

Palliative emergency stenting of the vertical vein in obstructive supracardiac pulmonary venous drainage prior to surgical repair



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Obstructed “total anomalous pulmonary venous connection” mostly presents with respiratory distress secondary to pulmonary congestion. We report two very sick patients who were referred to our cardiac center for intervention to relieve vertical vein (VV) obstruction. Due to hemodynamic instability and associated morbidity, the decision in our combined meeting was to go for interventional cardiac catheterization.

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1. Introduction

Obstructed total anomalous pulmonary venous connection (TAPVC) is an uncommon cyanotic heart lesion that frequently presents early within the first few hours of life with cyanosis and respiratory distress. Obstruction mostly occurs in the infracardiac type but may also occur in the supracardiac type. This obstruction is usually found along the vertical vein (VV) before draining into the innominate vein [1] Obstructed TAPVC frequently requires urgent treatment in

critical patients with significant VV stenosis. Surgical repair is the most common treatment for relieving the obstruction and improving the pulmonary congestion. However, surgery needs cardiopulmonary bypass, which is associated with serious complications, especially in neonates with pre-existing pulmonary edema and compromised pulmonary vascular bed. Moreover, in this critical subgroup of patients, pump-related events like bleeding, intracranial hemorrhage, thromboembolic events, and organ ischemia are poorly tolerated. Therefore, stent implantation could represent an alternative approach to treat an

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obstructed TAPVC with pulmonary congestion. It might be applied in extremely critical conditions as a rescue measure to stabilize the patient by relieving severe hypoxia and reducing pulmonary congestion. Stent implantation is a much less invasive procedure when compared to cardiopulmonary bypass. It may insure an immediate relief of symptoms with the minimum risk possible, which provides more stabilized conditions prior to surgical repair leading to a presumably better prognosis. Nevertheless, it still requires optimal preparation of the patient, sophisticated equipment, and an experienced operator with high and efficient technical skills, especially in unstable patients when the procedure constitutes a rescue [2]. We report regarding two infants in which this approach was implemented. In both cases, the procedure was urgently performed in a short order and bridged to the next surgical repair for TAPVC.

2. Case reports

2.1. Case 1:

A 41-day-old girl, with birth weight of 3 kg, was born full term by normal delivery. She was discharged home in stable conditions. However, after 6 weeks, she started to have progressive signs of respiratory distress. Subsequently, she was admitted to a local maternity and children hospital. Her initial chest X-ray (CXR) in the referring hospital showed left lower lobe lung collapse which improved by chest physiotherapy. The initial echocardiogram established the diagnosis of obstructed TAPVC, with all the pulmonary veins draining into the right superior vena cava (SVC) through a VV with an obstruction at the SVC–VV junction, with a mean gradient of 16 mmHg. Therefore, urgent referral to our cardiac center was arranged. On arrival, she was in moderate respiratory distress which rapidly worsened to respiratory failure. She needed intubation and mechanical ventilation. The CXR showed complete left lung collapse with typical air bronchogram (Fig. 1A). The patient's hemodynamic state worsened and oxygen saturation did not reach 80% even with maximum mechanical support with 100% fraction of inspired oxygen. After discussion with the surgeon, we agreed that the patient was too sick to tolerate surgery. Therefore, the consensus was to proceed with VV stenting. At the catheterization laboratory, the right SVC angiograms showed that all pulmonary veins were draining through the VV into the dilated SVC. The location of obstruction was confirmed at the right

SVC–VV junction (Fig. 1B). A Palmaz Genesis pre-mounted stent (Cordis/Johnson and Johnson; Baar, Switzerland) size 8 × 18 mm was deployed in the area of pulmonary vein obstruction. At the end of the procedure, the angiogram confirmed that the stent was in a good position and the obstruction was relieved (Fig. 1C). The patient was transferred in a stable condition to the pediatric cardiac intensive care unit. After 4 days, the left lung collapse was opened-up (Fig. 1D) and the patient extubated. The patient was discharged home within 1 week, and surgical repair was performed 2 weeks later. She is following-up in our pediatric cardiology clinic till now and she is doing well.

