

Elective Stent Implant in the Obstructed Vertical Vein of Supracardiac Total Anomalous Pulmonary Venous Connection Prior to Operative Repair

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Abstract

Background: Total anomalous pulmonary venous connection (TAPVC) comprises 2% of congenital heart disease cases. Obstructed TAPVC typically presents with respiratory distress secondary to pulmonary congestion. We report a case of an infant patient who was electively referred to catheterization for stent placement to relieve vertical vein (VV) stenosis. Our objective was to prevent the emergent need for surgical intervention while allowing additional growth before surgery.

Case Presentation: A 7-day-old, late pre-term, small for gestational age male infant was transferred from an outside institution. He was initially placed on nasal cannula due to oxygen saturation around 80% but progressed to continuous positive airway pressure and had a chest X-ray suggestive of pulmonary edema. Echocardiography revealed supracardiac TAPVC, a small apical muscular ventricular septal defect, and a moderate secundum atrial septal defect. On admission, the patient was clinically stable with a baseline oxygen saturation of 72% on 40% oxygen. Echocardiography confirmed supracardiac TAPVC and also showed an obstruction with a mean gradient of 22 mmHg in the VV. The desire to optimize the patient's clinical stability led to the decision to undergo cardiac catheterization for stent implantation in the VV. Immediately following the procedure, the patient's hemodynamics improved, with a pressure gradient between the pulmonary venous confluence and the left innominate vein of 4 mmHg.

Conclusions: Over the last decade, surgical outcomes

of TAPVC repair have improved with better control of pulmonary hypertension and preoperative clinical stabilization due to more aggressive medical management. This case presents an opportunity to consider an elective interventional strategy that palliates the disease to prevent an urgent need for definitive repair.

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Key Words

TAPVC • Interventional • Obstructed • Vertical vein • Elective

Introduction

Total anomalous pulmonary venous connection (TAPVC) is a rare cardiac defect that comprises 2% of congenital heart disease cases [1]. TAPVC encompasses different anatomic subtypes in which pulmonary veins fail to connect directly to the left atrium and drain to the right atrium via an anomalous venous connection [2, 3]. Supracardiac TAPVC is the most common type, comprising about 45% of cases [2]. A left-sided vertical vein (VV) accounts for 70% of the connections between the pulmonary confluence and the right atrium, and stenosis occurs in approximately 40% of cases [3].

Obstructed TAPVC typically presents with respiratory distress secondary to pulmonary congestion,



