients). In infracardiac TAPVC, only one patient (4%) had persistent unligated vertical vein and was awaiting surgical ligation. It is speculated that unligated vertical vein in infracardiac TAPVC closes due to the high resistance of the hepatic capillary bed [13].

To our knowledge, our patient is the first reported case of infracardiac TAPVC with a persistent unligated vertical vein causing hemodynamically significant left-to-right shunt in the early postoperative period, and requiring transcatheter closure. Transcatheter closure of vertical vein has been reported in two cases of supracardiac TAPVC. Hausdorf et al. reported a case in whom transcatheter closure of the vertical vein was performed using a11-Fr Rashkind PDA Occluder system 6 months after the repair of TAPVC [8]. Narula et al. reported an infant in whom transcatheter closure of vertical vein using the Amplatzer PDA device was performed at 5 years of age. A restricting band was placed surgically in the vertical vein at time of initial repair of TAPVC [7]. Accordingly, the stenotic waist in the vertical vein was easily accommodated by the device. In our case, 10-mm Amplatzer vascular plug was positioned in the 8.2-mm vertical vein. To ensure total occlusion, one Gianturco coil was deployed at the same time. Total occlusion of the vertical vein was confirmed at cardiac catheterization 1 month later.

CONCLUSION

We report the first case of successful transcatheter closure of persistent unligated vertical vein following repair of infracardiac TAPVC using an Amplatzer Vascular Plug-I and Gianturco coil. A persistent vertical

vein following repair of infracardiac TAPVC is very rare. In rare cases of preoperative obstructive infracardiac TAPVC, keeping the vertical vein open may help in perioperative management. Closure using a percutaneous technique in case of hemodynamically significant shunt in the postoperative period is technically feasible.

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