Life-saving urgent intervention in a low-birth-weight newborn with obstructed supracardiac total anomalous pulmonary venous connection: Stenting the vertical vein

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Introduction

In total anomalous pulmonary venous connection (TAPVC), pulmonary venous blood flow abnormally returns to join the systemic venous tributary or right atrium, and the pulmonary vascular resistance may be elevated if a form of obstruction occurs in this circuit (1). Neonates and infants with obstructed TAPVC can present with severe cyanosis, pulmonary hypertension, and low cardiac output, requiring emergency surgical intervention (2). The most common site of obstruction is the vertical vein at the level of the left pulmonary artery and the left main bronchus in patients with supracardiac TAPVC (1, 3). Immediate relief of the obstruction is required in these patients (3). Because surgical-intervention mortality is high, particularly in patients with obstructed TAPVC, preoperative stabilization by temporarily relieving the obstruction may improve outcomes in this population (3, 4). This case report describes stent implantation as a short-term palliative treatment for obstructed supracardiac TAPVC.

Case Report

A 3-day-old boy weighing 2.4 kg was referred to our clinic because of central cyanosis, respiratory distress, and hypoxia. Upon admission, he was gasping, with bradycardia and severe metabolic acidosis. The patient was immediately admitted to the pediatric cardiac intensive care, and endotracheal intubation and inotropic support were started. Chest X-ray showed severe pulmonary venous congestion. Echocardiogram revealed that the pulmonary veins were draining into a collector sac posteriorly, the junction of the collector sac and the vertical vein narrowed as it coursed behind the left pulmonary artery, and the vertical vein drained into the innominate vein, causing pulmonary hypertension (70 mm Hg) (Fig. 1a-c). The peak Doppler velocity in the

