Received: 26 July 2023 | Revised: 8 September 2023 | Accepted: 9 October 2023

DOI: 10.1002/ccr3.8079

Clinical Case Reports Open Access WILEY

CASE REPORT

Unobstructed infracardiac total anomalous pulmonary venous connection in a 7-month-old infant: A rare case report

Eman Shhada[1] | **Ali Alakbar Nahle**[2] | **Hussein Hamdar**[2] | **Ali Jawad**[2] | **Hasan Hasan**[3] | **Mohammad Shadi Hamra**[3] | **Mohannad Saleh**[3] | **Alwaleed Al-dairy**[3] | **Huda Daood**[1]

[1]Pediatric Intensive Care Department, Faculty of Medicine, Children's Hospital, Damascus University, Damascus, Syria

[2]Faculty of Medicine, Damascus University, Damascus, Syria

[3]Department of Cardiac Surgery, Faculty of Medicine, Pediatric Cardiac Surgery Unit, Damascus University, Damascus, Syria

Correspondence
Hussein Hamdar, Faculty of Medicine, Damascus university, AlMazzeh, Damascus, Syria.
Email: hussein_hamdar14@hotmail.com

Key Clinical Message

This case underscores the importance of early detection and treatment for total anomalous pulmonary venous connection, a rare congenital heart condition, through comprehensive newborn physical exams and prompt specialist referrals.

Abstract

Total anomalous pulmonary venous connection (TAPVC) is a rare form of congenital heart disease that is typically diagnosed in neonates. TAPVC has four subtypes, with the infracardiac type at risk of obstruction. TAPVC is usually diagnosed in newborns but can occur in other age groups. In this case, a 7-month-old male with recurrent cyanotic episodes was diagnosed with TAPVC of the infracardiac type using computed tomography angiography. The patient underwent successful surgical repair with a favorable postoperative course and was discharged in stable condition, and further follow-up was not possible beyond 2 months. This case emphasizes the importance of early recognition and management of this condition to prevent the progression of subsequent complications.

KEYWORDS

congenital heart disease, echocardiography—case report, infracardiac, total anomalous pulmonary venous connection

1 | INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a relatively infrequent form of cyanotic congenital heart disease (CHD) that typically manifests during the neonatal period. It represents between 1%–5% of all cases of CHD and has an incidence of 0.6–1.2 per 10,000 live births.[1–4] There are four types of TAPVC according to the connection between the pulmonary veins and the systemic venous circulation (Table 1).[1,2,5] It is possible for any subtype to present with obstruction, especially the infracardiac type. In neonatal cases, the survival of the affected individual relies on right-to-left shunting of blood through various openings, such as an atrial septal defect

TABLE 1 Showing the anatomical types of total anomalous pulmonary venous connection with their description and prevalence.

Type	Description	Incidence (%)
Supracardiac	Pulmonary veins anomalously connect, most commonly to the left innominate vein	45–55
Cardiac	Pulmonary veins drain into the coronary sinus or right atrium directly	20–30
Infracardiac	Pulmonary veins form a vertical vein and drain into the portal vein, hepatic vein, or IVC below the diaphragm	13–25
Mixed	A combination of veins that drain to more than one of the types mentioned above. This is the rarest form	less than 10

(ASD)/patent foramen ovale (PFO), or less commonly, a patent ductus arteriosus (PDA) or ventricular septal defect.[1,2,6,7] Neonates with TAPVC typically exhibit episodes of tachypnea and cyanosis, which indicate a lack of oxygen in the blood.[5,8–10]

Total anomalous pulmonary venous connection is diagnosed through the use of echocardiography. In some cases, computed tomography or magnetic resonance imaging may be necessary to provide a more comprehensive evaluation and details of the structural abnormalities that can be useful in the diagnosis and management.[2,4,11,12] Urgent surgery is necessary during the first few months of life, with a risk of mortality, as up to 80% of affected individuals die within the first year if left untreated.[2,5,13] Additionally, complications such as pulmonary hypertension and obstruction of the pulmonary veins may arise in some cases.[4,6,7] Herein, we report a case of an uncommon type of TAPVC, specifically the infracardiac subtype, without obstruction, in a 7-month-old patient who was diagnosed and underwent successful surgical repair upon diagnosis.

2 | CASE PRESENTATION

A 7-month-old male, born at full term with no obstetric complications, was admitted to our hospital due to a cyanotic episode. The patient had experienced five similar episodes in the month leading up to the diagnosis. The patient has no medical or surgical history, and the family history is significant for type 2 diabetes and hypertension. On examination, the infant displayed rapid breathing at a rate of 70 breaths per minute and increased effort in breathing. Oxygen saturation levels were recorded at 60%, and a systolic ejection murmur was detected on auscultation at the left side of the sternum. No indications of heart failure or shock were observed.

