

CASE REPORT

Emergency stenting of vertical vein in a neonate with obstructed supracardiac total anomalous pulmonary venous drainage

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SUMMARY

A late preterm newborn baby presented with respiratory distress and increasing cyanosis within 2 hours of birth. Bedside transthoracic echocardiography showed a critically obstructed vertical vein in a supracardiac total anomalous pulmonary venous drainage (TAPVD). Emergency stenting of the vertical vein was successfully performed at 24 hours of life.

KEY WORDS: Obstructed Total Anomalous Pulmonary Venous Drainage (TAPVD), Infracardiac, Supracardiac, Vertical Vein, Emergency Stenting, respiratory distress, cyanosis, Primary Pulmonary Hypertension of Newborn (PPHN)

INTRODUCTION

Premature babies may present with some degree of respiratory distress. A few conditions like transient tachypnea of newborn (TTN), hyaline membrane disease (HMD), primary pulmonary hypertension of newborn (PPHN), meconium aspiration syndrome (MAS) or pneumothorax can present in this way. However, conditions related to congenital heart defects that causes respiratory distress in a newborn neonate include obstructed total anomalous pulmonary venous drainage (TAPVD) or systemic outflow obstruction.

All TAPVD hearts are dilated. Both the right atrium and ventricle are hypertrophied with dilatation of the pulmonary artery, a normal-sized left ventricle, and reduced left atrial size and volume. There is usually a common chamber that is directly behind the left atrium that is formed by the confluence of the pulmonary veins. This pulmonary venous confluence is connected to the systemic venous system via an anomalous channel called a vertical vein. Darling¹ proposed a classification which is widely followed and consists of four types: supracardiac (55%), cardiac (30%), infracardiac (13%), and mixed (2-5%). TAPVD can further be classified by the presence or absence of obstruction.

Obstructed total anomalous pulmonary venous drainage (TAPVD) is an uncommon cyanotic heart lesion that frequently present early within first few hours of life with cyanosis and respiratory distress. Obstruction occurs more commonly in the infracardiac type but do also occur in the supracardiac type. This obstruction is usually found along the vertical vein before draining into the innominate vein.

Obstructed TAPVD causes increased pulmonary venous pressure. This usually presents with pulmonary oedema in neonates. As a compensatory mechanism, there will be increase in pulmonary lymphatic flow, recruitment of

pulmonary-bronchial venous anastomoses, and reflex pulmonary vasoconstriction. These will cause an increase in pulmonary artery and right ventricular pressures. As the right ventricular compliance decreases there will be more shunting across the atrial septal defect from right to left, causing severe cyanosis and hypoxaemia. This leads to acidosis, shock, and multiorgan dysfunction. In effect a neonate with obstructed TAPVD will present with pulmonary edema, pulmonary hypertension, reduced pulmonary blood flow, and severe cyanosis. In patients with severe hypoxaemia with respiratory failure and not responsive to initial medical therapy, urgent surgical repair is the only effective treatment.

Accurate recognition and clinical diagnosis of obstructed TAPVD is challenging if the clinician is unaware of this cardiac condition. Definitive management would be immediate surgical correction. However stenting of the vertical vein in obstructed TAPVD is not a well-established procedure; but anecdotal case reports^{2,3,4} have shown satisfactory outcome.

We report a case of successful pre-operative stabilization via transcatheter stenting of the obstructed vertical vein in a neonate with supracardiac TAPVD in our hospital where there is no on-site surgical service

CASE REPORT

EM was born in Sarawak General Hospital at 36 weeks gestation with a birth weight of 2.7 kg. He was treated as late preterm with presumed sepsis and was started on first line antibiotics. He was intubated soon after birth because of increasing respiratory distress and cyanosis. Despite high ventilatory setting, he remained severely hypoxaemic and acidotic. Bedside echocardiogram (Fig. 1) revealed the diagnosis of obstructed supracardiac TAPVD. The pulmonary veins formed a confluence behind the left atrium. This confluence then drained superiorly into the innominate vein via a vertical vein. Turbulence was seen on colour Doppler within the vertical vein with peak Doppler gradient of 25 mmHg.

As the baby was clinically unstable and logistically difficult to transfer him to a tertiary paediatric cardiac surgical center, transcatheter stenting of the obstructed vertical vein was attempted.