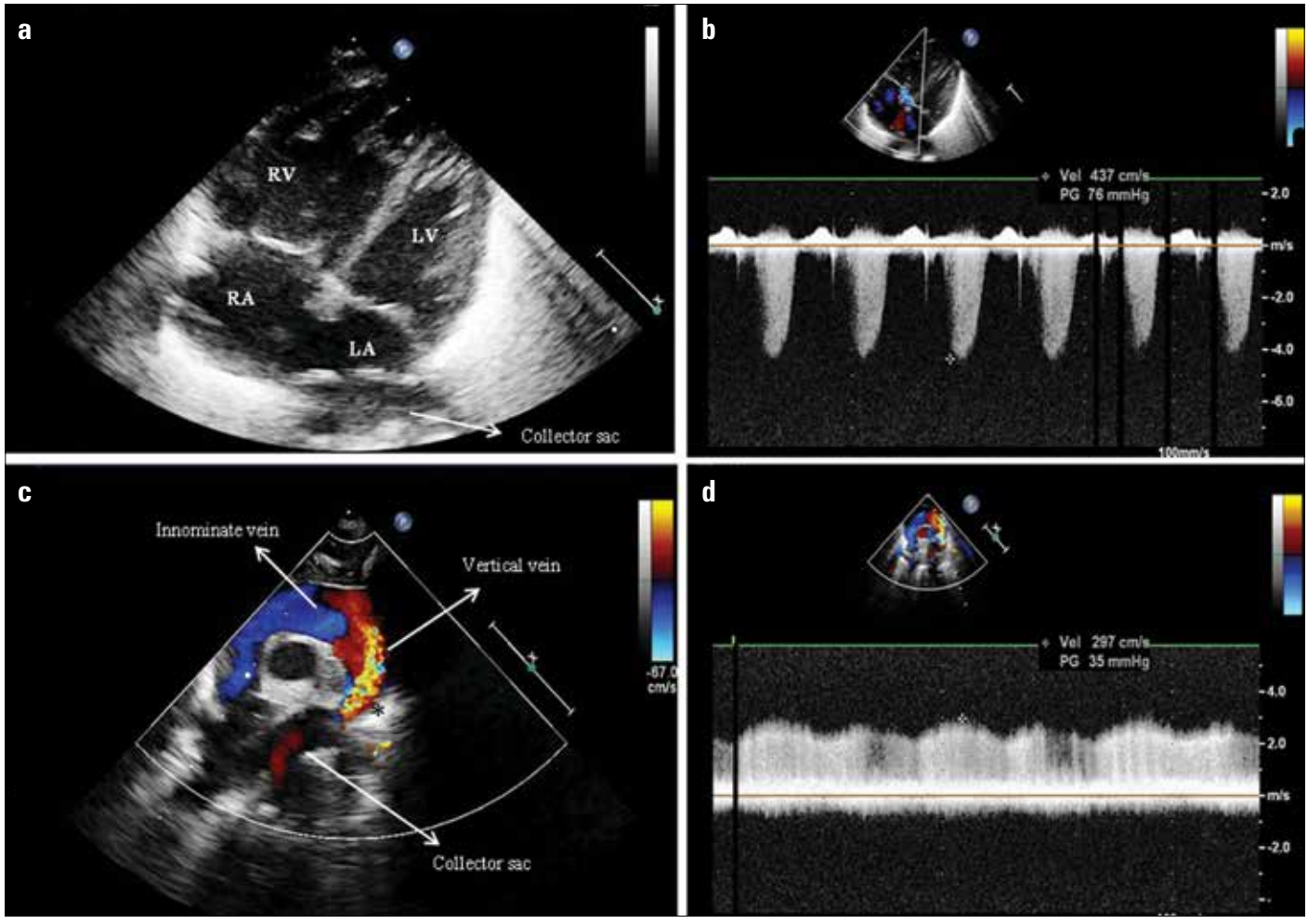

Figure 1. a-d. Echocardiographic views. (a) Four-chamber view, with emphasis on right ventricular enlargement. (b) Continuous wave Doppler of the tricuspid regurgitation. (c) Colour Doppler view of the vertical vein, with emphasis on the narrowed part. (d) Continuous wave Doppler of the narrowed part of the vertical vein

LA - left atrium; LV - left ventricle; RA - right atrium; RV - right ventricle, *shows narrowed part of the vertical vein

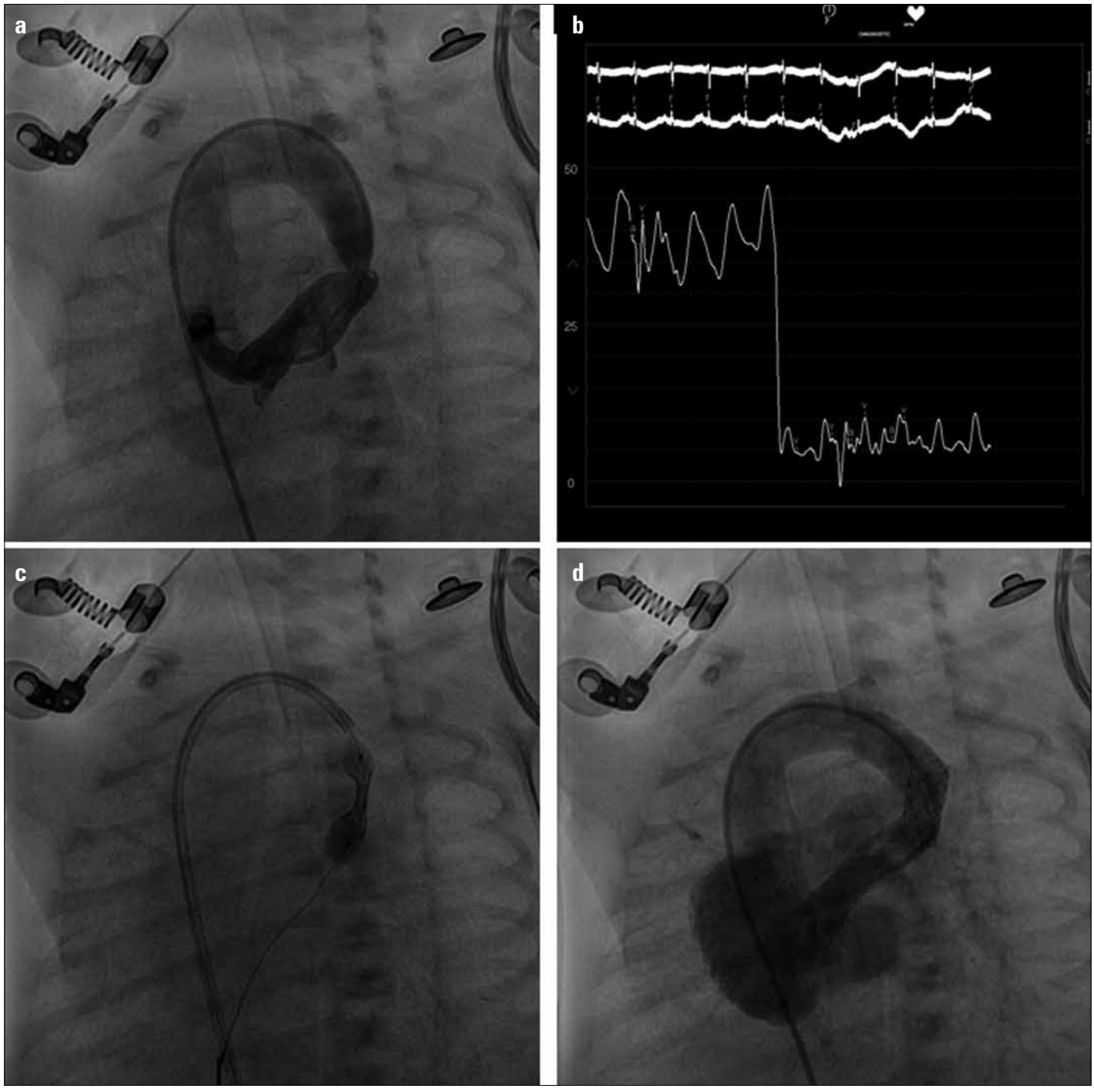

Figure 2. a-d. Angiographic evaluation. (a) Contrast injection into the collector sac, with emphasis on the narrowed part. (b) Pull-back pressure recording from the collector sac to the vertical vein. (c) Opening the stent, with emphasis on the stent waist. (d) Contrast injection into the collector sac after stenting the vertical vein

vertical vein was 3 m/s (Fig. 1d). The saturation was 70% on 100% oxygen.

