


Infracardiac Total Anomalous Pulmonary Venous Drainage: A Case Report

Journal of Diagnostic Medical Sonography
2022, Vol. 38(5) 452–456
© The Author(s) 2022
Article reuse guidelines:
sagepub.com/journals-permissions
DOI: 10.1177/87564793221091258
journals.sagepub.com/home/jdm


Sujith S. Pereira, FRCPCH, PhD^{1,2}  and
Sivathathishana Meinerikandathevan, MBBS, MRCPCH³

Abstract

Anomalous pulmonary venous drainage is a rare cause of congenital heart disease with varied clinical presentation. Symptoms can differ depending on the degree of venous obstruction. An 8-day-old term baby presented with tachypnea and was subsequently found to have an unobstructed infracardiac total anomalous pulmonary venous drainage. She underwent an uneventful surgical repair. The diagnosis of total anomalous pulmonary venous drainage can be delayed or missed, and a high index of suspicion is necessary.

Keywords

anomalous pulmonary venous drainage, congenital heart disease, newborn infant

Congenital heart disease (CHD) has a varied presentation. Total anomalous pulmonary venous drainage (TAPVD) can present as an isolated defect or it could be associated with syndromes such as cat eye syndrome, asplenia syndrome, or Holt-Oram syndrome.¹ In TAPVD, all the pulmonary veins bypass the left atrium and drain into the systemic veins or a chamber which subsequently connects to the right atrium either directly or indirectly. The obligate mixing of pulmonary and systemic venous blood results in cyanosis in the majority of cases.² The commonly used anatomic classification for TAPVD based on the site of connection between the pulmonary and systemic veins includes supracardiac (type I, with anomalous connection at the supracardiac level, is the most frequently encountered type), cardiac (type II, with anomalous connection to the coronary sinus), infracardiac (type III, with anomalous connection below the diaphragm), and mixed (type IV, with mixed types of connections, is the least common type) (see Figure 1). An interatrial communication is necessary to allow blood to enter the systemic circulation for the fetus to survive. Therefore, a patent foramen ovale or atrial septal defect is considered as part of the malformation.³ Heart failure can occur due to excessive pulmonary blood flow with features of pulmonary edema seen on a chest radiograph. An obstruction to pulmonary venous blood flow is seen more frequently in infracardiac TAPVD. This can be due to a number of reasons, such as intrinsic narrowing of the connecting vessel, interposition of the hepatic sinusoids between the pulmonary vascular drainage and the heart

for those that drain via the portal vein, and constriction of the ductus venosus.³ This case report is of an infant whose initial reason for presentation to hospital was tachypnea only.

Case Report

A female infant was born at 39 weeks with a birth weight of 3150 g. She was born by normal vaginal delivery with Apgar scores of 9⁵ and 10¹⁰ to a mother with a Group B Streptococcus colonization, for which the mother received intrapartum antibiotics. Due to the risk factors, the infant had initial observations (temperature of 36.5°C, heart rate [HR]: 125/min, and respiratory rate [RR]: 42/min) done soon after birth which were satisfactory. She continued to have regular observations for the first 12 hours of life all of which were within normal limits. She was breastfeeding well prior to discharge

¹Neonatal Unit, Homerton Hospital, London, UK

²Centre for Genomics and Child Health, Blizard Institute, Barts and the London School of Medicine and Dentistry, London, UK

³Starlight Ward, Homerton Hospital, London, UK

Received January 2, 2022, and accepted for publication February 12, 2022.

