CASE REPORT

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Obstructed infracardiac total anomalous pulmonary venous connection: The challenge of palliative stenting for the stenotic vertical vein

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Received: 31 March, 2020
Accepted: 18 May, 2020

ABSTRACT

Introduction: Obstructed total anomalous pulmonary venous connection (TAPVC) is one of the commonest seen emergencies in pediatric cardiology centers.

Case presentation: Our case was diagnosed to have this anomaly, showing early respiratory distress resulting from severe pulmonary congestion. Palliative stenting of the obstruction was done, which helped in stabilizing the debilitated hemodynamics of the baby before surgery, thus a good surgical outcome and prognosis are expected.

Conclusion: This intervention may be listed as a vital measurement in the preoperative cardiac stabilization plan for infants with obstructed TAPVC.

KEYWORDS
Obstruction, Total anomalous pulmonary venous connection (TAPVC), Palliative stent

INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a rare developmental cardiac anomaly representing 1%–2%.[1] The pulmonary veins drain the oxygenated blood into the right atrium either directly or indirectly through systemic veins. A right-to-left shunt is mandatory for survival. Nearly in 15% of cases, an obstruction of the pulmonary venous return is present.[2] Complete delineation of the drainage path of the TAPVC and the early management of the obstruction is essential for survival before planning to surgical correction.

Our case is one of these emergency cases that presented early within the first few hours of life with severe respiratory distress and cyanosis. The diagnosis consistent with infracardiac TAPVC, with a stenotic lower segment of the descending vertical vein (VV) draining into inferior vena cava (IVC), which could explain the patient's debilitation.

CASE REPORT

A 24-hour-old full-term male neonate, with birth weight 3.5 kg, was born in a tertiary pediatric hospital. The baby presented with severe respiratory distress, cyanosis, and profound metabolic acidosis despite mechanical ventilation. A systolic murmur was noticed at left upper sternal border. Chest x-ray revealed dextrocardia, and a diffuse haziness pattern indicating severe pulmonary venous congestion. Urgent preliminary two-dimensional echocardiography showed mirror-image dextrocardia, with atrioventricular concordance and double outlet right ventricle.

DOI: 10.1002/ped4.12204

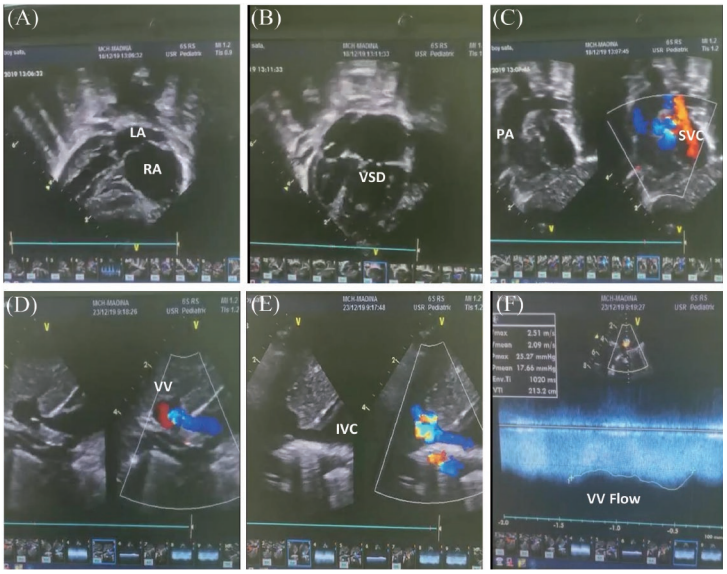

FIGURE 1 Echocardiography imaging of the neonate with obstructed infracardiac type total anomalous pulmonary venous connection (TAPVC) revealed (A) Mirror-image dextrocardia, dilated RA, ASD, and small-sized LA with no attached pulmonary veins. (B) Large inlet to outlet VSD. (C) Bilateral SVC attached to RA, and single VV going downward behind the heart. (D) VV tracked down to the IVC. (E) Turbulence at site of entry of VV into IVC. (F) Doppler of VV flow. RA, right atrium; LA, left atrium; ASD, atrial septal defect; VSD, ventricular septal defect; SVC, superior vena cava; IVC, inferior vena cava; VV, vertical vein.

Average sized atrial septal defect (measured 7 mm) with non-restrictive right-to-left shunt is present along with a large inlet to outlet ventricular septal defect. Bilateral superior vena cava (SVC) draining into left-sided atrium (morphologic right atrium). The picture of an obstructed infracardiac type TAPVC was suspected while observing all the pulmonary veins attaching separately a confluence behind the heart. Inotropic support was advised (Figure 1).

On the second day, the diagnosis was confirmed with computed tomography (CT) which revealed situs inversus totalis, infracardiac type TAPVC with a VV going downward with an infra-diaphragmatic path and a stenotic lowest segment of it draining into IVC (Figure 2). CT angiography videos were shown in the supporting information.