A chest radiograph (CXR) was performed, revealing normal results with notable pulmonary vascular prominence. Subsequently, a transthoracic echocardiogram was conducted, which indicated total anomalous pulmonary venous connection (TAPVC). The pulmonary veins drained into a confluence located behind the left atrium and then drained into a descending vertical vein that emptied into the portal vein, indicating an infracardiac type of TAPVC. The patient also had severe pulmonary hypertension, a sinus atrial septal defect, and patent ductus arteriosus. Further imaging with computed tomography angiography (CTA) was performed, confirming the diagnosis of infracardiac TAPVC with drainage to the portal vein through a vertical vein (Figure 1).

The patient underwent surgery via median sternotomy with total cardiopulmonary bypass. The common pulmonary vein was located behind the pericardium and drained through a vertical vein into the portal vein. Subsequently, the vertical vein was dissected and controlled (Figure 2). A transverse incision was made in the common pulmonary vein, and a parallel incision was performed on the opposing surface of the left atrium. These two incisions were then joined to create a wide anastomosis between the left atrium and the common pulmonary vein. The ventricle was closed using silk sutures and metal clips, and the remainder of the operation was completed without any complications.

Following the surgery, the infant was admitted to the pediatric cardiac intensive care unit, and a low dose of diuretics was prescribed. Within hours, the infant's general condition improved, reaching oxygen saturation levels between 93% and 95%. Two days later, the infant's condition had stabilized. CXR and transthoracic echocardiography both revealed normal results. Ten days postoperatively, the patient was discharged from the hospital. Over a 2-month monitoring period, the patient's postoperative course remained favorable, with no indications of cyanotic episodes or any other postoperative complications. The patient's general health status remained stable and satisfactory throughout this monitoring period. However, we were unable to maintain regular contact thereafter.

3 | DISCUSSION

Total anomalous pulmonary venous connection is a rare congenital cardiac malformation categorized into four anatomical subtypes based on the location of pulmonary venous drainage. In the supracardiac subtype, pulmonary

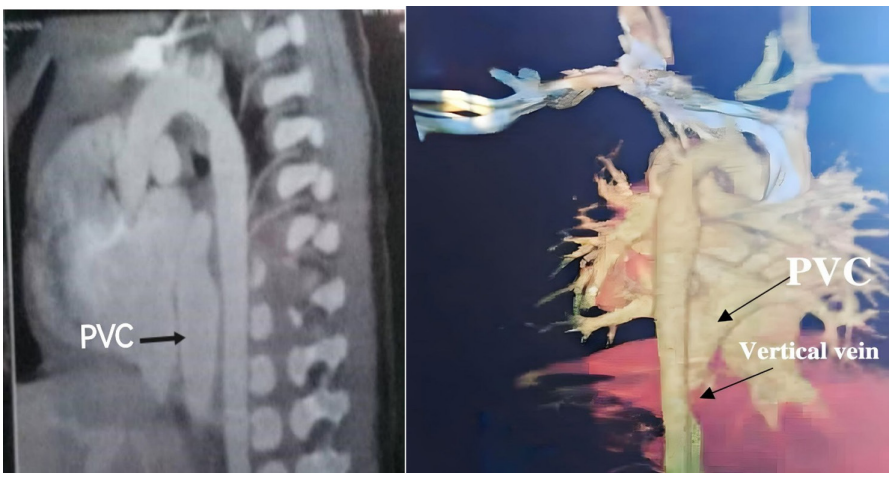

FIGURE 1 Computed tomography angiography showing the dilated pulmonary venous confluence (PVC) and the vertical vein.

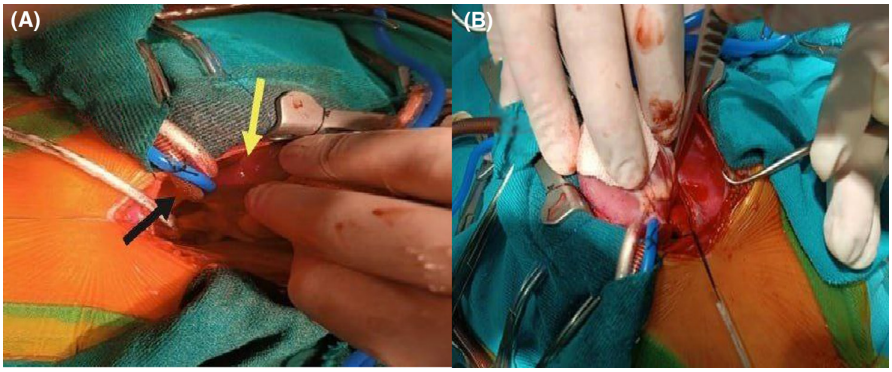

FIGURE 2 (A) Intraoperative image showing the vertical vein (black arrow) and the PVC (yellow arrow). The cannula in blue signifies the cannula for the inferior vena cava. (B) Intraoperative image after the correction.