2.2. Case 2:

An 18-hour-old boy, with birth weight of 3.6 kg, was born at the maternity and children hospital via spontaneous vaginal delivery. He was moderately cyanosed, hemodynamically unstable, and hypoactive. His oxygen saturation was 70–80% with mixed respiratory and metabolic acidosis in spite of mechanical ventilation and inotropic support. Urgent chest computed tomography confirmed the presence of obstructed supracardiac TAPVC. The patient was urgently shifted to the cardiac center. Initial echocardiography confirmed the presence of severely obstructed supracardiac TAPVC. The pulmonary veins were forming a confluence which ascended to the right of spine and drained into the right SVC–innominate vein junction, squashed left ventricle, and stretched the patent foramen ovale with a right to left shunt. Initial CXR showed congested both lung fields (Fig. 2A). The case was discussed in our combined team meeting, and we felt that the patient should undergo emergency VV stent to relieve obstruction and then ultimately, should undergo surgery at a later date after stent implantation. At the catheterization laboratory, under general anesthesia, pulmonary venous confluence hand injection angiograms showed that the pulmonary veins were found to form a confluence which ascended to the right of the spine and anteriorly, then made a loop and went posteriorly to join the back of the right SVC (Fig. 2B). Dilatation of the stenotic area prior to stent implantation was performed; therefore, we managed to cross the stenotic area with 5F guiding catheter. Pre-mounted coronary stent (4.5 × 20 mm) was deployed in the right position. A repeated angiogram after the procedure into the left upper pulmonary vein through the vein of Marshal (left VV) showed the stent in a good position with no obstruction (Fig. 2C). The patient

