

Infracardiac Total Anomalous Pulmonary Venous Drainage: A Diagnostic Challenge

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Infracardiac total anomalous pulmonary venous drainage can be erroneously diagnosed as respiratory distress of several different etiologies during the neonatal period. A cross-sectional echocardiography study with Doppler color flow mapping can preoperatively determine the precise drainage site and pulmonary venous return pattern, thereby allowing cardiac surgery to be performed promptly, prior to any clinical deterioration.

Introduction

Total anomalous pulmonary venous drainage (TAPVD) is a rare disease, accounting for just 1 to 3% of all congenital heart diseases¹. The infracardiac type is virtually always obstructive, while its clinical manifestations during the neonatal period can be misinterpreted as other causes of respiratory distress. TAPVD has been one of the most challenging diagnoses for neonatologists and pediatric cardiologists. In an autopsy series of 52 patients with total anomalous pulmonary venous drainage associated with fatal outcomes in infancy and early childhood, the diagnosis was unsuspected prior to death in half of the cases². Despite technological advances in echocardiography, the diagnosis of infracardiac total anomalous pulmonary drainage has remained a difficult challenge. Today however, cross-sectional echocardiography analysis with Doppler color flow mapping can provide an accurate delineation of drainage sites.

Surgical mortality has fallen by less than 5% in some institutions, although some factors, such as obstruction of the pulmonary pathway and critically ill infants can adversely affect the surgical outcome³. In contrast, early non-invasive diagnosis can improve surgical outcomes.

We describe a case of obstructive infracardiac TAPVD, diagnosed by echocardiography and sent for immediate surgery²⁻⁵.

Case Report

A three-day-old baby girl, with a term birth weight of 3.28

Key words

Pulmonary veins; echocardiography; heart defects, congenital.

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kg who was problem free during the pre-natal period and labor, developed progressive central cyanosis with respiratory distress during the early hours of life, and was referred to our institution.

On physical examination, the girl appeared severely ill, extremely dyspneic, cyanotic +++/4+ and presented poor peripheral perfusion. On auscultation, she had a regular cardiac rhythm, split second sound with a very loud pulmonary component and a harsh holosystolic heart murmur at the lower left sternal border. A massive hepatomegaly was found on abdominal palpation. The chest X-ray showed mild cardiomegaly, with dilated right heart chambers and pulmonary edema affecting both lung fields. Signs of right ventricle hypertrophy were found on the EKG. The echocardiogram showed infracardiac total anomalous pulmonary venous drainage to the portal vein system. Doppler color flow mapping clearly showed the pulmonary venous return site of the obstruction by the presence of a mosaic indicating a turbulent flow at the connection of the descending common pulmonary vein to the portal system (Figs.1A and 1B), and a high velocity and non-phasic flow was also detected at this site using a pulsed wave Doppler. There was a non-restrictive patent foramen ovale with a right to left shunt. Moderate tricuspid valve regurgitation was detected and a high pulmonary pressure was estimated from the Doppler spectral. The child was promptly referred to surgery, had an uneventful postoperative course and was discharged in a good state of health.

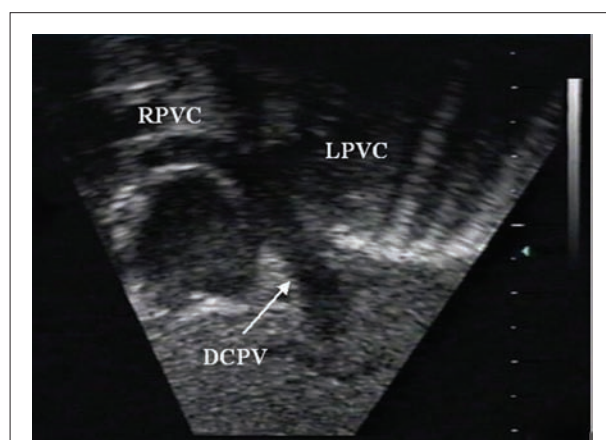


Fig. 1A - Subcostal view showing right and left pulmonary veins draining separately into a confluence chamber to a descending common pulmonary vein (arrow) to the portal venous system. RPVC (right pulmonary vein connection); LPVC (left pulmonary vein connection); DCPV (descending common pulmonary vein).

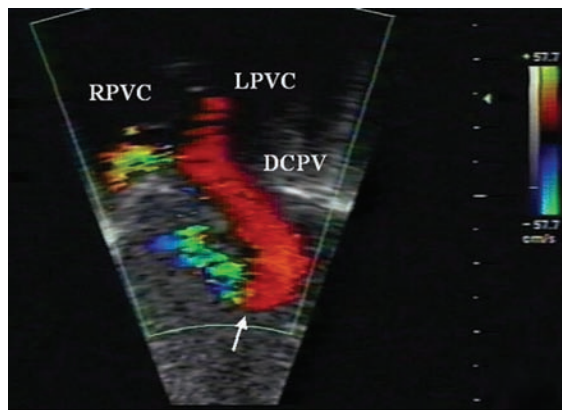


Fig. 1B - The same image with the Doppler colour flow mapping showing marked turbulence at the entry site of the descending vertical and portal venous system (arrow). RPVC (right pulmonary vein connection); LPVC (left pulmonary vein connection).

Discussion

When evaluating a cyanotic neonate with respiratory distress, it is often a challenge to rule out the diagnosis of obstructive anomalous pulmonary venous drainage, particularly the infracardiac type most commonly seen in these circumstances, which has proven to be more difficult to diagnose than other subtypes, even using an echocardiography.

The clinical presentation with mild cyanosis, severe respiratory distress, metabolic acidosis, and no murmur during cardiac auscultation may, in some cases, lead the neonatologist to erroneously diagnose the patient as having hyaline membrane, thereby delaying correct diagnosis and

treatment. Due to its grim and fast deterioration, the correct diagnosis must be made as quickly as possible. In these situations the experience of the cardiologist, and particularly the echocardiographer, is very important.

Based on the echocardiogram, the findings of enlarged right chambers, high pulmonary pressure and a right to left shunt through a patent foramen ovale should raise a high suspicion of this diagnosis, and warrant an exhaustive search for pulmonary venous return.

Since total anomalous pulmonary drainages are not often associated with other cardiac lesions, it is easy for the echocardiographer to misinterpret these findings as persistent fetal circulation¹⁻³. Obstruction is almost universally present in infracardiac pulmonary venous drainage, and in the case of connection to the portal vein when the venous duct is shut, the obstruction is located in the hepatic sinusoids.

Technological advances in echocardiography equipment, particularly Doppler color flow mapping, has made identification of this type of congenital heart disease both quicker and more straightforward⁴⁻⁸. Early recognition prevents inadequate management, which could in turn lead to severe adverse effects including death, or delayed surgical referrals, giving rise to clinical deterioration, respiratory tract infection, and severe pulmonary hypertension, which may have a major adverse effect on morbidity and outcome. Cardiac catheterization should be avoided, as this procedure can be risky for sick babies causing cardiopulmonary function to deteriorate and surgery to be delayed. Therefore, echocardiography has proven to be a reliable tool to detect drainage sites and pulmonary venous obstruction in total anomalous pulmonary drainage. Nowadays it is safe to send a neonate with infracardiac total anomalous pulmonary venous drainage for surgery based solely on echocardiogram findings.

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