Corresponding Author:

Sujith S. Pereira, FRCPCH, PhD, Neonatal Unit, Homerton Hospital, Homerton Row, London E9 6SR, UK.
Email: s.s.pereira@qmul.ac.uk

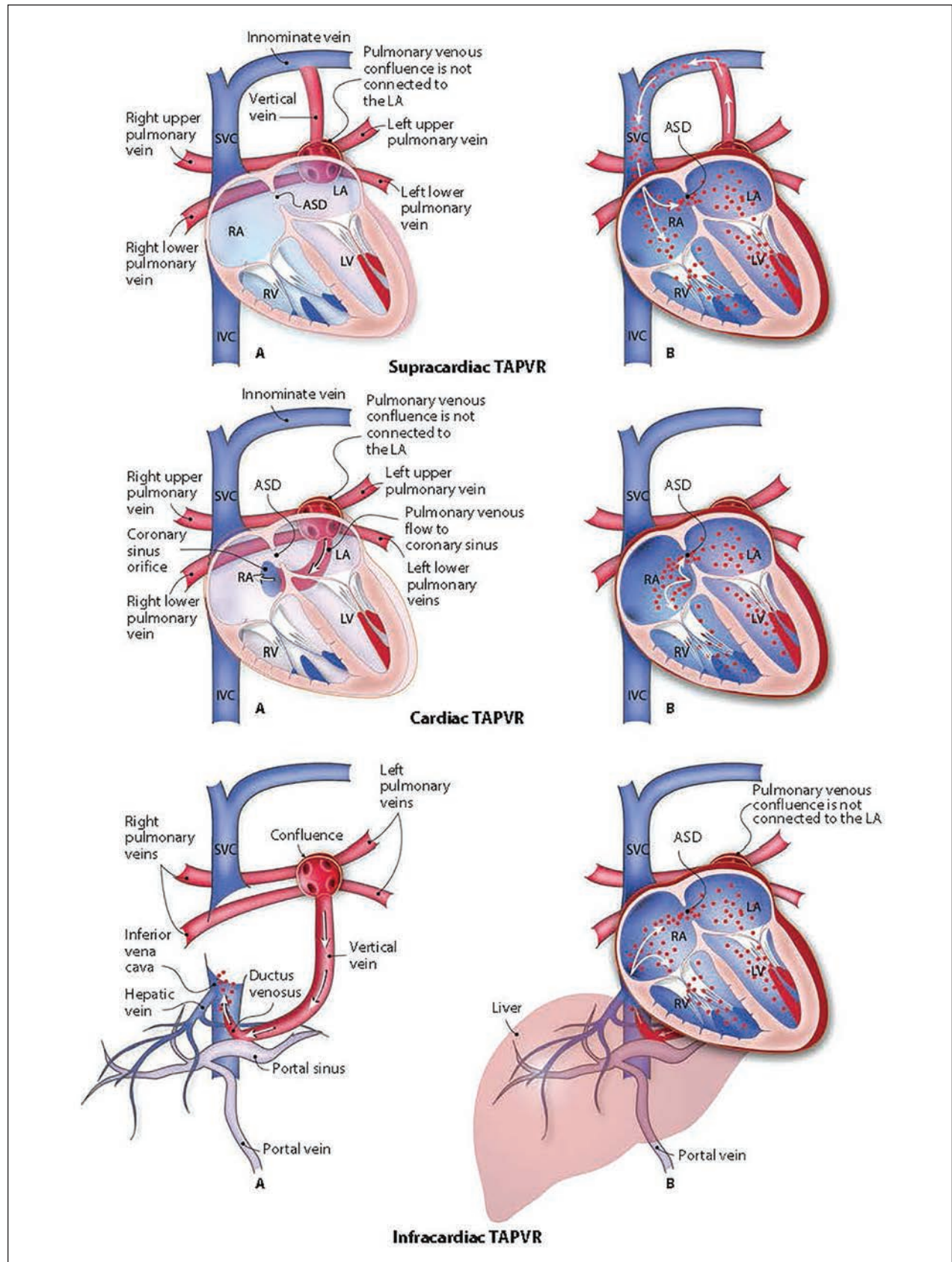


Figure 1. The different types of total anomalous pulmonary venous drainage. ASD, atrial septal defect; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; SVC, superior vena cava; TAPVR, total anomalous pulmonary venous return.

Source. Reproduced with permission from Siassi et al.¹(p259)

home on day 1. There was no family history of CHD or intercurrent viral illness in other family members.

