BAILOUT PROCEDURE IN OBSTRUCTED SUPRACARDIAC TOTAL ANOMALOUS PULMONARY VEIN DRAINAGE

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Abstract Total anomalous pulmonary venous drainage is a rare and diverse anomaly, accounting for 1% to 3% of patients with congenital heart disease. Newborns with diagnosis of an obstructed total anomalous pulmonary venous drainage are extremely ill soon after birth and often present with severe cyanosis, pulmonary hypertension and low cardiac output requiring urgent surgical intervention. Transcatheter palliative stenting of the obstructive vertical vein can be an acceptable alternative as a bailout intervention before complete surgical correction is undertaken. This report of two cases highlights the feasibility, safety and effectiveness of the interventional palliative procedure and confirms that this technique can be an acceptable and attractive bridge in the algorithm of medical decisions during the evaluation of these critical patients.

Key words: obstructed total anomalous pulmonary vein drainage, supracardiac, vertical vein obstruction, stent, bailout procedure

Resumen Procedimiento de rescate en drenaje venoso pulmonar anómalo total supracardíaco obstructivo. El drenaje venoso pulmonar anómalo total es una enfermedad poco frecuente y de presentación diversa y se observa en el 1% a 3% de las cardiopatías congénitas. Si se asocia a obstrucción, se convierte en una afección grave en el recién nacido, mostrando cianosis intensa, hipertensión arterial pulmonar y bajo gasto cardíaco con indicación de intervención quirúrgica de urgencia. El implante de stent por cateterismo de forma paliativa para aliviar la obstrucción puede ser una alternativa aceptable de tratamiento como intervención de rescate antes de la corrección quirúrgica definitiva. Presentamos dos casos de intervención percutánea paliativa mostrando que esta técnica puede ser eficaz como puente al tratamiento quirúrgico definitivo para ser incorporado en la toma de decisiones de estos pacientes críticos.

Palabras clave: drenaje venoso pulmonar total obstruido, supracardíaco, vena vertical obstruida, stent, procedimiento de rescate

Total anomalous pulmonary venous drainage (TAPVD) is a rare and diverse anomaly, accounting for 1% to 3% of patients with congenital heart disease1. It is characterized by failure of the pulmonary venous confluence to be absorbed into the dorsal portion of the left atrium in combination with a persistent splanchnic connection to the systemic venous systems. Improvement in diagnostic accuracy, advances in surgical techniques, and changes in perioperative management have led to a significant decrease in mortality2. Nevertheless, several factors such as neonatal surgical repair, poor preoperative condition, pulmonary venous obstruction type, mixed anatomic variation, single-ventricle physiology and heterotaxy syndromes have been identified as important risk factors for postoperative decreased survival1-5.

Although stenting of the vertical vein in obstructed TAPVD is not a standard of care procedure, few case reports have shown adequate outcome as a preoperative “bridge intervention”6-9.

Clinical case 1

A full term 1-day-old girl, with a birth weight of 3.7 kg, developed progressive respiratory distress, cyanosis, gasping, grunting, metabolic acidosis, decreased urine output and bradycardia. After endotracheal intubation and mechanical ventilation with 100% oxygen, intravenous administration of dopamine and epinephrine were started. A chest X-ray showed severe pulmonary venous congestion. A transthoracic color-Doppler echocardiogram demonstrated suprachardiad...
TAPVD with severe obstruction of the left vertical vein at the level of the left pulmonary artery. A moderate size, mildly restrictive, ostium secundum type atrial septal defect was also identified. The ductus arteriosus was patent and pulmonary artery pressure was estimated at systemic levels. The arterial oxygen saturation was persistently low in the range of 65 to 70% and the pH was 6.98. Due to progressive clinical status deterioration, we decided both to stent the obstructed vertical vein and to perform an atrial balloon septostomy to stabilize the patient prior to a corrective surgery.

The right femoral vein was cannulated and a 5-F short sheath was placed. A 4-F Judkins right catheter (Cordis Corporation, FL, USA) was advanced via the inferior vena cava, right atrium, superior vena cava, innominate vein into the vertical vein. A vertical vein angiogram showed a severe stenosis at the “anatomic vice” region. The caliber of the vessel in this site was 1.4 mm while the adjacent segment of the vertical vein was 3.8 mm wide. Four dilated pulmonary veins were identified draining into a confluence and from there, through the vertical vein, directly drained superiorly into the innominate vein (Fig. 1A).

A Rebel® coronary stent size 3.5 mm × 20 mm (Boston Scientific, USA) was positioned in the stenotic area and inflated at 10 ATM. Due to a mild kinking visualized in the central portion of the stent configuration, a postdilation was performed using a coronary balloon 4.0 mm × 20 mm inflated at 16 ATM achieving a homogeneous caliber of the stent. After final dilatation, an angiogram showed a significant improvement of the stenotic segment of the vertical vein with abolition of gradient through the lesion (Fig. 1B). Subsequently, an atrial balloon septostomy was carried out with a standard technique using a Z-5® atrioseptostomy catheter (Numed, Canada) with an adequate relief of gradient through the interatrial septum.

After the intervention, the patient showed a slowly but steady improvement of her clinical condition. At 20 days of life underwent a corrective surgery with optimal outcome. The vertical vein was ligated, the stent was left in situ and the pulmonary veins were reconnected through the confluence to the posterior wall of the left atrium. A small atrial septal defect (4 mm in diameter) was intentionally created in the atrial septum after patch closure. She was discharged after 15 days and since then remains asymptomatic growing appropriately. Her last bidimensional (2D) and color-Doppler transthoracic echocardiogram showed no obstruction at the level of the pulmonary veins drainage into the left atrium and the pulmonary artery pressure was estimated within the normal range.