veins drain into a vertical vein leading to the left brachiocephalic vein. However, in rare cases, pulmonary veins may drain into the superior vena cava, left superior vena cava, or azygous system. In the cardiac subtype, pulmonary veins either drain directly into the right atrium or the coronary sinus. In the infracardiac subtype, drainage is below the diaphragm, potentially flowing into systemic veins like the inferior vena cava, hepatic veins, or azygous system, or into the portal venous system. Lastly, when pulmonary veins drain to two or more locations, above or below the diaphragm, it is classified as a mixed type (Table 1).[14] Obstruction of pulmonary veins may arise in any subtype, but it is most frequently observed in the infracardiac subtype.[1,3,4,8,11] However, in our case, it was unobstructed infracardiac TAPVC, a rare manifestation of this condition. Although most cases of cyanotic heart disease are typically identified during the neonatal period, as indicated by the literature, TAPVC may also present in other age groups. Our case involved a diagnosis of cyanotic heart disease during infancy, identified at a later stage than typically observed in the literature.[1–4] Patients with obstructed TAPVC typically exhibit severe cyanosis and experience low cardiac output and respiratory distress soon after birth.[15] Conversely, patients with TAPVC without pulmonary venous obstruction, as observed in our case, may not exhibit symptoms at birth due to the presence of a systemic pulmonary shunt, which can occur through an atrial septal defect or patent ductus arteriosus (less commonly).[1,2,4,11] However, our case involved both of these conditions, making it difficult to detect cyanosis.

TAPVC is typically associated with high mortality rates in the first few weeks of life. The long-term survival of patients has shown improvement with advancements in medical and surgical management.[16] Complications linked to this condition encompass pulmonary infections, heart failure, and dysrhythmias.[17] Additionally, complications such as pulmonary hypertension and obstruction of the pulmonary veins may arise in some cases.[4,6,7]

In order to plan surgical procedures effectively, one must possess a comprehensive understanding of the anatomy and drainage locations. The initial diagnostic evaluations typically consist of CXR and transthoracic echocardiography. However, in certain cases, echocardiography may have limited utility, primarily due to its restricted viewing field and sonic window. Consequently, CTA and magnetic resonance angiography (MRA) offer significant advantages, providing detailed anatomical information crucial for presurgical planning and postsurgical follow-up.[2] In the presented case, we conducted CXR, echocardiography, and CTA to confirm the diagnosis and gain insight into the anatomy and drainage location.

Total anomalous pulmonary venous connection necessitates immediate surgical intervention within the first few months of life to prevent severe complications. Whenever possible, corrective surgery should occur once the patient's clinical condition stabilizes. For infracardiac and supracardiac TAPVC, surgeons frequently perform procedures to create an anastomosis between the left atrium and the pulmonary venous confluence. Although TAPVC can manifest beyond the neonatal period, our case demonstrates that surgical repair can still succeed, involving the creation of an anastomosis between the left atrium and the common pulmonary vein.[14,18] Our study found that infracardiac TAPVC had successful surgical outcomes, and there was a low incidence of postoperative complications such as pulmonary hypertension and pulmonary venous obstruction. These results are in line with previous studies, and our follow-up evaluations did not reveal any significant issues.[8,11]

4 | CONCLUSION

This case report emphasizes the importance of early diagnosis and management of total anomalous pulmonary venous connection (TAPVC), a rare form of congenital heart disease that can have serious consequences if left untreated. TAPVC is typically diagnosed during the neonatal period, making this case noteworthy due to the later age of diagnosis. Delayed diagnosis can lead to poor outcomes, highlighting the need for physicians to consider TAPVC in patients presenting with symptoms such as tachypnea and cyanosis. Thorough physical examinations from birth can aid in early identification, and prompt referral to a specialist can facilitate appropriate management, including surgical correction. Efforts to improve recognition and management of congenital heart diseases in patients of all ages are crucial.

AUTHOR CONTRIBUTIONS

Eman Shhada: Data curation; writing – original draft; writing – review and editing. **Ali Alakbar Nahle:** Writing – original draft; writing – review and editing. **Hussein Hamdar:** Writing – original draft; writing – review and editing. **Ali Jawad:** Writing – review and editing. **Hasan Hasan:** Resources; writing – review and editing. **Mohammad Shadi Hamra:** Resources; writing – review and editing. **Mohannad Saleh:** Resources; writing – review and editing. **Alwaleed Al-Dairy:** Supervision; visualization. **Huda Daood:** Supervision; visualization.

ACKNOWLEDGMENTS

None.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

Data are available upon request due to privacy/ethical restrictions. The data that support the findings of this study are available upon request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

CONSENT

Written informed consent was obtained from the patient's guardian to publish this report in accordance with the journal's patient consent policy.

