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CASE REPORT

Persistent Patent Vertical Vein After Repair of Total Anomalous Pulmonary Venous Connection (TAPVR): A Rare Cause of Hypoxemia Post-Fontan Procedure

Aditi Gupta[1] · Megan Gunsaulus[1] · Alexandra Erdmann[1] · Mario Castro Medina[1] · Tarek Alsaied[1] · Jacqueline Kreutzer[1]

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Abstract
Vertical vein (VV) ligation during total anomalous pulmonary venous return (TAPVR) repair is controversial. While some surgeons prefer ligation of the VV to prevent adverse sequelae of shunting across it and to promote flow through the newly created anastomosis, others leave it to serve as a "pop off valve" to the left heart structures, which are believed to be hypoplastic and noncompliant, presumably contributing to a more favorable post-operative outcome. We report two patients post-Fontan procedure, who underwent cardiac catheterization to explore the etiology of hypoxia and were found to have a persistent VV responsible for right to left shunting. Both patients underwent closure of the VV with improvement in the cyanosis and clinical course. These cases provide evidence supporting surgical ligation of the VV at the time of TAPVR repair, especially in patients with single ventricle.

Keywords Heterotaxy · Total anomalous pulmonary venous return (TAPVR) · Fontan procedure · Cardiac cath · Vertical vein

Case 1

A 16-year-old male with asplenia syndrome, right atrial isomerism, unbalanced common atrioventricular canal, double outlet right ventricle, pulmonary valvar and subvalvar stenosis, infracardiac total anomalous pulmonary venous return (TAPVR), and diaphragmatic hernia initially underwent bilateral bidirectional Glenn procedure, pulmonary artery banding, TAPVR repair, and diaphragmatic hernia repair at an outside institution at 1 year of age. He then underwent his pre-Fontan cardiac catheterization and was not deemed to be a suitable candidate for Fontan. He was subsequently referred to our center for further evaluation. He underwent diagnostic cardiac catheterization which was suggestive of overall favorable hemodynamics, significant aortopulmonary collateral (APC) burden, and bilateral pulmonary arteriovenous malformations (PAVMS). He also underwent preoperative cardiac magnetic resonance and after review of all diagnostic studies, it was decided to proceed with fenestrated extracardiac Fontan procedure. His immediate post-operative course was complicated by severe hypoxemia and bleeding requiring ECMO cannulation. Early postoperative cardiac catheterization was performed to explore the possible etiology of his severe hypoxemia.

Findings

Inferior vena cava (IVC) angiography revealed a large connection from the IVC to the common atrium which represented the residual TAPVR vertical vein (VV) to the IVC now communicating with the left atrium after TAPVR repair. The blood was preferentially shunting through the VV into the left atrium, acting as a large pop-off and right to left shunt. There was notably minimal antegrade flow into the Fontan conduit and bilateral pulmonary arteries despite a wide open connection, which explained the profound cyanosis. Initially, the VV was closed with an Amplatzer muscular VSD occluder in the catheterization laboratory. However,

✉ Jacqueline Kreutzer
jacqueline.kreutzer@chp.edu

[1] Division of Pediatric Cardiology, Department of Pediatrics, University of Pittsburgh, Heart Institute, UPMC Children's Hospital of Pittsburgh, Pittsburgh, PA, USA

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given persistent residual flow through the device after 24 h, he underwent surgical removal of the device and patch closure of the entry of the VV into the left atrium. Following this procedure, the patient had significant improvement in his saturations, and he was successfully decannulated and extubated few days later. His subsequent course was complicated by hemoptysis in the setting of a significant APC burden requiring multiple transcatheter embolization procedures. He eventually was discharged home and has done clinically well at follow-up with no cardiovascular symptoms and oxygen saturation in low 90s in the setting of a patent fenestration (Figs. 1, 2, 3).