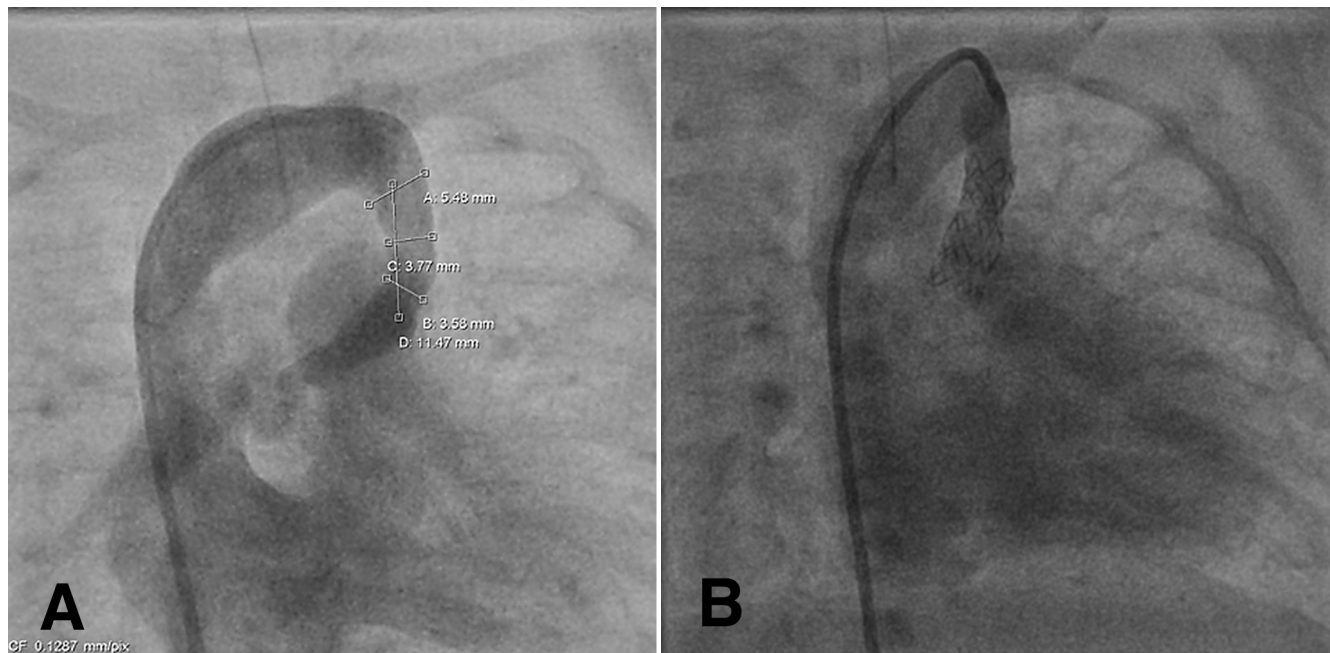


Figure 1. Panel A. Cardiac angiography prior to stent placement. The confluence narrowing is 3.6mm with the superior aspect of the vertical vein measuring 5.5mm. Panel B. Cardiac angiography after the two stents were implanted, with the second stent telescoped proximally within the initial stent.

cyanosis, and metabolic acidosis [4]. It is traditionally considered a surgical emergency. Emergent stenting of the VV is rarely reported but has been performed when a patient is considered a poor candidate for surgical repair [4-6]. Pulmonary venous obstruction is associated with poor prognosis and a high risk of operative mortality [7, 8].

Here, we report a case of an infant patient who was electively referred to catheterization for stent placement to relieve VV stenosis. Our objective was to prevent the emergent need for surgical intervention while allowing additional growth before surgery.

Case Presentation

A 7-day-old, former 36 6/7 week, 2.4 kg, small for gestational age male infant was transferred from an outside institution. At the time of delivery, he was noted to have poor respiratory effort and a heart rate below 100 beats per minute. Chest compressions were provided for less than 1 min, and his APGAR scores were 5 and 7 at 1 and 5 min, respectively. The patient was noted to have oxygen saturation of roughly 80%, requiring blow by oxygen. He was initially placed on

nasal cannula, but over the next 7 days progressed to continuous positive airway pressure and had a chest X-ray suggestive of pulmonary edema. An echocardiogram at that time revealed supracardiac TAPVC, a small apical muscular ventricular septal defect with right-to-left shunt, and a moderate secundum atrial septal defect with right-to-left shunt.

On admission, the patient was stable with a baseline oxygen saturation of 72% on 40% oxygen. An echocardiogram was performed that confirmed supracardiac TAPVC but demonstrated an obstruction in the VV with a mean gradient of 22 mmHg. All pulmonary veins drained to a confluence behind the left atrium and communicated to the innominate vein via a VV.

The patient's size and the desire to optimize his clinical stability led to the decision to undergo cardiac catheterization for stent implantation in the VV (Figure 1A). Femoral vein access was achieved, and a 4-F angled Glide catheter (Terumo, Somerset, NJ, USA) was advanced prograde into the VV. A 17 mmHg pressure gradient between the pulmonary venous confluence and the left innominate vein was recorded. A V-18 Control wire (Boston Scientific, Marlborough,