ORCID

Eman Shhada https://orcid.org/0000-0001-9286-0945
Ali Alakbar Nahle https://orcid.org/0009-0003-9163-4024
Hussein Hamdar https://orcid.org/0009-0002-5986-4383
Ali Jawad https://orcid.org/0000-0002-7100-8419
Alwaleed Al-dairy https://orcid.org/0000-0002-2239-646X

REFERENCES

1. Samyn MM, Plymale JM, Cousineau AJ, Tweddell JS. TAPVR in a kindergartner presenting with fatigue. *BMJ Case Rep*. 2014;2014:bcr2014206231.
2. Jaramillo FA, Hernandez C, Garzón JP, Herrera APS, Morales MLV. Infracardiac type total anomalous pulmonary venous return with obstruction and dilatation of portal vein. *Radiol Case Rep*. 2017;12(2):229-232.
3. Fu C-M, Wang J-K, Lu C-W, et al. Total anomalous pulmonary venous connection: 15 years' experience of a tertiary care center in Taiwan. *Pediatr Neonatol*. 2012;53(3):164-170.
4. Munsi AS, Hussain M, Rima R, Biswas R, Sayeed A. Clinical profile of patients with Total anomalous pulmonary venous return and their short term outcome in pediatric cardiac Centre at Dhaka Shishu hospital. *Bangladesh J Med Sci*. 2015;14(3):270-273.
5. Reddy KP, Nagarajan R, Rani U, et al. Total anomalous pulmonary venous connection beyond infancy. *Asian Cardiovasc Thorac Ann*. 2011;19(3–4):249-252.
6. Shi G, Zhu Z, Chen J, et al. Total anomalous pulmonary venous connection: the current management strategies in a pediatric cohort of 768 patients. *Circulation*. 2017;135(1):48-58.
7. Sakamoto T, Nagashima M, Umezu K, et al. Long-term outcomes of total correction for isolated total anomalous pulmonary venous connection: lessons from 50-years' experience. *Interact Cardiovasc Thorac Surg*. 2018;27(1):20-26.
8. Chowdhury UK, Airan B, Malhotra A, et al. Mixed total anomalous pulmonary venous connection: anatomic variations,

surgical approach, techniques, and results. *J Thorac Cardiovasc Surg*. 2008;135(1):106-116. e105.
9. White BR, Ho DY, Faerber JA, et al. Repair of total anomalous pulmonary venous connection: risk factors for postoperative obstruction. *Ann Thorac Surg*. 2019;108(1):122-129.
10. Honjo O, Atlin CR, Hamilton BC, et al. Primary sutureless repair for infants with mixed total anomalous pulmonary venous drainage. *Ann Thorac Surg*. 2010;90(3):862-868.
11. Kok LW, Liong WMN, Kong TS. Emergency stenting of vertical vein in a neonate with obstructed supracardiac total anomalous pulmonary venous drainage. *Med J Malaysia*. 2014;69:138.
12. Yoshimura N, Fukahara K, Yamashita A, et al. Current topics in surgery for isolated total anomalous pulmonary venous connection. *Surg Today*. 2014;44:2221-2226.
13. Dillman JR, Yarram SG, Hernandez RJ. Imaging of pulmonary venous developmental anomalies. *AJR Am J Roentgenol*. 2009;192(5):1272-1285.
14. Han D, Pan S, Li H, Meng L, Luo Y, Ou-Yang C. Prognostic value of cardiac cycle efficiency in children undergoing cardiac surgery: a prospective observational study. *Br J Anaesth*. 2020;125(3):321-329.
15. Güzeltaş A, Tanıdır İC, Kasar T. Life-saving urgent intervention in a low-birth-weight newborn with obstructed supracardiac total anomalous pulmonary venous connection: stenting the vertical vein. *Anatol J Cardiol*. 2015;15(4):337-339. doi:10.5152/akd.2015.5938
16. Seale AN, Uemura H, Webber SA, et al. Total anomalous pulmonary venous connection: morphology and outcome from an international population-based study. *Circulation*. 2010;122(25):2718-2726.
17. Burroughs JT, Edwards JE. Total anomalous pulmonary venous connection. *Am Heart J*. 1960;59(6):913-931.
18. Najm HK, Ahmad M, Salam Y, et al. Early outcomes for in situ pericardial roll repair for distant anomalous pulmonary venous return. *Ann Thorac Surg*. 2021;111(1):169-175.

How to cite this article: Shhada E, Nahle AA, Hamdar H, et al. Unobstructed infracardiac total anomalous pulmonary venous connection in a 7-month-old infant: A rare case report. *Clin Case Rep*. 2023;11:e8079. doi:10.1002/ccr3.8079