CASE REPORT

Transcatheter occlusion of vertical vein that opened up after surgical repair of obstructed infracardiac total anomalous pulmonary venous connection

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INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is the most important anomaly of the pulmonary veins in which there is no direct connection between any pulmonary vein and the left atrium. Infracardiac TAPVC usually presents as a surgical emergency due to its inherent immediate risk of obstruction to the vertical vein (VV). However risk is inevitable when the vertical vein drains into ductus venosus or portal vein, however, if it drains into inferior vena cava (IVC) or right hepatic vein (HV), the chances are lesser. Surgical correction of infracardiac TAPVC with sutureless technique has shown better outcome than conventional technique in international setting[1,2]. The sutureless technique involves creating a new left atrium (neoatrium) by suturing the edge of the left atrial wall to the pericardium around the pulmonary veins, without any direct suturing of the pulmonary veins and the neo-left atrium thus formed is contained by the posterior pericardium and mediastinal soft tissue. As the descending vertical vein in the infracardiac TAPVC is usually obstructed, and to minimize the dissection, the vertical vein can be left alone. The intact vertical vein may help to decompress the small left atrium. However, on some occasions where vertical vein drains into HV or IVC, the unligated vertical vein reopens resulting in significant left to right shunt. We report a neonate whose unligated vertical vein reopened in the post operative period with significant haemodynamic instability and later required transcatheter occlusion with embolization coils.

Keywords: Infracardiac TAPVC, sutureless technique, descending vertical vein, transcatheter occlusion

CASE HISTORY

A one-week-old newborn underwent emergency surgical correction of partially obstructed infracardiac TAPVC and was recovering in the cardiothoracic intensive care unit. The surgery was performed with the sutureless technique, and the vertical vein was left open because it was significantly obstructed at the entry point into right hepatic vein (RHV). Initially, she showed a good progression but, from post op day 3 onwards she deteriorated into a low cardiac output status and echocardiographic examination revealed a significant left to right shunt across the vertical vein. On the post op day 5, as the baby was critically ill to undergo open surgery, it was decided by the multidisciplinary team to go ahead with minimally invasive percutaneous occlusion (Figure 1).

PROCEDURE DETAILS

The procedure was performed under general anaesthesia after obtaining informed written consent. Left femoral venous access was obtained. A 4-Fr RCA catheter was passed into the main pulmonary artery (MPA) and the pressure was recorded as 44/21 (29) mmHg. MPA angiogram showed small branch pulmonary arteries with good bilateral lung arborization. The levo phase showed unobstructed pulmonary venous return to the left atrium via the common chamber. However, there was a significant egress of contrast into the dilated vertical vein that drained into the right RHV and then into the right atrium (Figure 1).

The vertical vein was entered using a 0.038/150cm terumo exchange length guidewire and a 4-Fr RCA catheter. Two 0.038" x 5cm x 5mm Cooks embolization coils were deployed using bare-release technique at the entry point of vertical vein to RHV. Post occlusion angiogram showed near-complete occlusion of the vertical vein proximal to its drainage point

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into the RHV (Figure 2). Post occlusion, there was no significant rise in the pulmonary artery pressure. The infant was successfully extubated 2 days later and discharged home 4 days after the coil embolization.

The infant has undergone regular follow-up visits for the past year following the procedure and has exhibited satisfactory growth and development.

