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Case report

Extracorporeal membrane oxygenation as a bridge to surgery in a neonate with total anomalous pulmonary venous return

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Abstract

Total anomalous pulmonary venous return (TAPVR) is a life-threatening condition that requires emergency surgical correction. The diagnosis of TAPVR can be challenging, especially in non-tertiary centers lacking pediatric cardiology support. Extracorporeal membrane oxygenation (ECMO) has been used in some patients as a bridge to cardiac surgery or for postoperative support. We describe a term neonate with severe pulmonary hypertension who was placed on ECMO, which facilitated the diagnosis and subsequent surgical correction of obstructive TAPVR with excellent outcomes.

Keywords

Total anomalous pulmonary venous return, extracorporeal life support, neonatal intensive care.

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Introduction

Total anomalous pulmonary venous return (TAPVR) is a critical cyanotic heart disease that usually presents in the neonatal period [1]. TAPVR is found in 0.6-1.2 per 10,000 live births and accounts for 1-3% of all cases of congenital heart disease [2-4]. Because oxygenated blood returning from the lungs mixes with systemic unoxygenated blood, the affected newborns present with cyanosis.

The four main anatomic types of TAPVR are supracardiac (type 1), intracardiac (type 2), infracardiac (type 3), and mixed (type 4) [2, 5]. In the infracardiac type, which accounts for 22% of cases of TAPVR, the pulmonary veins usually drain into a collector vein, which generally drains into the portal vein [2, 6]. This increases the likelihood of obstruction of the collector vein, which may increase the pulmonary artery pressure and cause secondary pulmonary hypertension.

It may be difficult to distinguish TAPVR from persistent pulmonary hypertension of the newborn (PPHN) because the clinical and echocardiographic features of these disorders may seem similar to untrained professionals. In critically ill neonates presenting with severe respiratory distress without an obvious cause, it is vital to consider TAPVR, although this diagnosis is challenging.

Extracorporeal membrane oxygenation (ECMO), which was initially developed for the support of infants with severe respiratory failure, has been shown to be an excellent pre- and postoperative support technique in patients with congenital heart disease with pulmonary hypertension refractory to treatment [7, 8].

Case report

We present the case of a female born at 39 weeks' gestation by a healthy gravida 1, para 1 mother. Elective cesarean section was performed due to breech presentation at a non-tertiary hospital. The parents were nonconsanguineous and their family history was unremarkable. The pregnancy was uneventful and normal prenatal screening tests (including ultrasounds) were normal. No fetal echocardiogram was performed. Her birth weight was 3,145 g and the Apgar score was 10 at 1, 5, and 10 minutes, with no need for resuscitation. At 6 hours of life, she presented with severe cyanosis (preductal oxygen saturation of 50%) refractory to supplementary oxygen administration, and she developed progressive respiratory distress that required invasive mechanical ventilation. Chest radiography revealed moderate pulmonary congestion (**Fig. 1**) without any other abnormalities. A postnatal screening echocardiogram revealed severe pulmonary hypertension (right-to-left shunting at the level of the patent foramen ovale [PFO] and patent ductus arteriosus [PDA]). Severe PPHN was deemed likely and inhaled nitric oxide therapy was started. Due to hypotension at 24 hours of life, dopamine was started followed by the addition of epinephrine and dobutamine, but with little clinical effect. The serum acute-phase reactants and blood cultures were all negative.

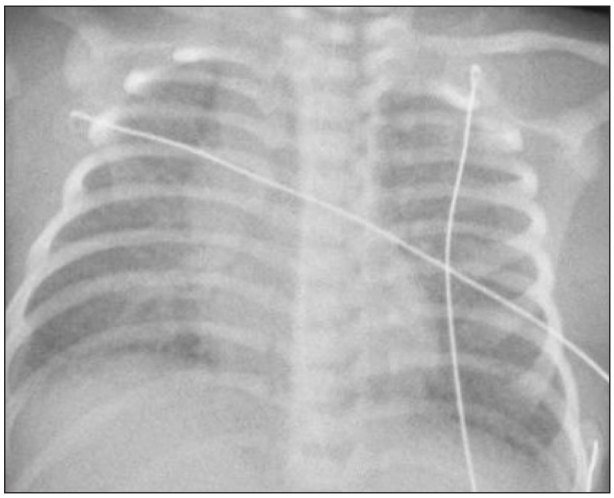

Figure 1. Chest radiograph showing pulmonary congestion without any other abnormalities.