PROCEDURE

The right femoral vein was cannulated. Right heart catheterization was performed with 4F Judkins right coronary catheter. The vertical vein was entered retrogradely from the innominate vein. Contrast injection revealed a discrete stenosis

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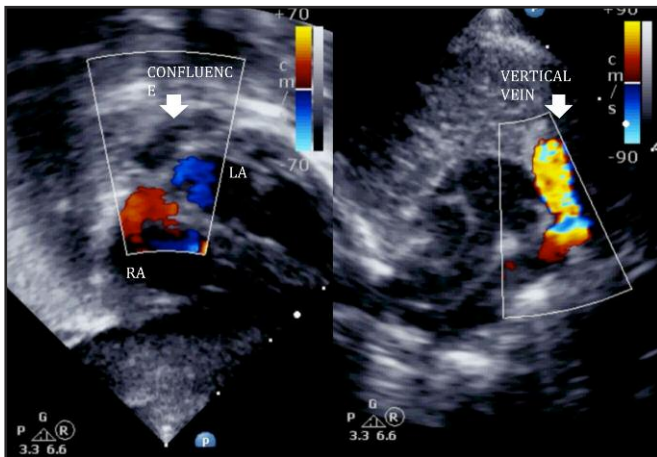


Fig. 1: Confluence seen forming behind the Left Atrium and Vertical Vein flowing superiorly seen.

at the mid-segment of the vertical vein (Fig. 2a) between the pulmonary venous confluence and the innominate vein entrance. Pulmonary artery pressure was suprasystemic.

The catheter was then exchanged to 6F Judkins right coronary guiding catheter. A 0.014" BMW PTCA guidewire (Abbott Vascular, USA) was used to cross the obstruction at the vertical vein. Balloon angioplasty of the vertical vein stenosis was first performed with Tyshak Mini balloon 5 mm (NuMed, USA) but the waist could not be abolished at maximal inflation pressure. Two overlapping Omega Platinum Chromium 4.5 mm x 12 mm bare metal coronary stents (Boston Scientific, USA) were deployed consecutively to cover the obstructed segment. Contrast injection post stenting (Fig. 2b) showed good flow of contrast across stents. Balloon atrial septostomy was performed as an adjunctive measure. There was minimal blood loss and the vitals improved dramatically post vertical vein stenting.

RESULTS

Post-stenting angiography revealed no residual obstruction. There was smooth flow of contrast draining from the pulmonary veins into the innominate vein. Post-procedure, the infant improved dramatically, both haemodynamically and in oxygenation status. His inotropic support was reduced gradually over 3 days and was successfully extubated 3 days post procedure. He was transferred to a tertiary paediatric cardiac surgical center for surgery.

DISCUSSION

For TAPVD patients, medical management is only supportive with the aim of stabilising the patient before surgery. This includes correcting acidosis, hypotension, judicious fluid management, and mechanical ventilation. Prostaglandin is occasionally helpful in keeping ductus venosus open in infra-diaphragmatic TAPVD, but in other forms it is harmful. Surgery for TAPVD is aimed at achieving unobstructed pulmonary venous drainage to the left atrium. The clinical condition of the patient will usually decide the timing of surgery.

In general, the diagnosis of TAPVD of any type is an indication to proceed with early surgery. An emergency operation is required in neonates with obstructed forms of TAPVD. However

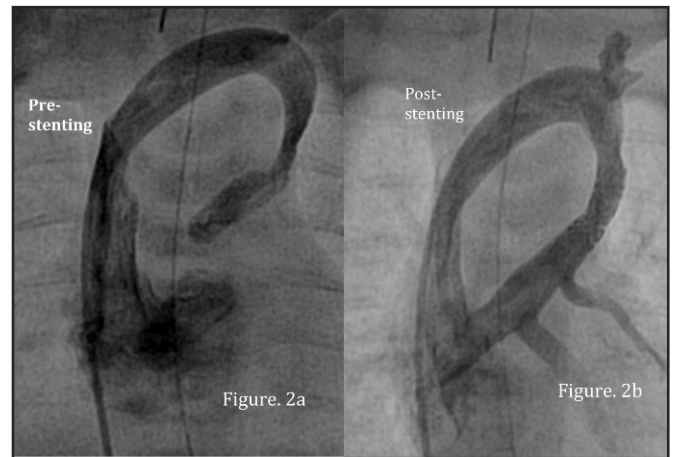


Fig. 2: Pre and post stenting of the vertical vein.

this was not feasible as the nearest paediatric cardiothoracic centre that could handle this condition was logistically inaccessible for our patient.

Thus the only option available would be a transcatheter stenting of the obstructed vertical vein followed by arrangements later to transport the patient to the definitive paediatric cardiothoracic centre. This condition warrants an urgent intervention by paediatric cardiologist who already has appropriate training for this procedure.

A search via PubMed revealed few case reports that have shown improvement in clinical conditions following stenting of the obstructed TAPVD^{2,3,4}. This report highlights the feasibility and the effectiveness of transcatheter stenting of the vertical vein for obstructed supracardiac TAPVD as an emergency measure to stabilize a sick neonate. This procedure can serve as a bridge to subsequent surgical repair that can be performed under a more stable and controlled condition.

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