Dual pathology causing severe pulmonary hypertension following surgical repair of total anomalous pulmonary venous connection: Successful outcome following serial transcatheter interventions

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ABSTRACT
Surgical repair of total anomalous pulmonary venous connection (TAPVC) can be complicated by the development of pulmonary venous stenosis later on. In addition, the vertical vein, if left unligated, can remain patent and lead to hemodynamically significant left to right shunting. We report an infant who required transcatheter correction of both these problems after surgical repair of TAPVC.

Keywords: Amplatzer vascular plug, balloon dilatation, left vertical vein, pulmonary vein stenosis, total anomalous pulmonary venous connection

INTRODUCTION
Total anomalous pulmonary venous connection (TAPVC) constitutes 1–1.5% of children with congenital heart disease.1 Surgical outcomes of isolated TAPVC have improved with an operative mortality of as low as 10%, however, postoperative pulmonary artery hypertension can be problematic especially in obstructed TAPVC.1 Etiology of pulmonary hypertension could range from pulmonary venous hypertension secondary to chronically under filled and small left-sided chambers, pulmonary venous chamber to left atrial anastomosis site stenosis or individual pulmonary vein stenosis to pulmonary arterial (PA) hypertension as a result of large left to right shunt across the patent vertical vein (VV). We report a rare experience of managing both pulmonary venous hypertension followed by PA hypertension by transcatheter measures in a single patient.

CASE REPORT
A 10-month-old boy underwent surgical correction of supracardiac type of TAPVC at 3 months of age when he was weighing 3.2 kg. The left VV was left open at surgery. After an initial uneventful recovery, the child developed shortness of breath, difficulty in feeding, and cough 4 weeks later. Clinical examination revealed tachypnea with respiratory distress, tachycardia, parasternal heave, and loud pulmonic component of the second heart sound. There were bilateral basal crepitations with evidence of mild hepatomegaly. Two-dimensional echocardiography with color Doppler revealed severe PA hypertension secondary to a severe stenosis involving the right common pulmonary vein [Figure 1]. A mean Doppler gradient of 14 mmHg was recorded across this vein. The left VV was patent and draining into the right atrium through the left innominate vein and the right superior vena cava. The patient was taken up for cardiac catheterization with a view to dilate the right-sided pulmonary venous confluence. The right femoral vein was accessed with a 6F venous sheath, and the right pulmonary veins were entered from the left VV through the confluence using a 4F 3.5 Judkins right coronary catheter and a 0.014” BW angioplasty wire (Guidant Corp., Santa Clara, CA, USA). After recording the pressures, diagnostic angiography with a hand

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The pressure in the common confluence was noted to be high (mean = 19 mmHg) without any gradient across the right upper and lower pulmonary veins and across the LA to the venous confluence. The PA pressure was elevated (mean PA pressure was 43 mmHg) and about 2/3 of the systemic pressure (mean = 61 mmHg). Administration of 100% FiO$_2$ for 15 min resulted in a fall in the mean PA pressure to 38 mmHg. After occluding the VV with a low-pressure inflation of a 10 mm Tyshak Balloon (NuMED, Inc., Hopkinton, NY, USA), there was a further fall in the mean PAP to 30 mmHg. Angiogram in the venous confluence now incorporated into the LA revealed a left VV measuring 9 mm draining freely into the left innominate vein [Figure 3a]. In view of the favorable hemodynamics, it was decided to close this communication with a 12 mm Amplatzer Vascular Plug II (St. Jude Medical, St. Paul, MN, USA) which was delivered through a 6F Judkins right coronary guide catheter. The catheter was adjusted to avoid blocking the entrance of the pulmonary veins distally and avoid interfering with the entry of the left jugular or subclavian vein into the innominate vein proximally [Figure 3b]. After performing check angiography to confirm proper position, the device was released. The final angiogram revealed near total occlusion of flow in the left VV and unobstructed flow from the left subclavian vein into the innominate vein [Figure 3c]. The child has remained asymptomatic and continues to show good growth.
and development. The latest echocardiogram revealed normal biventricular dimensions and contractility and unobstructed left internal jugular venous flow.