Case 2

A 20-month-old male with history of hypoplastic left heart syndrome and obstructed infracardiac TAPVR initially underwent Norwood procedure with Blalock Taussig Thomas shunt and TAPVR repair at 1 week of life. This was followed by a bidirectional Glenn procedure with take down of the aortopulmonary shunt at 9 months of age and a fenestrated lateral tunnel Fontan procedure at 1.5 years of age at an outside institution. His post-operative course was complicated by chronic hypoxia and plastic bronchitis

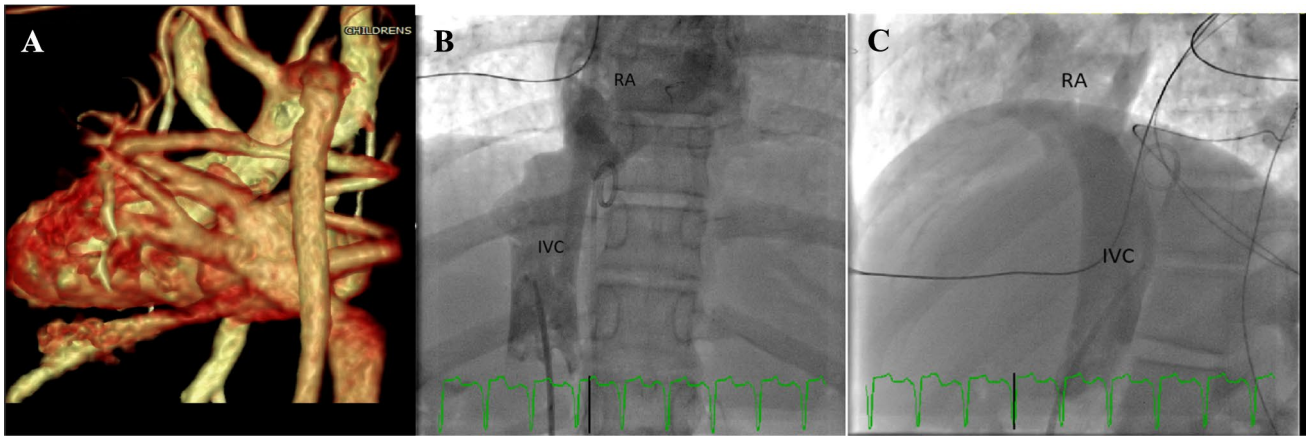

Fig. 1 **A** 3D reconstruction of pre-Fontan cardiac MRI demonstrating the IVC draining into the atrium. **B, C** AP and lateral view of preoperative Cath showing unobstructed flow of contrast from IVC to right atrium with filling of VV posteriorly but given the lack of septation between the two pathways within the atrium, the contrast swirls and the VV presence is missed