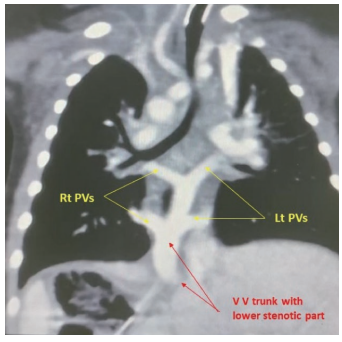

FIGURE 2 Computed tomography angiography revealing infracardiac type total anomalous pulmonary venous connection (TAPVC) with a vertical vein going downward (with a lower stenotic segment), and four pulmonary veins joining it separately. Lt PVs, left pulmonary veins; Rt PVs, right pulmonary veins; VV, vertical vein.

The hemodynamic status of the baby worsened and the oxygen saturation failed to exceed 75% even on the maximal settings of the ventilation support. Urgent transfer of the case to the locality cardiac center was done. After exhaustive discussion with the pediatric cardiac surgeon, a verified decision was taken to stent the VV as the baby was too sick to tolerate surgery. This hoped to relieve the obstruction and consequently to be weaned safely from mechanical ventilation. Later, the baby would undergo surgery after stabilization.

Under general anesthesia, at the catheterization laboratory, a pulmonary artery angiograms with levo-phase revealed that all pulmonary veins were opening separately into one confluent, descending posteriorly as VV to join the IVC with a severe stenotic lowest segment (Figure 3).

Using sterling balloon (5 mm × 40 mm) passing from the IVC; dilation of the lowest stenotic area of the VV prior to stent implantation was done (Figure 4). Then, 6F guiding catheter crossed the obstructed part of the VV allowing implantation of Sirry Omnilink stent (10 mm × 29 mm) (Figure 5). A repeated hand-injection angiogram into the confluent after the procedure showed a good positioned patent stent with no IVC obstruction, and good flow to right atrium (Figure 6).

After the procedure, the baby admitted to the pediatric cardiac intensive care unit. A low dose of diuretics advised after changing the pathophysiology from obstructive to non-obstructive. Hours later, the baby general condition showed significant improvement (his oxygen saturation reached 93% on 100% oxygen), and he was rapidly

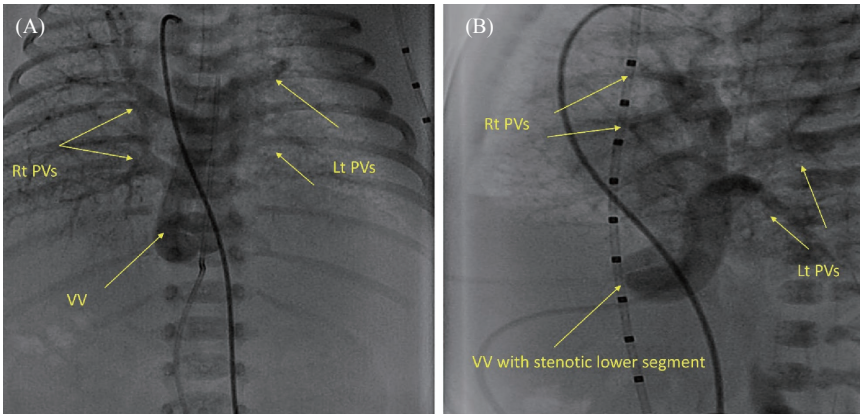

FIGURE 3 Pulmonary artery angiograms with levo-phase showing vertical vein joining the inferior vena cava with a stenotic lowest segment in the neonate with total anomalous pulmonary venous connection. (A) Anteroposterior view. (B) Lateral view. Lt PVs, left pulmonary veins; Rt PVs, right pulmonary veins; VV, vertical vein.

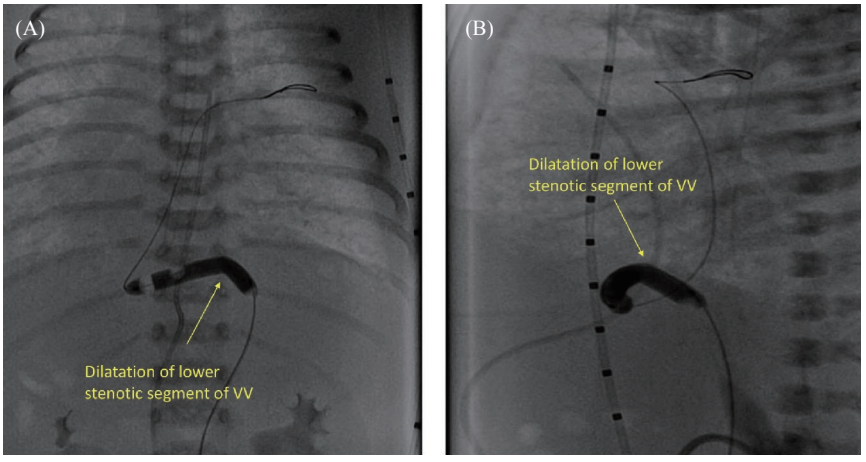

FIGURE 4 Dilation of the lower stenotic segment of the vertical vein prior to stent implantation in the neonate with total anomalous pulmonary venous connection. (A) Anteroposterior view. (B) Lateral view. VV, vertical vein.