DISCUSSION

The early presentation and postoperative pulmonary hypertension have the greatest adverse impact on surgical outcomes after repair of TAPVC. Of these, failure to achieve a low-pressure pulmonary vascular system seems to be the variable that most strongly prevents survival and hence needs to be aggressively tackled.[2]

This patient is an unusual case in that he developed two distinct postoperative complications both of which resulted in significant pulmonary hypertension and were severe enough to necessitate a cardiac intervention. Pulmonary venous obstruction ± confluence stenosis is a well-known complication occurring in about 8%-15% of patients after surgical correction of TAPVC.[3] This usually develops within 6 months of primary repair.[2] The site of obstruction is either at the anastomotic site of the confluence with the LA or within the pulmonary veins.[4] It seems that the intimal hyperplasia process probably starts at the anastomotic suture site and extends towards the pulmonary ostia located very close by.[4] The prognosis of the latter as with congenital pulmonary vein stenosis is often poor leading to worsening pulmonary hypertension and eventual death.[4,5] Even with early treatment, the overall results remain disappointing.

The optimal treatment for PVS is yet to be established.[5] Conventional balloon angioplasty, stent implantation, and surgery have all been attempted with limited success.[6-8] More recently, a novel sutureless technique has been introduced for the surgical management of postoperative PVS occurring after the repair of total anomalous pulmonary venous drainage as well as for primary PVS with encouraging results.[3]

Fenestration in the interatrial septum has replaced leaving a patent VV as a pop off in the event of left atrial hypertension secondary to small left heart structures. However, a patent VV can be useful for approaching the LA for the purpose of intervention on the pulmonary veins in the event of development of stenosis.[6]

The results of balloon dilatation of congenital pulmonary vein stenosis are usually unsatisfactory with frequent suboptimal results or rapid recurrence with restenosis. It is possible that conventional balloon dilatation fails because the area of stenosis is transiently stretched during the procedure without rupture of the intima, which is necessary for balloon dilatation to be successful. Recently, the use of cutting balloons has been attempted with some success.[9] Our patient was unusual in that despite the use of the regular balloon technique; there was no recurrence at short-term follow-up. Perhaps the cause of the stenosis was at the suture line and not in the pulmonary veins and therefore responded well to balloon dilatation.

The second unusual feature about our patient was the persistence of the unligated left VV leading to a large shunt and reactive pulmonary hypertension. It is well known that the VV may have to be left open after repair of a TAPVC in children especially in those with preoperative obstruction or whose left heart chambers are small.[10] An unligated VV has been found to reduce PA pressure, decrease perioperative pulmonary hypertensive crisis, and provide better hemodynamics postoperatively.[11]
Although these, VVs are expected to close later, they may remain patent in about half of these cases\(^\text{12}\) and rarely may lead to significant and symptomatic left-to-right shunting as was in our case.

There have been three prior reports of transcatheter closure of this persistent communication using various devices and coils.\(^\text{13-15}\) We decided on an Amplatzer™ plug as it was felt its shape would make it more suitable to totally occlude the tubular VV. Kobayashi et al. used a similar device to close the persistent VV in an infradiaphragmatic TAPVC along with an additional single Gianturco coil deployment to ensure complete occlusion of the communication. We strongly feel that before embarking on transcatheter closure of the unligated vein, it is mandatory to make certain that the hemodynamics are favorable for closure of the communication. The persistent VV sometimes acts as a pop off especially in those with pulmonary venous obstruction or restrictive left heart chambers. Therefore, before we proceeded with closure of the VV at the second procedure, we ruled out any pulmonary venous or confluence stenosis by recording the gradients across the pulmonary veins and confluence in addition to performing pulmonary venous angiography. Furthermore, temporary balloon occlusion of the VV resulted in fall of PA pressure suggesting that at least part of pulmonary hypertension was reactive.

Although access to the left VV is not a problem, tracking a large sheath all the way into the vein could be. Therefore, our preference was for a right coronary guiding catheter to deliver the device. Finally, accurate placement of the plug well within the VV is imperative to avoid blocking the pulmonary veins distally and entry of left jugular or subclavian vein into innominate vein proximally. The dramatic clinical response of the baby to closure of the VV vindicated the decision to close it.

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Conflicts of interest
There are no conflicts of interest.

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