Due to progressive hemodynamic instability and cyanosis refractory to medical therapy, she was placed on veno-arterial ECMO at 36 hours of life with blood pressure stabilization. She was then transferred to a tertiary Pediatric Intensive Care Unit.

Assessment by the pediatric cardiologist at 48 hours of life led to the diagnosis of obstructive infracardiac TAPVR. Echocardiography revealed an infracardiac collector vein coursing through the liver and draining in the portal vein, without a connection between the pulmonary veins and the left atrium (**Fig. 2** and **Fig. 3**). Additionally, there were several indirect signs of pulmonary hypertension: dilated right chambers, impaired right ventricular function, increased pulmonary pressure, and an exclusive right-to-left shunt in the PFO (**Fig. 4** and **Fig. 5**).

She was then transferred to a Pediatric Cardiac Intensive Care Unit and emergency surgical correction of TAPVR was performed. During surgery, the ECMO cannulas were used for cardiac

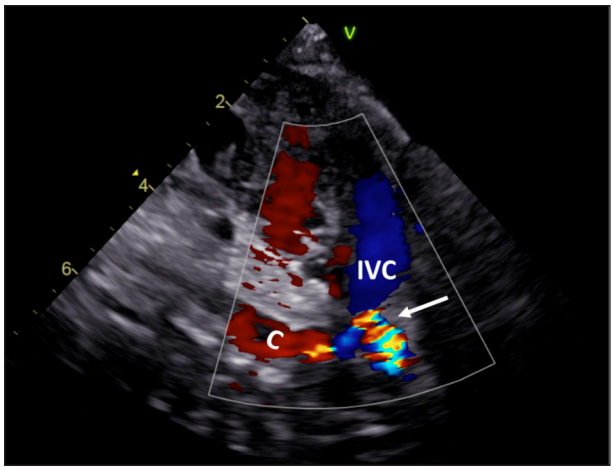

Figure 2. Subxiphoid view showing the venous collector with turbulence at the site of anastomosis with the portal vein (white arrow).
IVC: inferior vena cava; C: venous collector.

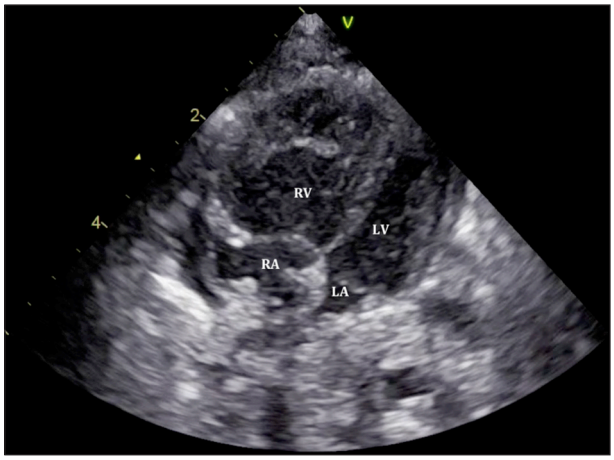

Figure 4. Four chamber view demonstrating indirect signs of pulmonary hypertension, with dilation of the right atrium and the right ventricle. Note the almost entirely collapsed left atrium and the presence of pericardial effusion (an additional sign of right ventricular failure).
LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

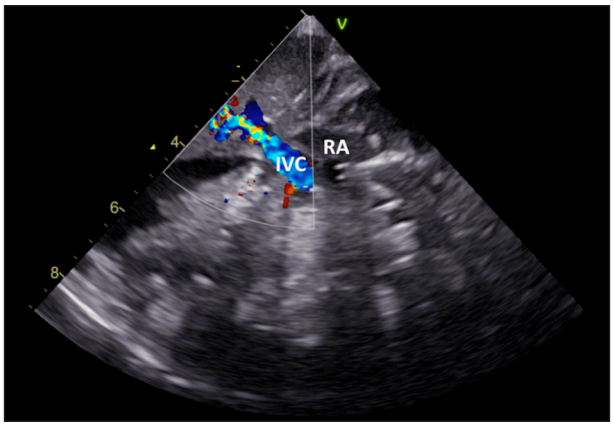

Figure 3. Subxiphoid view showing the inferior vena cava with increased blood flow.
IVC: inferior vena cava; RA: right atrium.

