Turkish Journal of Pediatric Disease

Case Reports

Pulmonary venous return anomalies simulating primary pulmonary disease in term neonates: Two case reports

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Received : 27.07.2026, Accepted : 10.11.2026
DOI: 10.12956/TJPD.2026.1413

ABSTRACT

Anomalous pulmonary venous return comprises a spectrum extending from partial (PAPVR) to total (TAPVR) drainage of the pulmonary veins into the systemic venous circulation. We report two term neonates who developed respiratory distress, cyanosis, differential oxygen saturation and reticulogranular chest radiographs within hours of birth, closely simulating primary pulmonary disease, although neither had risk factors for respiratory distress syndrome (RDS). In the first infant two consecutive echocardiograms failed to identify a cyanotic defect; computed tomography angiography (CTA) demonstrated cardiac-type TAPVR. He was referred for surgical correction and died within the first 48 postoperative hours. In the second infant echocardiography could not demonstrate right pulmonary venous drainage into the left atrium; CTA showed that both right pulmonary veins drained anomalously, confirming PAPVR. He underwent successful repair and remains alive and asymptomatic. Presented together, these cases show that the RDS-like picture is generated by the transitional pulmonary circulation rather than by the anatomical extent of the anomaly, and that echocardiography may be inconclusive at either end of the spectrum. Anomalous pulmonary venous return should be considered early in term neonates with unexplained hypoxaemia, and CTA obtained promptly when echocardiography is inconclusive.

Keywords: Congenital, heart defects, infant, pulmonary veins, respiratory distress syndrome, tomography

Introduction

Congenital heart defects affect approximately 8 per 1000 live births and remain an important cause of neonatal morbidity and mortality worldwide (1). Anomalous pulmonary venous return forms a continuous anatomical spectrum: in total anomalous pulmonary venous return (TAPVR) all pulmonary veins drain into the systemic venous circulation rather than the left atrium, the presentation varying with the drainage pattern and the severity of pulmonary venous obstruction; in partial anomalous pulmonary venous return (PAPVR) one or more, but not all, veins drain into the right atrium or a systemic vein, producing a left-to-right shunt (2).

Respiratory distress and cyanosis developing shortly after birth are commonly attributed to pulmonary disorders, yet congenital cardiovascular anomalies should also be considered, particularly in term neonates without typical risk factors for respiratory distress syndrome (RDS). A systematic assessment incorporating physical examination, preductal and postductal pulse oximetry, chest radiography and echocardiography is essential to distinguish pulmonary disease from congenital heart disease (3).

Herein we present two term neonates from opposite ends of this spectrum, one with TAPVR and one with PAPVR, whose presentations in the first hours of life were almost indistinguishable and were initially attributed to primary pulmonary disease. Reporting them together is instructive for three reasons. First, the RDS-like picture is produced by pulmonary venous congestion within the transitional circulation rather than by the anatomical extent of the anomaly, so the severity of the presentation cannot be used to estimate how much of the venous return is anomalous. Second, echocardiography was inconclusive at both ends of the spectrum but in opposite ways: a false-negative study in TAPVR and a non-specific finding in PAPVR. Third, computed tomography angiography (CTA) defined the anatomy within hours in both, supporting a low threshold for cross-sectional imaging across the whole spectrum.

Case Reports

Case 1: A male neonate weighing 3020 g was delivered by caesarean section at 37 weeks of gestation to a 31-year-old mother (G4P4). APGAR scores were 6 and 8 at one and five minutes. Shortly after birth he developed tachypnoea, intercostal retractions and peripheral cyanosis.

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Arterial blood gas analysis revealed a pH of 7.13, a pCO_2 of 86.5 mmHg, a pO_2 of 31 mmHg, and a lactate level of 2.8 mmol/L. Due to progressive respiratory distress, the patient was intubated and transferred to the NICU.

Upon admission, a 20% difference in oxygen saturation was noted between the upper and lower extremities, raising suspicion of a congenital heart anomaly. Chest radiography demonstrated bilateral reticulogranular opacities suggestive of RDS, together with elevation of the right hemidiaphragm and a mediastinal shift to the left (Figure 1). In light of the differential saturation, a pulmonary venous return anomaly was considered.