She presented to the hospital on day 8 with fast breathing reported by her mother. She was afebrile with an RR 68/min, HR 168/min, and saturation of 97% to 98% in air initially. However, after 3 hours, she required 0.3 L/min of nasal cannula oxygen to maintain saturations >95% (85% in air). Capillary blood gases were normal. She received antibiotics, her investigations for infection (blood, urine, and cerebrospinal fluid culture; respiratory panel; and COVID-19 screen) were negative. The chest radiograph showed reticular shadowing with bilateral pleural effusion (see Supplemental figure). A cardiovascular examination revealed normal peripheral pulses with normal heart sounds and no audible murmur. Her clinical picture remained unchanged throughout admission to the hospital.

An echocardiogram was performed using Vivid S70N Ultra Edition (GE Healthcare, Buckinghamshire UK) with the 6 MHz sector array probe. Imaging using multiple views is crucial for diagnosing CHD. Subcostal imaging is important to visualize the atrial septum and to determine the direction of shunting using color Doppler. This provides an important clue in the diagnosis of TAPVD. In addition to subcostal views, apical, left parasternal, and suprasternal views were also performed along with other standard and nonstandard echocardiography views to study the cardiac anatomy very carefully. To diagnose TAPVD, it is important to trace the ascending and descending veins carefully using the suprasternal and subcostal views, respectively. Nyquist limits were set at 60 cm/s and adjusted if indicated. Simultaneous two-dimensional (2D) echocardiography and color Doppler were used to interrogate the anatomy and direction of blood flow, respectively.

The echocardiogram revealed a pure right to left flow across the interatrial communication with absent pulmonary venous drainage into the left atrium (see Figure 2). The right side of the heart was enlarged compared with the left side with evidence of ventricular septal flattening (see Figure 3). The regurgitant jet across the tricuspid valve measured nearly 5 m/s (indicating a pulmonary pressure gradient of around 100 mmHg; see Figure 4). In addition, there was also evidence of pulmonary regurgitation (see Figure 5). These findings are in keeping with elevated right-sided pressures. A large vessel was also noted which was consistent with a descending vein, with venous flow profile, traversing the liver to join the hepatic veins and ductus venosus which subsequently drained via the inferior vena cava into the right atrium (see Figure 6). The suprasternal short axis view demonstrated a dilated pulmonary artery with a dilated inferior vena cava draining into the right atrium (see Figure 7). The echocardiographic findings supported an unobstructed infracardiac TAPVD. There was no obvious evidence of acceleration of blood seen on color Doppler. Her case was discussed



Figure 2. Subcostal imaging demonstrating blood flow from enlarged RA to LA across the interatrial communication (see Complimentary Video 1, Supplemental Material). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.



Figure 3. An apical four-chamber cardiac view illustrates the enlargement of the right atrium and right ventricle, along with ventricular septal deviation toward the left side (see Complimentary Video 2, Supplemental Material). IAS, interatrial septum; IVS, interventricular septum; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

with the cardiologists, and she was transferred to a local cardiac center. The infant underwent an uneventful surgical repair of the total anomalous venous connection and patch closure of the secundum atrial septal defect. She

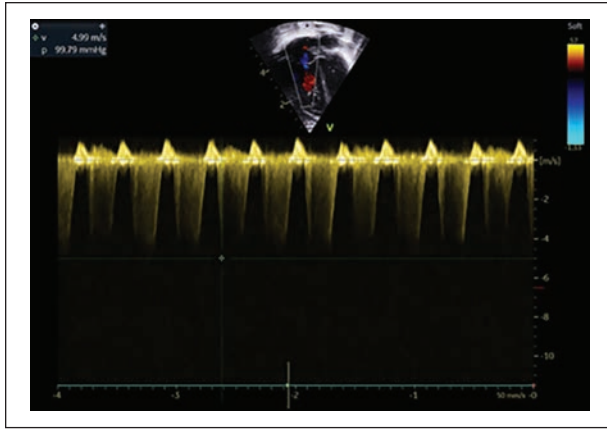


Figure 4. Doppler spectrogram demonstrating the regurgitant jet across the tricuspid valve.