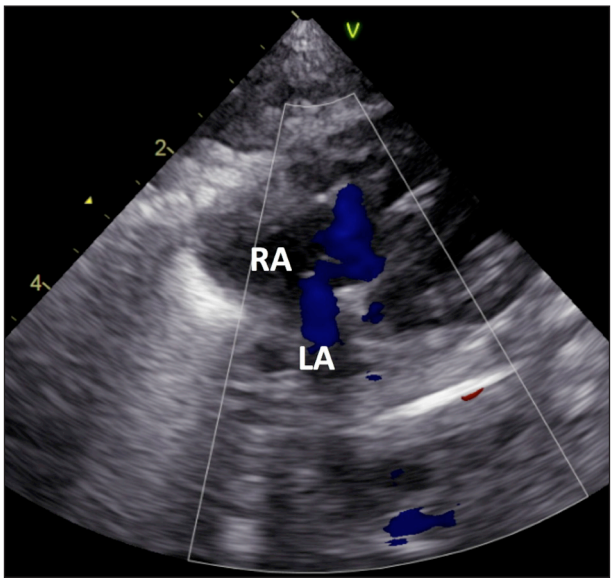

Figure 5. Subxiphoid view showing the right-to-left shunt through the patent foramen ovale (PFO).
LA: left atrium; RA: right atrium.

bypass. The extracorporeal bypass circulation time was 82 minutes and the aortic cross-clamping time was 23 minutes. Postoperatively, junctional ectopic tachycardia without hemodynamic repercussion was noted. On postoperative day 5, she was successfully weaned off ECMO.

The baby was discharged home at 23 days old. At a recent follow-up, at 2 years old, she is completely asymptomatic with normal growth and neurodevelopment for her age.

Discussion

TAPVR is a life-threatening condition that usually presents in the neonatal period. It is crucial to establish an early diagnosis because emergency surgery is necessary to correct the defect. The initial presentation depends on the degree of obstruction of the pulmonary veins. Because obstruction is more common in the infracardiac type, affected neonates are generally in a critical state or even in shock. This is mainly due to the increased pulmonary pressures and decreased cardiac output, presenting as tachypnea, respiratory distress, severe hypotension, and shock.

Upon examination, neonates with TAPVR may have minimal cardiovascular findings. Audible murmurs are rare and the pulmonary component of the second heart sound is louder. They may have signs of right heart failure, such as hepatomegaly and pulmonary rales. The differential diagnosis of

obstructive TAPVR and PPHN can be challenging because the clinical manifestations of these disorders are similar.

The electrocardiogram presents with signs of right ventricular hypertrophy, a non-specific finding. Obstructive TAPVR presents with echocardiographic signs of volume overload of the right ventricle. This means that the right heart chambers and pulmonary artery are dilated and the interatrial and interventricular septum bulges to the left side of the heart during systole. By contrast, PPHN presents with signs of pressure overload of the right ventricle. Opposite to TAPVR, in PPHN, the septum bulges during diastole instead of during systole. However, this difference is particularly difficult to discern in a tachycardic neonate with a low cardiac output. Other echocardiographic features, such as right ventricle hypertrophy, major tricuspid regurgitation, and the obligatory right-to-left shunt in the PFO and/or PDA are merely indicators of high right-side pressures and are not specific to either TAPVR or PPHN. Thus, a structural and functional echocardiogram is necessary to properly identify the anomalous return and the collector vein. Elucidation of the structure will help exclude other causes of cyanotic heart disease and pulmonary venous obstruction.

The use of cardiac computed tomography has increased in recent years, especially in cases in which the echocardiogram cannot establish a definite diagnosis [9]. This modality shows high diagnostic accuracy and can clearly delineate venous malformations, allowing more detailed surgical planning.

ECMO is a successful support therapy, particularly because many patients present with refractory pulmonary hypertension. In such patients, ECMO not only provides a successful bridge to surgery but it also supports patients in the postoperative period until the pulmonary hypertension resolves. Successful pre- and postoperative ECMO support has been reported in several patients with congenital heart diseases, including TAPVR. The usefulness of veno-arterial ECMO, especially in undiagnosed patients presenting with shock, has also been reported [7]. Ishino et al. [8] described 3 neonates with TAPVR in whom veno-arterial ECMO was used as preoperative support and offered fairly good outcomes. Their findings suggest that ECMO provides adequate support to such cases and that it can provide more time to reach a definitive diagnosis.

In conclusion, in critically ill neonates presenting with respiratory distress without an obvious cause, TAPVR should be considered, although this can be an elusive diagnosis. In the present case, ECMO provided more time for the definitive diagnosis to be made, offered a successful bridge to surgery, and was valuable as postoperative support until pulmonary hypertension had resolved. No major complications from this technique were noted and the child presented with no sequelae at her last follow-up visit.

Declaration of interest

The Authors declare that there is no conflict of interest. This research received no specific grant from any funding agency, commercial, or not-for-profit sector.

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