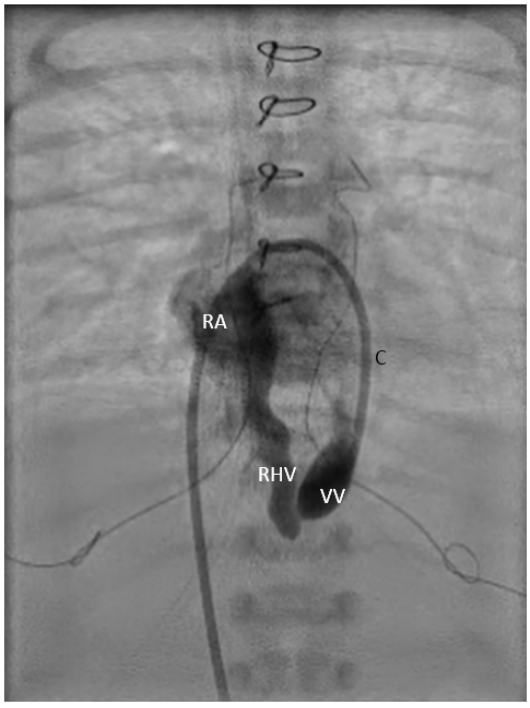

Figure 1. Angiogram showing the patient's descending vertical vein draining into RHV which in turn drains into right atrium causing significant left to right shunt. The catheter has been passed through LFV, IVC, RA, LA, common chamber, into VV. LFV- left femoral vein, IVC- inferior vena cava, RA- right atrium, LA- left atrium, VV- vertical vein, C- RCA catheter, RHV- right hepatic vein.

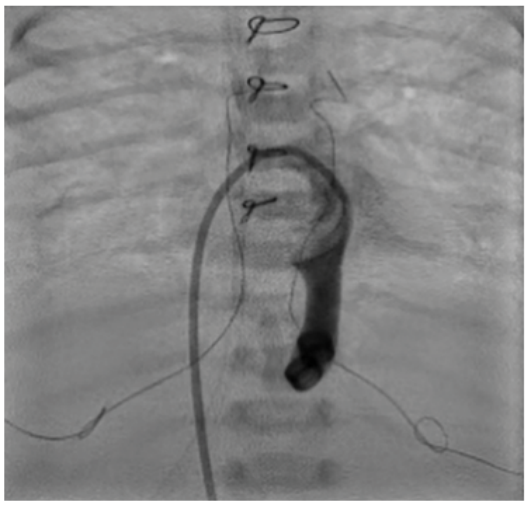

Figure 2. After coil embolization of the descending vertical vein. Note the complete occlusion of the vertical vein.

DISCUSSION

TAPVC is the most important anomaly of the pulmonary veins in which there is no direct connection between any pulmonary vein and the left atrium. All the pulmonary veins connect to the right atrium or one of its tributaries, mixing oxygenated and deoxygenated blood, reducing oxygen delivery to the body. TAPVC accounts for 1-1.5% of congenital heart diseases and is divided into four main types. Infracardiac TAPVC is usually associated with obstruction of the vertical vein leading to a surgical emergency. International as well as local data at our institution suggest that surgical outcome following sutureless technique for the correction of supracardiac and infracardiac variants is superior compared to conventional repair[1,2].

Traditionally, the vertical vein is ligated at the time of surgical repair to avoid any significant left to right shunt post-operatively. Conversely, there is evidence to suggest that an unligated vertical vein could improve the morbidity and mortality because of its ability to unload the small, non-compliant left sided cardiac chambers by acting as a temporary reservoir for pulmonary venous blood[3]. In infracardiac TAPVC repair with sutureless technique, the vertical vein can be left untouched to preserve the pericardial continuity especially if there is some evidence of obstruction on preoperative echocardiographic examinations.

In our case, the vertical vein had shown evidence of moderate obstruction at the entry point to the right hepatic vein. Almost all infracardiac TAPVC cases we have operated in our institution were obstructed partially or completely at the level of ductus venosus, portal vein, IVC or hepatic veins. In the index case, however, the vertical vein reopened postoperatively, and it resulted in a significant left right shunt leading to prolonged ICU stay and delayed recovery. Therefore, it was decided to occlude the vertical vein. Surgical closure would have resulted in significant morbidity as the baby was unstable to undergo another bypass, the sutureless technique precludes the thoracotomy approach and the need for peritoneal dialysis precludes abdominal approach. Therefore, the better option was to obliterate the vertical vein by the percutaneous embolization and thereafter the patient had smooth recovery and follow up afterwards[4,5].

Learning points

- We highlight the need for continuous monitoring of patients with unligated vertical veins during TAPVC repair as it can sometimes lead to potential serious post op complications.
- Transcatheter device occlusion is a feasible option for such patients with significant left to right shunt through the vertical vein.

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