During cardiac catheterization, selective right and left pulmonary angiography and direct injection of contrast into the proximal segment of the vertical vein showed narrowing in the mid-portion of the vertical vein, with a length of 6 mm and a minimum diameter of 3.6 mm (Fig. 2a). A 4F JR4 catheter was advanced into the collector sac through the femoral vein, inferior vena cava, right atrium, superior vena cava, left innominate vein, and vertical vein. The baseline pressures were as follows: left innominate vein, 8 mm Hg; right and left atrial, 8 mm Hg; right ventricle, 60/9 mm Hg; pulmonary artery, 58/32 (mean: 44) mm Hg; and pulmonary venous confluence mean, 29 mm Hg. The patient was hypotensive from a low cardiac output despite inotropic support (Fig. 2b).

The 4F JR4 was exchanged for a 6F MPA 3.5 guiding catheter. The wire was then exchanged for an extra-support 0.014-inch coronary guidewire to enable the advancement of a 7-mm ×15-mm Palmaz Blue balloon-expandable peripheral stent (Cordis Endovascular, Warren,

NJ). The stent was placed across the stenosis and dilated until the waist completely disappeared (Fig. 2c-d, Video 1). After stent implantation, the pressure gradient across the stent dropped from 21 mm Hg to zero. Echocardiographic and angiographic evaluation of the left pulmonary artery revealed no signs of obstruction. The saturation increased to 80% after the procedure. The patient was extubated soon after the procedure and discharged 4 days later with 85%-90% saturation in the room air. He had a surgical TAPVC repair at the age of 2.5 months (weight 5 kg). He was discharged from the hospital 12 days after the surgery.

Discussion

In patients with TAPVC, the most common drainage site is to the left innominate vein through the left vertical vein. The vertical vein courses posterior to the left pulmonary artery and anterior to the left main bronchus; it then ascends to the anterior mediastinum where it joins the left innominate vein. Obstructed TAPVC can cause severe hypoxia and acidosis because of pulmonary venous congestion (1, 3). Although surgical repair is the primary treatment modality for TAPVC patients, in critically ill patients, pulmonary vein obstruction or low body weight at the time of operation are potential risk factors for hospital mortality (3-6). The operative mortality is up to 7% for isolated TAPVC; this rate is higher in obstructed patients (5). Several previous cases have reported transcatheter interventional procedures as alternative therapeutic options for relief of obstruction and stabilizing clinical status before surgery (1, 3-4).

Conclusion

Stenting of the vertical vein is an effective therapy to acutely stabilize a sick neonate with obstructed supracardiac TAPVC. Catheter intervention should be considered as part of the preoperative cardiovascular stabilization strategy for high-risk infants with obstructed supracardiac TAPVC.

Video 1. Angiographic views.

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Stuck aortic valve treated by reteplase in a Bentall patient

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Introduction

Reteplase, a recombinant plasminogen activator, is indicated for the thrombolytic treatment of suspected myocardial infarction with persistent ST elevation or recent left bundle-branch block within 12 h; however, its use on stuck mechanical valves remains controversial.

Case Report

A 55-year-old woman was admitted to the cardiology clinic with acute onset chest pain, shortness of breath, and palpitation. She had undergone a Bentall operation because of type 1 aortic dissection 3 years ago; however, she did not use her warfarin properly. She had 1 mm of ST elevation in AVR and V1 and 2 mm ST depression in D2, D3, aVF, and V4-V6. She was emergently taken to an angio laboratory, and a huge thrombus material strictly blocking the movement of the aortic valve with resultant severe aortic stenosis was detected. The coronary angiogram was normal. The ejection fraction was 25%. She was sent to the cardiovascular surgery intensive care unit for emergent re-do surgery for the stuck valve. Because the patient had previously undergone the Bentall operation, operative mortality was thought to be considerably high.

Until the patient was prepared for operation, the medical treatment strategy was decided. Reteplase (Rapilysin 10U, Actavis Group, Hafnarfjordur, Iceland) was administered as a 10 U bolus dose followed by a second 10 U bolus dose 30 min later. Each bolus was administered as a slow intravenous injection within 2 min. Heparin and acetylsalicylic acid were also administered before and following the administration of reteplase to reduce the risk of rethrombosis. The patient's clinical status dramatically improved within hours, and the surgery was postponed. Initially, the shortness of breath partially resolved and oxygen saturation began to increase from 85% to 91% in the initial 3 h. During this period, hourly bedside echocardiographic controls were made and the partial movement of the mechanical valve was confirmed. The heparin administration was continued for the following 48 h with control of aPTT every 6 h, with concomitant echocardiographic bedside evaluation of the valve movements. These movements were also confirmed by fluoroscopy at the end of the second day of treatment (Fig. 1). Video 1 shows the recovery of leaflet motion of the