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Delayed Clinical Presentation of Infracardiac Total Anomalous Pulmonary Venous Return with Portal Venous Drainage

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Abstract

Background: Infracardiac total anomalous pulmonary venous return (TAPVR) is a rare congenital cardiac anomaly that is usually associated with pulmonary venous obstruction and presents early in the neonatal period with severe respiratory distress and hemodynamic compromise. Delayed clinical presentation is uncommon, particularly in cases with portal venous drainage.

Case Presentation: A 19-day-old female neonate presented with sudden cyanosis following feeding after remaining asymptomatic during the first 18 days of life. Chest radiography demonstrated pulmonary congestion. Echocardiography revealed a small left atrium, right-sided cardiac chamber dilatation, severe tricuspid regurgitation, and a posterior pulmonary venous confluence. Subcostal imaging identified a dilated vascular structure at the portal hilum, raising suspicion of anomalous pulmonary venous drainage. Cardiac computed tomography confirmed infracardiac TAPVR, with all pulmonary veins draining through a large vertical vein into the portal venous system. The vertical vein showed marked aneurysmatic dilatation measuring 18 mm. Surgical repair consisting of confluence-to-left atrial anastomosis was successfully performed, while the vertical vein was intentionally left patent. The postoperative course was uneventful, and the patient recovered without major complications.

Conclusion: This case demonstrates that infracardiac TAPVR may occasionally present later than expected despite anatomically severe disease. Awareness of atypical presentations, careful echocardiographic assessment, and prompt confirmation with advanced imaging are essential for timely diagnosis and surgical intervention. Early recognition remains critical to prevent rapid clinical deterioration and potentially fatal outcomes.

Keywords: İnfracardiac, Portal Vein, Total Anomalous Pulmonary Venous Return

Introduction

Total anomalous pulmonary venous return (TAPVR) is a rare congenital cardiac anomaly caused by failure of the common pulmonary vein to incorporate into the left atrium, resulting in drainage of all pulmonary veins into the systemic venous circulation [1]. It accounts for 1–2% of congenital heart diseases and requires surgical correction for survival [1, 2]. The infracardiac subtype, representing 15–20% of cases, typically drains through a descending vertical vein into the portal venous system. The long descending pathway and potential narrowing at the diaphragm or portal level often cause pulmonary venous obstruction, leading to early severe neonatal presentation [2, 3]. Most patients present within the first days of life with respiratory distress and pulmonary hypertension, but rare delayed presentations have been described [4]. We report a neonate with isolated infracardiac TAPVR draining into the portal system via a markedly aneurysmatic vertical vein, who remained relatively stable for the first two postnatal weeks and was diagnosed on day 19, illustrating an unusual clinical course and a rare anatomical feature contributing to delayed presentation.

Case Report

A 19-day-old female neonate was referred to our neonatal intensive care unit due to sudden cyanosis shortly after feeding. Aspiration was initially suspected at the referring center. She was born at 37 weeks via cesarean section, weighing 2205 g, as the first living child of a 22-year-old mother with a history of congenital heart disease and prior open-heart surgery, though no records were available. Routine prenatal follow-up had been uneventful, and she remained asymptomatic until day 18.

On admission, the infant appeared clinically stable, with a pulse oximetry oxygen saturation (SpO_2) of 97% on room air, a heart rate of 132 beats/min, and normal blood pressure. Body weight was 2400 g and length was 47 cm. Physical examination revealed no cardiac murmur or dysmorphic features, and peripheral perfusion was preserved. Venous blood gas analysis demonstrated a pH of 7.40, pCO_2 of 38.4 mmHg, pO_2 of 41.5 mmHg, and a lactate level of 2.2 mmol/L. Chest radiography demonstrated bilateral pulmonary congestion. Respiratory distress syndrome was initially suspected, and surfactant therapy was administered. However, respiratory distress rapidly progressed, necessitating endotracheal intubation.

Transthoracic echocardiography revealed marked right atrial and ventricular dilation with severe tricuspid regurgitation (peak velocity 4.1 m/s), small left atrium, and a posterior confluent pulmonary venous chamber suggestive of anomalous pulmonary venous return. Subcostal imaging showed a dilated venous structure (~14 × 11 mm) at the portal hilum. Abdominal ultrasonography confirmed a dilated venous sac extending from the portal confluence to the hepatic hilum (maximal diameter 11.5 mm) (Fig 1A, 1B).

Cardiac CT angiography demonstrated all pulmonary veins converging posterior to the left atrium without direct connection. A large vertical vein descended through the diaphragmatic hiatus into the portal vein, with aneurysmatic dilatation (~18 mm) (Fig 1C, 1D; Fig 2), consistent with isolated infracardiac TAPVR with portal venous drainage.