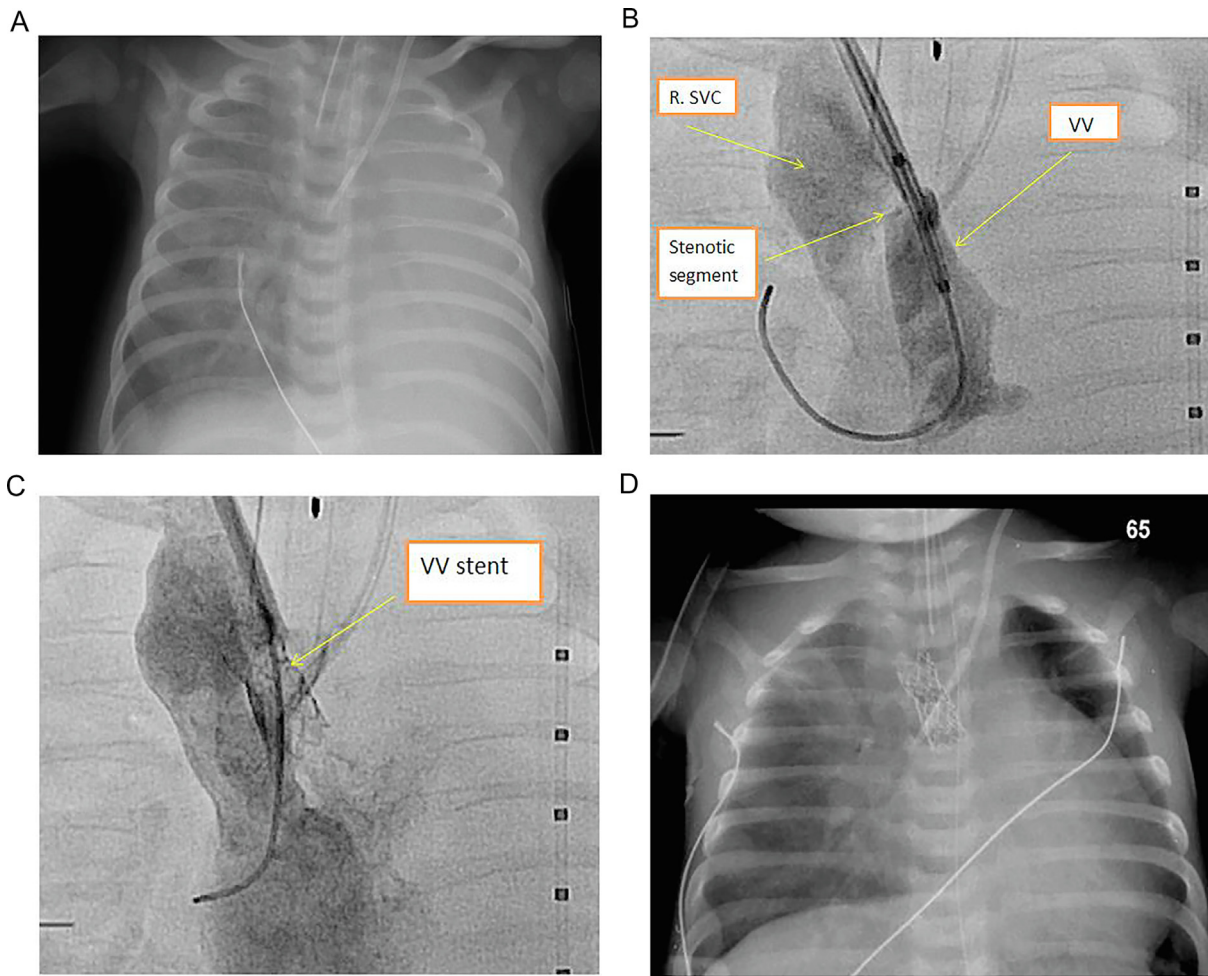


Figure 1. (A) Chest radiography shows total left lung collapse; (B) anteroposterior view, right superior vena cava (SVC) angiogram shows SVC-vertical vein VV junction obstruction; (C) lateral view, right SVC angiogram shows patent SVC-VV stent with good flow; and (D) chest radiography shows resolved left lung collapse, VV stent. SVC = superior vena cava; VV = vertical vein.

showed significant improvement in blood pressure, saturation (from 40% to 95% on oxygen), and blood gas. Lung congestion also improved (Fig. 2D). The patient was extubated and discharged from hospital. Surgical repair is planned in the near future.

3. Discussion

TAPVC is a rare cardiac defect that comprises 1.5% of all congenital heart disease cases. Since the first surgical attempt to correct TAPVC was reported in 1951, improved surgical techniques and hospital care have led to significantly better outcomes of TAPVC surgery in children [3]. However, a review of the literature suggests that neonates represent the most severe end of the TAPVC spectrum and may have higher rates of death. Risk factors for mortality include preoperative clinical status, prolonged bypass time, persist-

ing micro-obstruction, and low operative weight. Reduction in the mortality rate will likely require development of effective medical management for patients who have peripheral pulmonary vein obstruction not amenable to surgical repair [3]. 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. Supracardiac TAPVC is the most common type, comprising about 45% of cases. A left-sided VV accounts for 70% of the connections between the pulmonary confluence and the right atrium, and stenosis occurs in approximately 40% of cases [4]. VV obstruction represents one of the most malicious conditions in the newborn causing severe respiratory distress refractory to conservative urgent measures. Severe pulmonary congestion due to pulmonary venous obstruction, in particular, leads to a critical condition, resulting in poor