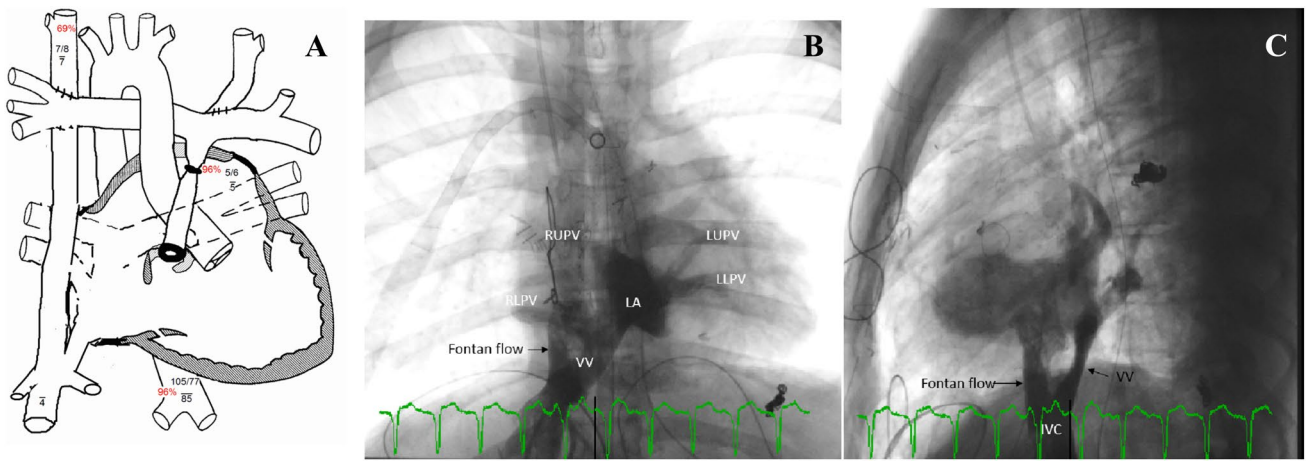

Fig. 2 **A** Hemodynamic diagram for cardiac catheterization performed post-Fontan surgery demonstrating—underlying anatomy (s/p right bidirectional Glenn and pulmonary artery band, right atrial isomerism, unbalanced atrioventricular canal, double outlet right ventricle,valvar and subvalvar pulmonary stenosis, infradiaghragmatic TAPVR repair) with patent Fontan baffle and VV connection from IVC to atrium which was diagnosed on the cardiac catheteriation done after Fontan. **B, C** AP and lateral view: IVC injection demonstrates large connection between IVC and atrium consistent with remnant VV serving as a right to left shunt with minimal antegrade filling of Fontan conduit

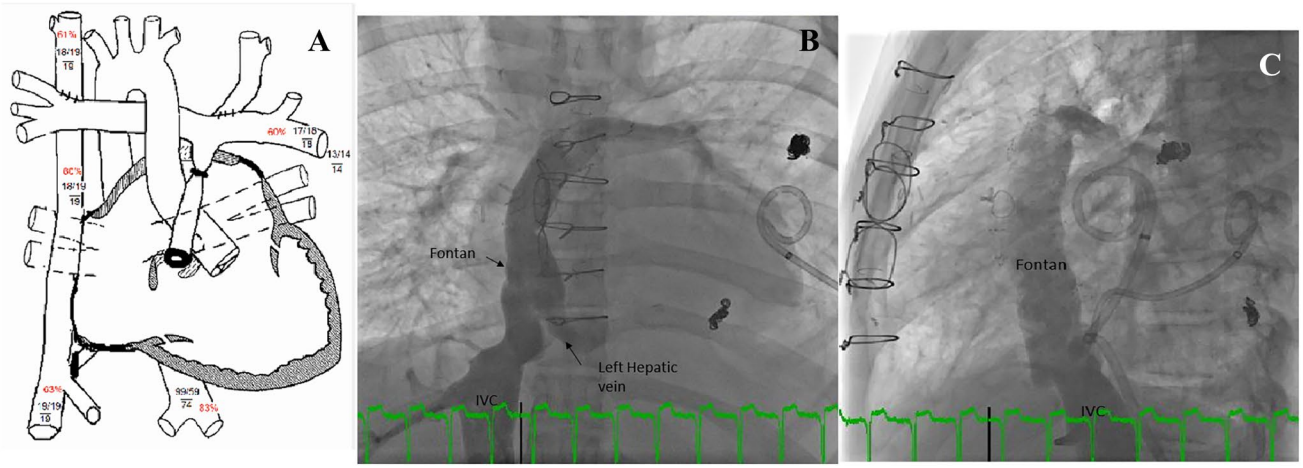

Fig. 3 **A** Hemodynamic diagram demonstrating underlying anatomy s/p surgical ligation of the VV and removal of the plug. **B, C** AP and lateral view of IVC injection: post-surgical ligation of the VV, there is unobstructed flow of IVC into the Fontan conduit and no filling of the VV is seen

and was transferred to our center for heart transplant evaluation. His echo showed a patent pulmonary venous confluence into the left atrium and a patent Fontan fenestration. He underwent cardiac catheterization as a part of his transplant evaluation at our center.

Findings

A catheter was advanced from the IVC through the Fontan fenestration to the right atrium and into the left atrium in an attempt to access the pulmonary veins. The catheter took an unusual course that projected inferiorly and crossed the diaphragm revealing a portal vein communication to the left atrium, consistent with a VV. Angiography was performed using a balloon wedge catheter to occlude the VV which revealed complete filling of the portal vein and branches within the liver. When the balloon was deflated, the contrast flowed to the left atrium consistent with a right to left shunt which was likely contributing to chronic hypoxia and need for oxygen. In addition this was a porto-left atrial shunt, leading to reversal of flow in the portal vein being hepatofugal, rather than hepatopedal (normal portal flow). This anatomy was consistent with a persistent VV that drained the pulmonary venous confluence to the portal vein which had been left unligated at the time of the TAPVR neonatal repair. Test occlusion of the VV was performed with no increase in portal vein pressures and therefore this connection was successfully closed with a 6 mm × 6mm Amplatzer vascular plug II. The arterial saturations improved following the procedure. However his course continued to be complicated by plastic bronchitis refractory to medical management and therefore he eventually underwent orthotopic heart transplant at 5 years of age and has done well thereafter, with normal oxygen saturations (Fig. 4).