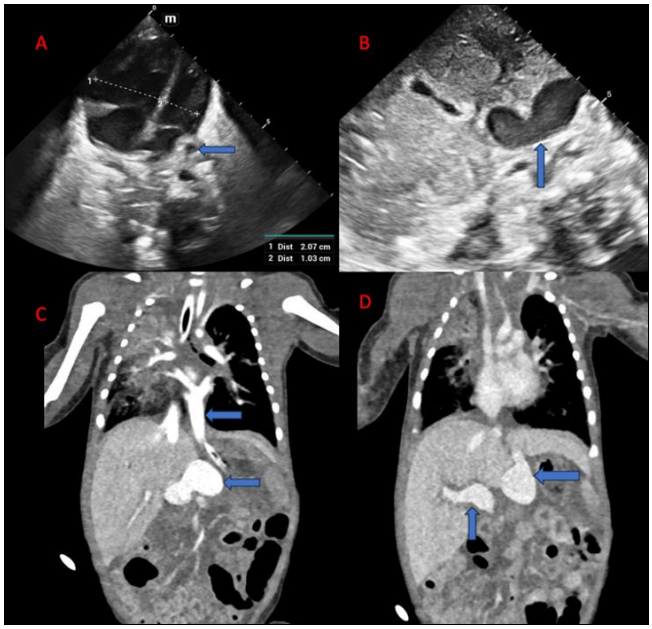

Fig 1: 1A: Transthoracic echocardiography in the apical four-chamber view showing dilation of the right heart chambers and a pulmonary venous confluence posterior to the left atrium. **1B:** Transthoracic echocardiography in the subcostal view demonstrating a venous confluence visible within the liver. **1C:** Cardiac computed tomography showing the vertical vein formed by the convergence of the pulmonary veins. **1D:** Cardiac computed tomography demonstrating the vertical vein and the venous confluence resulting from drainage of the vertical vein into the portal system

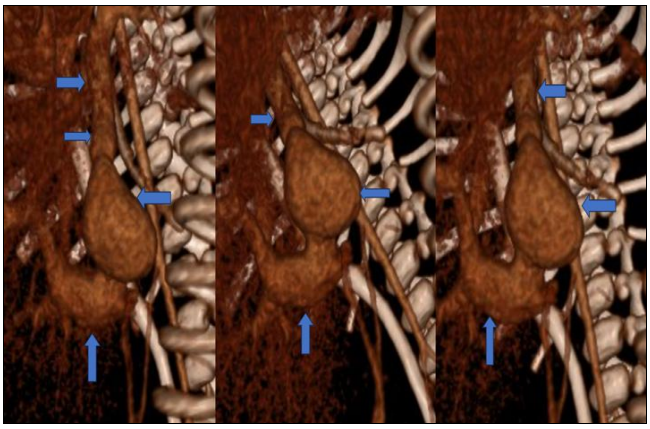

Fig 2: Three-dimensional cardiac computed tomography showing the pulmonary venous confluence and a descending vertical vein draining into the portal venous system, consistent with infracardiac total anomalous pulmonary venous return (blue arrows)

The patient was referred to a tertiary pediatric cardiovascular surgery center for definitive repair. Surgical correction consisted of anastomosis between the pulmonary venous confluence and the left atrium. According to the operative summary provided by the referral surgical team, the vertical vein was intentionally left patent because of elevated pulmonary arterial and right-sided pressures and concern for postoperative pulmonary hypertensive crises. The patient recovered uneventfully after surgery, was successfully extubated, and was discharged from the intensive care unit without reported early postoperative complications before transfer to the pediatric ward. Because the operation and postoperative follow-up were performed at an external referral center, detailed operative records and postoperative echocardiographic data—including cardiopulmonary bypass and aortic cross-clamp times, management of the atrial septum or patent foramen ovale, Doppler gradients across the pulmonary venous anastomosis, and pulmonary artery pressure measurements—were not available despite attempts to obtain additional documentation. This limitation has been acknowledged in the present report.

Discussion

Total anomalous pulmonary venous return (TAPVR) is a rare but life-threatening congenital cardiac anomaly, and the infracardiac subtype is particularly prone to early clinical deterioration due to pulmonary venous obstruction. In a large population-based study, obstruction was significantly more frequent in infracardiac TAPVR compared with supracardiac or cardiac forms [1]. The long descending vertical vein draining into the portal venous system is vulnerable to both extrinsic compression and intrinsic narrowing at the diaphragmatic hiatus or portal entry, resulting in rapid elevation of pulmonary venous pressure. Consequently, most patients with obstructed infracardiac TAPVR present within the first days of life with severe respiratory distress, pulmonary hypertension, and hemodynamic compromise [2, 5]. Pulmonary venous obstruction is a well-recognized predictor of early mortality and postoperative complications [3, 5].