Clinical case 2

A 3 days-old male infant, weighing of 3.2 kg, born after an uncomplicated pregnancy was admitted to the Neonatology Department due to impending circulatory collapse 12 h previously. The patient had been discharge two days after been delivered from a regional hospital and developed respiratory distress soon after arriving home. He experienced feeding intolerance with excessive sweating, irritability, poor color, progressive tachypnea and cyanosis. His peripheral saturation was in the low 80’s and his urine output was decreased. A transthoracic color-Doppler echocardiogram confirmed the diagnosis of obstructed TAPVD (supracardiac type) with severe obstruction located at the anatomic vice site. Pulmonary artery pressure was estimated at supra systemic level and a large ostium secundum type atrial septal defect was also observed. Due to clinical instability, we judge high surgical risk and decided palliative endovascular stenting of the obstructed vertical vein prior to a corrective surgery. The narrowest point of the vertical vein at the anatomic vice location was 3.6 mm with the adjacent vessel measuring 7.2 mm (Fig. 2A). With the aid of a Judkins right coronary artery catheter, a 0.014” extra support coronary wire (Boston Scientific, USA) was parked distally into the right lower pulmonary vein. A Express vascular® peripheral stent SD 7.0 mm × 18 mm (Boston Scientific, USA) was implanted with the aid of a insufflator reaching 14

Fig. 1.– Patient one. Selective pulmonary vein angiogram in anteroposterior projection. Four dilated pulmonary veins are visualized with severe obstruction of the collector at the anatomic vice segment with improvement of caliber distally, draining into the innominate vein (A). Similar angiogram showing the improvement of caliber of the narrowest portion of the collector after stent implantation (B)
ATM. Post intervention angiogram showed a significant improvement of the stenotic segment, elimination of the gradient (Fig. 2B) and the patient showed a prompt recovery being extubated two days later. One month after the interventional palliative procedure, he underwent complete surgical correction that included sutureless technique anastomosis of the collector to the posterior wall of the left atrium and vertical vein ligation. The stent was left in situ and the atrial septal defect was partially closed. His last follow up visit informed a well-nourished infant, with normal growth and development. His pulmonary artery pressure estimated by transthoracic 2D and Doppler color echocardiography was within normal limits. No stenosis was observed in the pulmonary veins.

Discussion

Supracardiac TAPVD is the most common type of this anomaly representing about 42% of the cases. A left-sided vertical vein accounts for 70% of the connections and obstruction occurs in approximately 40% of patients; being the infracardiac type the most common group associated with obstruction. The vertical vein can be obstructed as it passes through a narrow space formed by the ipsilateral pulmonary artery anteriorly, ligamentum or patent ductus arteriosus medially and the ipsilateral main bronchus and descending aorta posteriorly, forming what is called the “anatomic vice”.

A diagnosis of preoperative obstructed TAPVD can be made by a combined evaluation of clinical status, oxygen saturation and echocardiography data, which shows a non-phasic flow velocity > 1.8 m/s in an individual vein or in the obstructed vertical vein itself. Morphological features of preoperative TAPVD can also be visualized by contrast enhanced MRI, cardiac CT, angiography and finally with a detailed intraoperative assessment. The variety of malformations causing obstruction includes intrinsic stenosis with pulmonary vein ostial stenosis, pulmonary vein hypoplasia, extrinsic compression with obstruction within the anomalous connecting vein course or at its connection to the systemic circulation or a restrictive atrial septal defect level.

Early corrective surgery is the gold standard to manage obstructed supracardiac TAPVD. However, when corrective surgery is deemed high-risk or not possible or unavailable, palliative endovascular stenting of the obstructed vertical vein is an option as a bridge to surgical correction. These patients have poor prognosis due to severe pulmonary congestion secondary to vertical vein obstruction. Therefore, urgent intervention is crucial to relieve obstruction.

According to our experience, at the time of the surgical procedure and after the vein confluence was opened, the proximal end of the stent was visualized in both cases and appeared completely endotelized so they were left in situ. Ligation of the stented vertical vein was uneventful. Stent removal was considered unnecessary and dangerous due potential vein laceration. No additional surgical difficulty was noticed correcting this congenital heart anomaly after stent implantation.

Most frequently, the vertical vein obstruction is related to extrinsic compression so we did not consider just balloon dilatation of the lesion due to the recoil phenomenon that most probable might occur in this situation. Since we choose this procedure as a bridge to surgical repair, a bare
metal stent was implanted knowing that re-stenosis in the following short time-period would not be a problem. The size of the stent implanted was selected according to the adjacent diameter of the vertical vein in a relation 1 to 1 to improve both the caliber and the gradient through the vessel. Our ultimate goal with this approach was to avoid urgent corrective surgery, allowing time for planning and improvement prior to complete surgical repair.

In summary, transcatheter palliative stenting of the vertical vein can be an acceptable alternative as a bailout intervention before complete surgical correction is performed in critical newborns presenting with an obstructed TAPVD. This report highlights the feasibility, safety and effectiveness of the interventional palliative procedure and confirms that this technique can be an acceptable bridge in the algorithm of medical decisions during the evaluation of these critical patients.

**Conflict of interest:** None to declare

**References**