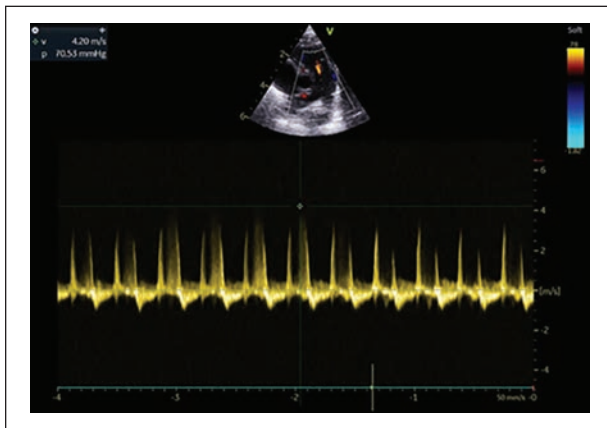


Figure 5. Doppler spectrogram illustrating the pulmonary regurgitant jet.

has been progressing well and thriving. Her cardiology follow-up at 6 months showed unobstructed pulmonary venous flow with no acceleration, no residual atrial communication, and good biventricular systolic function.

Discussion

TAPVD is a rare form of CHD in which all pulmonary veins that bypass the left atrium connect to the systemic veins, right atrium, or coronary sinus.² TAPVD represents 1.5% of CHD⁴ with a reported incidence of 7.1 per 100 000 live births.² Among cases of TAPVD, majority were supracardiac (43%), followed by cardiac (27%), infracardiac (19%), and mixed (11%) types.⁵ Symptoms include cyanosis (43%), respiratory distress (32%),



Figure 6. An echocardiographic view that demonstrates the presence of a descending vein (with red flow) traversing the liver to join the hepatic veins which subsequently joins (blue flow) the inferior vena cava to enter the right atrium (see Complimentary Video 3, Supplemental Material). AA, abdominal aorta; DV, descending vein; HV, hepatic vein; LA, left atrium; RA, right atrium.



Figure 7. A suprasternal cardiac view illustrates blood flowing into the right atrium (see Complimentary Video 4, Supplemental Material). Ao, aorta; IVC, inferior vena cava; PA, pulmonary artery; RA, right atrium; SVC, superior vena cava.

failure to thrive (11%), circulatory collapse (4.5%), murmur (0.5%), and supraventricular tachycardia (0.5%).²

In unobstructed TAPVD, saturated blood from the inferior vena cava is preferentially guided to flow from the right atrium into the left atrium via the atrial septal defect as demonstrated in this case with pure right to left (blue flow) on color Doppler. Dilatation of the right side of the heart with ventricular septal flattening and a dilated pulmonary artery all support elevated pulmonary pressures. In addition, the interatrial septum was thick with a small interatrial communication. This may explain the cyanosis and tachypnea. The tachycardia could be due to reduction in the size of the atrial communication and ventricular septal flattening which compromised left ventricular filling resulting in low cardiac output.

Clinical presentation and pathophysiology vary depending on the presence of pulmonary venous obstruction (PVO), which is frequently seen with infracardiac TAPVD (75%).⁶ In the absence of obstruction, it is not uncommon for the infant to be acyanotic. As resistance to pulmonary circulation is significantly lower than systemic circulation, significant pulmonary over-circulation can ensue with resulting right ventricular dilatation and hypertrophy mimicking varying degree of pulmonary hypertension.³ Severe PVO presents with cyanosis, respiratory failure, and shock. In the absence of PVO, infants may present with failure to thrive, tachypnea, or mild hypoxia.

In a large multicenter case series² examining 422 cases of TAPVD, 110 (26.1%) infants were found to have infracardiac type of TAPVD; 215 (50.9%) of 422 cases had PVO. Severe preoperative PVO was found in 32.3% who presented at a median age of 3 days. On multivariable analysis in this series, infracardiac TAPVD was found to be a significant risk factor for preoperative PVO with an odds ratio of 2.21 (95% confidence interval [CI] = 1.16–4.21).