In contrast to this typical clinical course, our patient remained clinically stable during the first two postnatal weeks and presented on day 19 with preserved oxygen saturation. Although presentation during the third postnatal week cannot be considered exceptionally late, it is nevertheless uncommon for an infant with infracardiac TAPVR and significant pulmonary venous obstruction to remain relatively asymptomatic for such a prolonged period [4]. The apparent discrepancy between the preserved pulse oximetry oxygen saturation (97%) and the measured pO_2 also deserves clarification. Review of the original medical records confirmed that the blood gas sample obtained at admission was venous rather than arterial. Therefore, the reported venous pO_2 of 41.5 mmHg reflects mixed venous oxygen tension and should not be directly compared with pulse oximetry-derived arterial oxygen saturation. Accordingly, these findings are physiologically compatible and do not represent an inconsistency in the patient's oxygenation status.

The mechanism underlying the delayed clinical presentation was most likely multifactorial. Previous reports have suggested that a large patent ductus arteriosus with right-to-left shunting may temporarily delay symptom onset by decompressing the pulmonary circulation and maintaining systemic cardiac output. However, no patent ductus arteriosus was present in our patient, excluding this well-recognized protective mechanism. Another important physiological consideration is the ductus venosus. In neonates with infracardiac TAPVR draining into the portal venous system, functional patency of the ductus venosus during the early postnatal period may temporarily provide a lower-resistance venous pathway, thereby reducing resistance to pulmonary venous return. As physiological closure of the ductus venosus progresses during the first weeks of life, resistance within the portal venous circulation may gradually increase, leading to worsening pulmonary venous obstruction and delayed clinical deterioration. Although the ductus venosus was not directly visualized on the available imaging studies, the patient's relatively stable clinical course until day 19 is consistent with this physiological mechanism. Furthermore, cardiac computed tomography demonstrated a markedly enlarged (18 mm) aneurysmal vertical vein at the portal venous confluence. This unusual anatomical finding may have provided additional transient decompression of pulmonary venous return by acting as a temporary capacitance reservoir before progressive obstruction became clinically significant. Therefore, the delayed presentation in our patient was most likely the result of the combined effects of transient ductus venosus patency and temporary decompression provided by the aneurysmal vertical vein rather than either mechanism alone. Reports describing aneurysmal enlargement of the vertical vein in infracardiac TAPVR are extremely limited, making this anatomical feature particularly distinctive [1, 6, 7].

Another important clinical aspect relates to diagnostic delay. Initial suspicion of aspiration and respiratory distress syndrome postponed cardiac evaluation. However, unexplained pulmonary congestion associated with right-sided chamber enlargement in a neonate should prompt careful evaluation for pulmonary venous anomalies, even when pulse oximetry oxygen saturation appears relatively preserved [3, 8]. Early comprehensive echocardiographic assessment remains the cornerstone of diagnosis, whereas multimodality imaging, particularly cardiac computed tomography angiography, provides detailed anatomical information regarding pulmonary venous drainage, the site of obstruction, and surgical planning in complex cases [9].

Surgical repair remains the definitive treatment and involves anastomosis of the pulmonary venous confluence to the left atrium. Management of the vertical vein remains controversial; some centers advocate routine ligation, whereas others recommend leaving it patent in selected patients with elevated pulmonary arterial pressure to provide temporary postoperative decompression [2, 4, 5]. In our patient, according to the operative report received from the referral center, the vertical vein was intentionally left patent because of elevated pulmonary arterial and right-sided pressures. The postoperative course was uneventful, supporting the effectiveness of this individualized surgical approach. Recent surgical series have likewise suggested that selective preservation of the vertical vein may be beneficial in carefully selected patients with elevated pulmonary vascular resistance, although the decision should be individualized according to intraoperative hemodynamic findings [10].

A limitation of this report is the unavailability of detailed operative records and postoperative echocardiographic data because definitive surgical management and follow-up were performed at an external referral center.

Conclusion

Although infracardiac TAPVR usually presents early due to pulmonary venous obstruction, atypical presentations may occur. In this patient, the markedly aneurysmal vertical vein may have partially decompressed pulmonary venous flow, delaying the onset of severe symptoms.

This case highlights the importance of considering pulmonary venous anomalies in infants with unexplained respiratory findings or right-sided cardiac dilation. Early recognition through detailed echocardiographic assessment is essential for timely surgical intervention and favorable outcomes.

List of Abbreviations

TAPVR: Total Anomalous Pulmonary Venous Return
CT: Computed Tomography

Declarations

Ethics approval: Ethical approval was waived by the Akdeniz University Faculty of Medicine Clinical Research Ethics Committee because this study is a single-patient case report and does not constitute human subjects research requiring institutional ethics committee approval under national regulations. All procedures performed were in accordance with the ethical standards of the institutional and national research committees and with the principles of the Declaration of Helsinki.

Consent to participate: Written informed consent was obtained from the patient's parents/legal guardians.

Consent for publication: Written informed consent for publication of this case report and the accompanying images was obtained from the patient's parents/legal guardians.

Clinical trial number: Not applicable.

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Data availability: No datasets were generated or analyzed during the current study.

Conflict of interest: The authors declare that they have no competing interests.

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