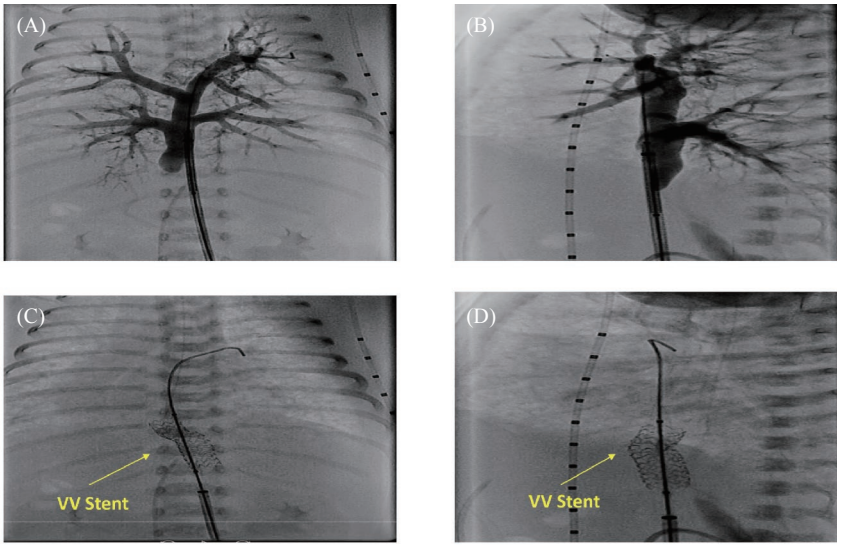

FIGURE 5 Vertical vein angiogram of the neonate with total anomalous pulmonary venous connection. (A) Anteroposterior view. (B) Lateral view. Implantation of vertical vein stent. (C) Anteroposterior view. (D) Lateral view. VV, vertical vein.

extubated. Two days later, the baby's condition was stable and a pulmonary artery banding was performed. Thereafter, at age of 2 months, an elective total surgical repair done successfully and baby now is stable at home.

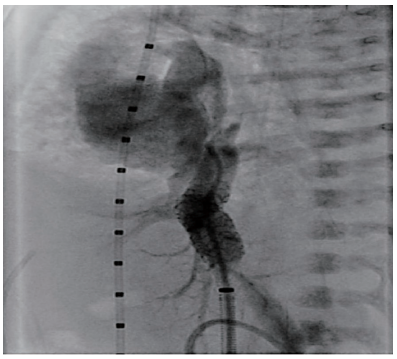

FIGURE 6 Confluent angiogram with a good positioned patent stent with no inferior vena cava obstruction, and good flow to right atrium in the neonate with total anomalous pulmonary venous connection.

DISCUSSION

TAPVC is a common differential diagnosis of a newborn presenting with respiratory distress and cyanosis. This is classified into four anatomic types according to the site of pulmonary venous drainage. An obstruction to the pulmonary veins may occur with any type, with the highest incidence encountered in the infracardiac type.[3]

Severe pulmonary congestion secondary to obstructed VV drainage, in particular, leads to a critical respiratory distress condition refractory to conservative medical management.[4] The preoperative clinical status, failure to wean from ventilation, persisting micro-obstruction, and low operative weight are all risk factors for poor surgical outcome.[5] On the other hand, the elective surgical intervention in TAPVC without pulmonary venous obstruction has a good prognosis. Therefore, urgent intervention is vital to alleviate the pulmonary congestion, relieve the critical condition and improve the patient prognosis.[6,7]

Relieving the stenotic VV obstruction by a palliative stent implantation could be a life-saving measure, and a rescue treatment for the worsening patient's condition. The stent can represent a bridge to a definitive safe repair by allowing some physical growth, improving the critical condition of the baby, and thereby reducing mortality and morbidity of an urgent non-avoidable surgery.[7,8]

In conclusion, although not widely performed, palliative stent implantation procedure could represent a safe, emerging strategy for neonates presenting with any type of obstructed TAPVC.[7,8] The goal is to reverse the pathophysiology of a debilitated neonate to become more stable and to turn the emergency surgical repair to semi-elective one. This intervention may be listed as a vital measurement in the preoperative cardiac stabilization plan for infants with obstructed infracardiac TAPVC to achieve more stable course.

CONSENT FOR PUBLICATION

Informed consent was taken from the participant's parents.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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SUPPORTING INFORMATION

Additional Supporting Information may be found online in the supporting information tab for this article.

How to cite this article: Al-Mutairi M, Aselan A, Al-Muhaya M, Abo-Haded H. Obstructed infracardiac total anomalous pulmonary venous connection: The challenge of palliative stenting for the stenotic vertical vein. Pediatr Invest. 2020;4:141-144. https://doi.org/10.1002/ped4.12204