Conclusion

Prenatal diagnosis of TAPVD can be challenging^{7,8} but has been associated with improved postoperative clinical status and reduced length of hospital stay. Early and accurate diagnosis reduces morbidity and mortality. A high index of suspicion is required in any infant presenting with tachypnea and hypoxia with absent infection markers.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

ORCID iD

Sujith S. Pereira  <https://orcid.org/0000-0001-7104-6872>

Supplemental Material



Supplemental material for this article is available online. Scan the QR code with your smart device to view the online supplemental videos available at <https://journals.sagepub.com/doi/suppl/10.1177/87564793221091258>.

References

1. Siassi B, Noori S, Acherman RJ, Wong PC: *Practical Neonatal Echocardiography*. China: McGraw-Hill Publication, 2019.
2. Seale AN, Uemura H, Webber SA, et al: British Congenital Cardiac Association. Total anomalous pulmonary venous connection: morphology and outcome from an international population-based study. *Circulation*. 2010;122:2718–2726.
3. Lai WW, Mertens LL, Cohen MS, Geva T: *Echocardiography in Pediatric and Congenital Heart Disease*. 2nd ed. Singapore: Wiley Blackwell, 2015.
4. Dillman JR, Yarram SG, Hernandez RJ: Imaging of pulmonary venous developmental anomalies. *AJR Am J Roentgenol*. 2009;192:1272–1285.
5. Lemaire A, DiFilippo S, Parienti JJ, et al: Total anomalous pulmonary venous connection: a 40 years' experience analysis. *Thorac Cardiovasc Surg*. 2017;65:9–17.
6. Karamlou T, Gurofsky R, Al Sukhni E, et al: Factors associated with mortality and reoperation in 377 children with total anomalous pulmonary venous connection. *Circulation*. 2007;115:1591–1598.
7. Laux D, Fermont L, Bajolle F, Boudjemline Y, Stirnemann J, Bonnet D: Prenatal diagnosis of isolated total anomalous pulmonary venous connection: a series of 10 cases. *Ultrasound Obstet Gynecol*. 2013;41:291–297.
8. Seale AN, Carvalho JS, Gardiner HM, et al: British Congenital Cardiac Association. Total anomalous pulmonary venous connection: impact of prenatal diagnosis. *Ultrasound Obstet Gynecol*. 2012;40:310–318.

JDMS Article – SDMS CME Credit

SDMS members can earn SDMS CME credit by successfully completing the complimentary online CME test in the SDMS Learning Center at learn.sdms.org. Non-members may access the online CME test for a fee. **Note: questions may appear in random order online.**

Article: Infracardiac Total Anomalous Pulmonary Venous Drainage: A Case Report
Authors: Sujith S. Pereira, FRCPCH, PhD, and Sivathatishana Meinerikandathevan, MBBS, MRCPCH
Category: Cardiac (Pediatric) [PE]
Credit: 0.50 SDMS CME Credit

Objectives: After studying the article you will be able to:

- Recognize that symptoms of anomalous pulmonary venous return vary depending on the degree of venous obstruction.
- Identify syndromes associated with total anomalous pulmonary venous return.
- Describe the classifications of total anomalous pulmonary venous return.

-
1. Which syndrome can be associated with total anomalous pulmonary venous return?
 - A. Noonan
 - B. Holt-Oram
 - C. Turner
 - D. Williams
 2. What is the primary sign of total anomalous pulmonary venous return due to the mixing of pulmonary and systemic venous blood?
 - A. Tachycardia
 - B. Bradypnea
 - C. Edema
 - D. Cyanosis
 3. What is the most common type of total anomalous pulmonary venous return?
 - A. I
 - B. II
 - C. III
 - D. IV
 4. What is necessary for a fetus to survive when total anomalous pulmonary venous return is present?
 - A. Interventricular communication
 - B. Presence of azygos vein
 - C. Interatrial communication
 - D. Persistent left superior vena cava
 5. Which finding can be seen on chest radiograph when total anomalous pulmonary venous return results in heart failure?
 - A. Pulmonary edema
 - B. Cardiomegaly
 - C. Pleural effusions
 - D. Displaced cardiac apex