Discussion

Ligation of the anomalous VV during TAPVR repair is controversial. While some centers propose ligation of the VV to prevent adverse sequelae of residual shunting across the VV, others leave it unligated to serve as a "pop off valve" to the left heart chambers, which are believed to be hypoplastic and noncompliant.

Cope et al. [1] in a review of 23 infants concluded that VV ligation at the time of repair is not necessary and is in fact undesirable especially if the left heart structures are small. In another review by Caspi et al. [2] of 34 patients with TAPVR, the group concluded that a patent VV, especially in infracardiac TAPVR contributes to a favorable outcome following surgery.

While unligated VV has been found to be associated with better postoperative outcomes after TAPVR repair [2], long-term sequelae of persistent cardiac shunting through these unligated VVs have also been reported. While most patients with these residual shunts can be observed, some of them may become hemodynamically significant and require surgical or transcatheter closure. [3–5]. In a series of four infants in whom, the VV was left open at the time of surgery, three underwent delayed surgical/transcatheter closure of the VV in view of persistent left to right shunt [6].

The debate on whether or not to ligate the VV at the time of TAPVR repair has been centered primarily on patients with biventricular circulation. There is very limited data on relevance of patency of a VV in patients with univentricular hearts. Overall, the population of patients with TAPVR and

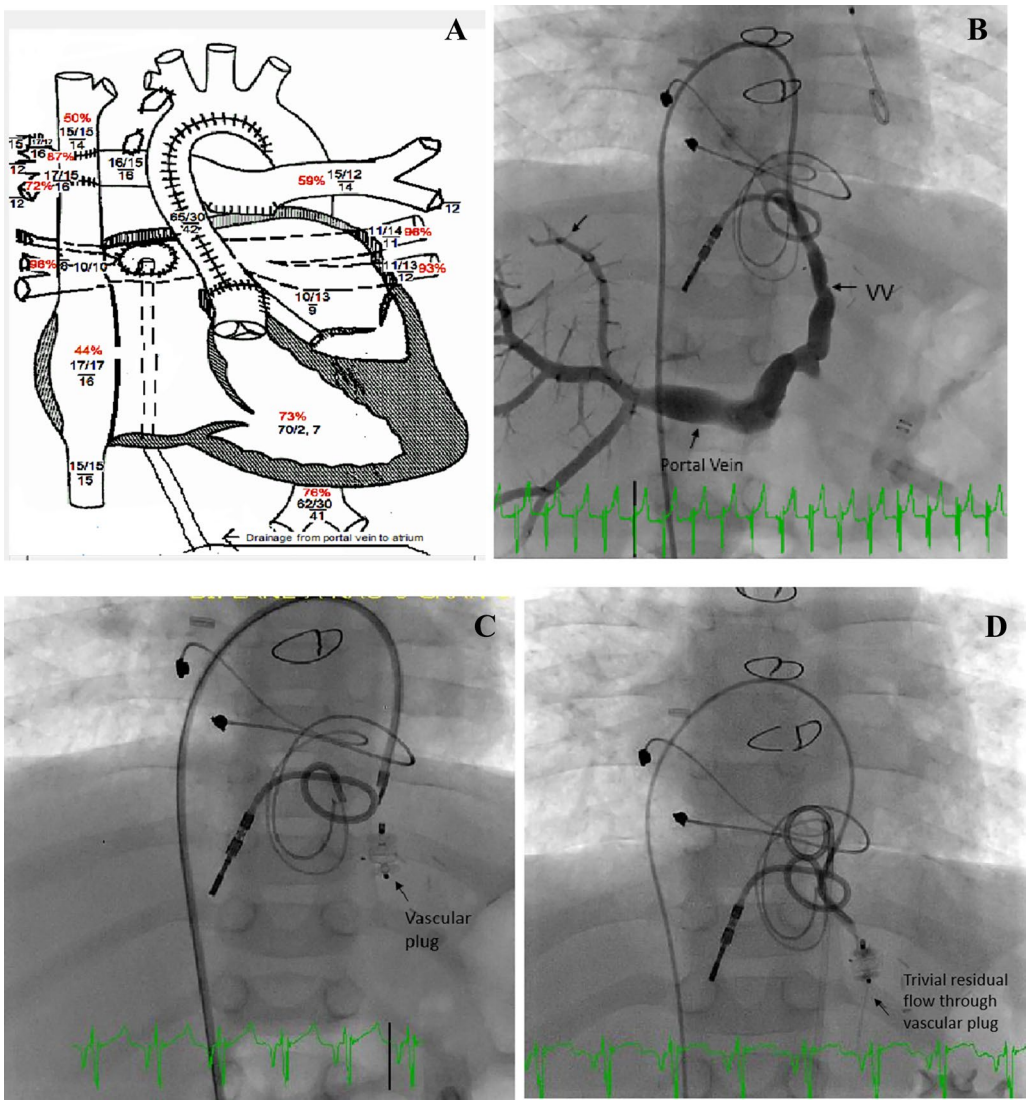

Fig. 4 **A** Hemodynamic diagram demonstrating HLHS s/p Norwood and infradiaphragmatic TAPVR repair most recently Fontan completion. **B** Catheter course from lateral tunnel Fontan through the fenestration into the right atrium to left atrium and into the portal vein showing communication of portal vein to the left atrium s/o residual VV serving as a right to left shunt. **C** Deployment of vascular plug in this connection. **D** Injection in VV post vascular plug release s/o trivial residual flow through the vascular plug

single ventricle physiology has been reported to have poor outcomes (survival of 50–80%) [7–9]. In a review of 207 patients with TAPVR repair, 10 years survival was only 51% in those who have univentricular hearts as compared to 85% in biventricular hearts [10, 11].