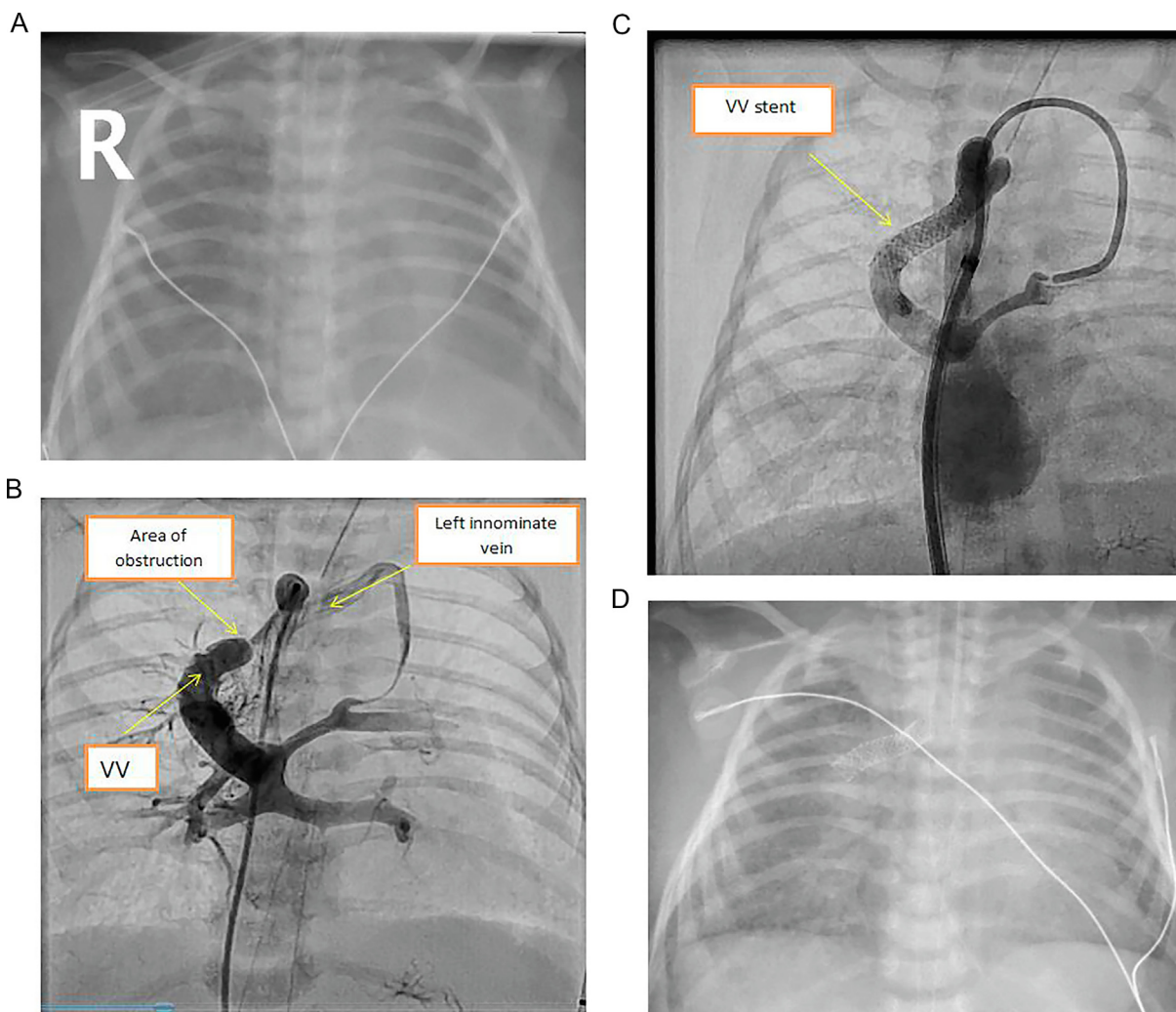


Figure 2. (A) Chest radiography shows congested lung; (B) anteroposterior view, left innominate vein angiogram shows superior vena cava (SVC)–vertical vein (VV) junction obstruction; (C) anteroposterior angiogram view shows VV stent; and (D) chest radiography shows better lung aeration, VV stent. SVC = superior vena cava; VV = vertical vein.

prognosis if obstruction is unrelieved. Urgent intervention is vital in TAPVC with pulmonary venous obstruction to reduce pulmonary congestion and improve the critical condition, while elective surgical intervention is applicable in TAPVC without pulmonary venous obstruction [2]. Palliative stent implant in the VV and relieving the obstruction has many advantages. It could be a life-saving measure and may allow some physical growth of the newborn, thereby reducing mortality and morbidity at the time of surgery, and could represent a bridge to a management decision. Therefore, urgent surgery is avoided, with time allowing for planning and resolution of symptoms prior to definitive repair. Kilgore et al [5] reported stent implantation into the VV of a 6-week-old infant in a critical condition, followed by surgical correction after 1 week. Lo-A-Njoe et al [6]

described a critical neonate with TAPVC who received a coronary stent implantation to prepare before surgery. Kyser et al [7] presented a stent implantation under extracorporeal membrane oxygenation in a newborn with TAPVC to stabilize a lung condition [2]. All the aforementioned cases were critical, and all survived. This suggests that this procedure can be an acceptable alternative choice for rescue in the treatment of TAPVC with severe pulmonary venous stenosis if the patient's condition is critical.

4. Conclusion

Stent implantation as a palliative procedure, performed in a few cases, could represent a safe, reproducible strategy for babies with obstructive TAPVC. The aim is to change the pathophysiology

of a sick patient to become more stable and to change the emergency surgical repair to semi-elective. Catheter intervention may be considered as part of the preoperative cardiovascular stabilization strategy for high-risk infants with obstructed supracardiac TAPVC to achieve more stable intensive care unit course.

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