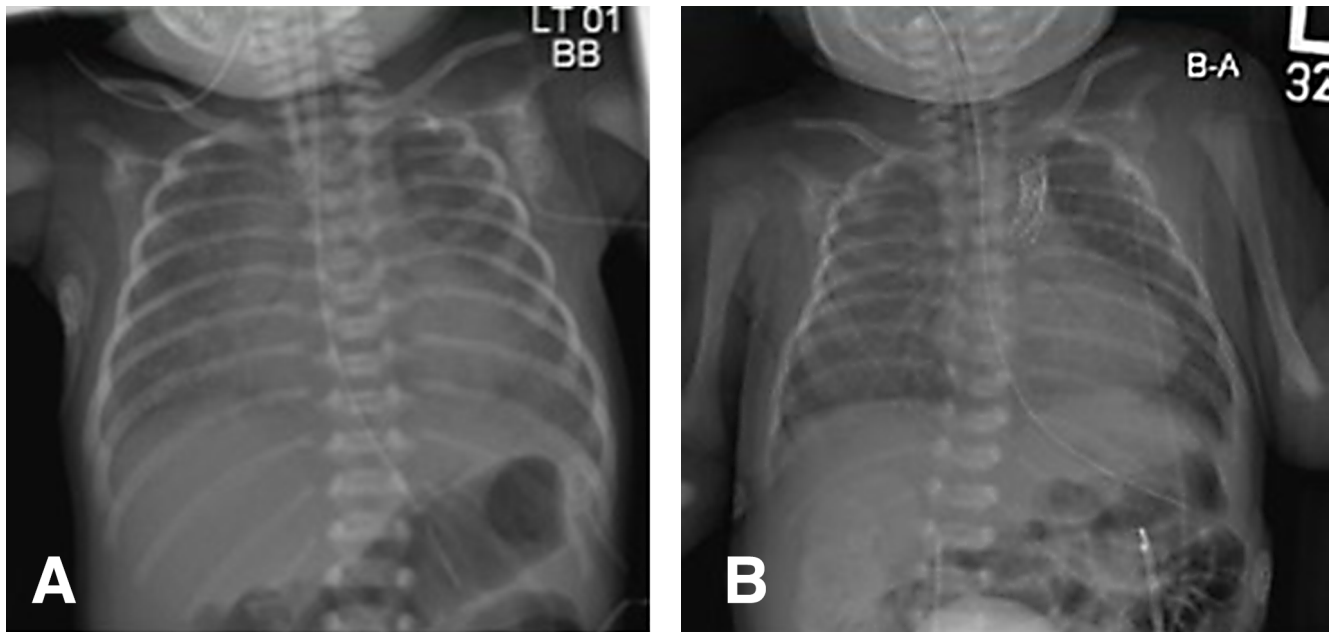


Figure 2. *Panel A.* Chest radiograph 1 day prior to stent placement. Diffuse pulmonary edema is seen. *Panel B.* Chest radiograph 1 day after stent placement in the vertical vein. The stents are visible along the left sternal border. There is improvement in the pulmonary edema.

MA, USA) was advanced into the right lower pulmonary vein, and the Glide catheter was exchanged for a Palmaz Blue 6 × 16 mm stent (Cordis, Fremont, CA, USA) that was deployed in the VV. Due to a persistent residual gradient, a Palmaz Blue 6 × 12 mm stent was telescoped proximally within the prior stent to ensure resolution of the obstruction within the VV (Figure 1B). Repeat hemodynamics demonstrated a pressure gradient between the pulmonary venous confluence and the left innominate vein of 4 mmHg. Oxygen saturation improved to 95% on 50% oxygen, which was reduced to room air over 48 hours. Repeat chest X-ray showed improvement of the pulmonary edema (Figure 2A and 2B).

Over the following 3 weeks, pulmonary edema developed again despite medical management with diuretics. At 26 days of age, the patient had gained approximately 200 g and underwent TAPVC repair. He recovered well, was extubated, and weaned to room air by postoperative day 5. He was discharged on postoperative day 21 on once daily furosemide.

Discussion

Low birth weight is an independent risk factor in the operative management of obstructive TAPVC [5, 6, 9]. Patients who undergo emergent VV stent implantation prior to definitive surgery often present with respiratory distress and cyanosis secondary to pulmonary congestion [4]. In this case, the patient showed no evidence of pulmonary venous obstructive disease, the presentation of which leads to emergent surgery. The patient's respiratory support was more suggestive of pulmonary overcirculation due to the large left-to-right shunt produced by the anomalous pulmonary venous return. Our clinical strategy was to allow the patient to achieve additional somatic growth to mitigate the increased morbidity and mortality observed in low birth weight neonates with this disease.

Over the last decade, surgical outcomes of TAPVC repair have improved with better control of pulmonary hypertension and preoperative clinical stabilization due to more aggressive medical management [6, 9]. Our case presents an opportunity to consider an interventional strategy that palliates the disease to prevent an urgent need for definitive repair. Stent

implantation in the VV provides a bridge that allows further somatic growth and improved pulmonary mechanics from obstruction relief.

Supracardiac TAPVC is a rare congenital cardiac defect in which obstruction of the VV approaches 40% [3]. Elective stent implant in the VV as a palliative procedure may allow additional somatic growth to reduce mortality and morbidity at the time of surgery. Therefore, urgent surgery is avoided, with time allowing for planning and resolution of symptoms prior to definitive repair.

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Conflict of Interest

The authors have no conflict of interest relevant to this publication.

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