It is important to understand that, while in biventricular hearts the residual VV will act as a source of left to right shunting due to higher LA pressures, the physiology is very different in single ventricle circulation. Depending on the anatomic location, there may be no shunting whatsoever across the residual VV during the first two stages of single ventricle repair as the pulmonary veins and unligated VV are draining to chambers of equal pressure. This makes diagnosing the presence of a residual VV a challenge in these surgical stages if detailed operative information from the TAPVR repair is not available. After the Fontan procedure, the systemic venous pressure is higher than the left atrial pressure leading to flow reversal through the VV, still connected to the pulmonary venous confluence at the left atrium. This can result in significant right to left shunting acting as a large Fontan fenestration leading to hypoxia postoperatively (12).

In our first case, the VV caused a significant right to left shunting in the immediate post-operative period with decreased antegrade flow across the Fontan conduit leading to severe hypoxemia. Ligation of the VV, improved both the antegrade blood flow across the Fontan conduit

and oxygen levels leading to successful ECMO decannulation and progression towards extubation.

In the second case, the VV was smaller in size but still was a significant contributing cause of chronic hypoxemia. In this particular variant, when a VV drains to the portal system and is left unligated, it may lead to late portosystemic shunts—with secondary development of PAVMS. However, in this particular case, we did not diagnose these, possibly secondary to there not having been enough time for them to develop. While there was mild improvement in saturations after the transcatheter closure of the VV, this patient had failing Fontan physiology for other reasons and eventually required heart transplantation given other serious manifestations of post Fontan lymphatic dysfunction including plastic bronchitis.

In these illustrative cases of persistent patency of the VV post-TAPVR repair, we demonstrate sizable right to left shunts, causing significant hemodynamic burden after the Fontan procedure supporting surgical ligation at the time of original repair.

Conclusion

Patency of a VV after the TAPVR repair can have a significant detrimental impact on patients with single ventricle physiology. Therefore surgical ligation of the residual VV is recommended.

Authors Contribution All authors were involved in clinical care of the patients and contributed to conception, image acquisition and reviewed the final manuscript.

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Declarations

Conflict of interest The authors declare no conflict of interest.

Availability of Data and Material Patient records.

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