Echocardiography at 6 hours of life revealed a 3 mm interatrial communication, dilatation of the right atrium and right ventricle, and tricuspid regurgitation indicating pulmonary artery pressure at systemic level. The pulmonary venous anatomy could not be established: the individual veins could not be traced into the left atrium, no vertical vein was identified, and the suprasternal and subcostal windows were limited by mechanical ventilation and lung hyperinflation. The study was reported as pulmonary hypertension without a demonstrable cyanotic defect, and oral sildenafil and inhaled nitric oxide were initiated.

Because hypoxaemia persisted, prostaglandin E1 infusion was started empirically to maintain ductal patency while a duct-dependent lesion was excluded. Overall saturation improved, but the gradient between the extremities persisted and the clinical picture was otherwise unchanged. The patient was re-evaluated by the pediatric cardiology team.

Repeat echocardiography again failed to demonstrate a cyanotic defect despite a clinical picture consistent with cyanotic congenital heart disease. CTA was therefore performed and showed that all four pulmonary veins joined a confluence draining into the right atrium, establishing cardiac-type TAPVR. No site of pulmonary venous obstruction was identified, so that the 3 mm interatrial communication, restrictive for a neonate of this size, was the only route of systemic output.

He was transferred to an external pediatric cardiovascular surgery centre at 48 hours of life, where surgical correction was performed, and he died within the first 48 postoperative hours. As the operation and postoperative care took place at another institution, the operative findings and the immediate cause of death were not available to us.

Case 2: A male neonate weighing 2685 g was delivered vaginally at 39 weeks of gestation to a 33-year-old mother (G6P6); APGAR scores were 9 at one and five minutes. Early CPAP was started in the delivery room for grunting and tachypnoea. The respiratory rate was 48/min, with intercostal retractions and peripheral cyanosis, and a 2/6 systolic murmur was audible in the pulmonary area.

Arterial blood gas analysis showed a pH of 7.15, a pCO_2 of 67.8 mmHg, and a lactate level of 4.8 mmol/L. The patient was admitted to the NICU for further evaluation. Pulse oximetry revealed a 15% saturation difference between the upper and lower extremities. Chest radiography demonstrated cardiomegaly and a reticulogranular appearance, suggestive of RDS.

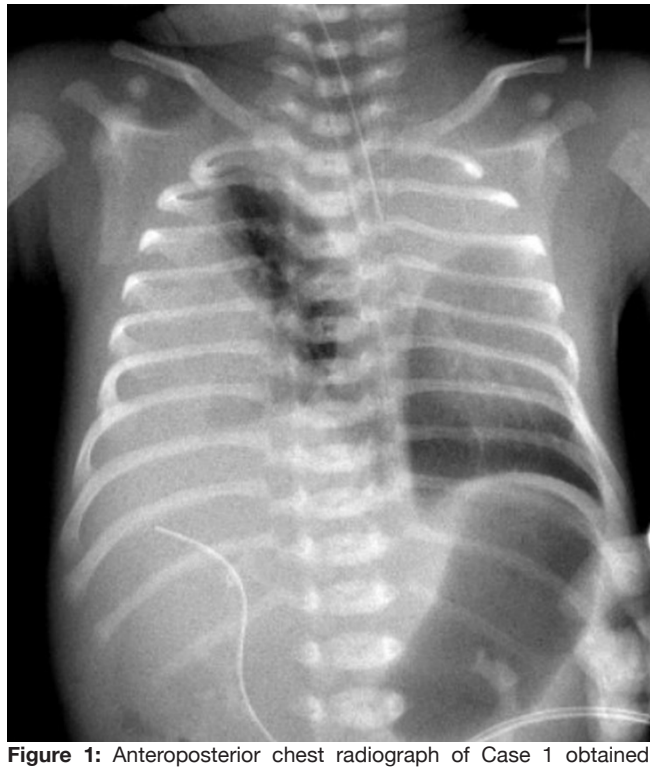


Figure 1: Anteroposterior chest radiograph of Case 1 obtained on admission. Bilateral reticulogranular opacities are present in the mid and lower zones; in this patient they reflect interstitial pulmonary oedema secondary to pulmonary venous congestion and are indistinguishable from the granular pattern of surfactant-deficient respiratory distress syndrome, which is why the radiograph supported rather than challenged the initial diagnosis. The right hemidiaphragm is elevated, with a radiodense area corresponding to the liver projecting over the middle and lower zones of the right lung, and the mediastinum is shifted to the left. Neither of these two findings is a feature of anomalous pulmonary venous return; they reinforced the impression of a primary thoracic abnormality and illustrate how coexisting non-cardiac findings may delay recognition of the underlying venous anomaly.

As in the first patient, this neonate lacked identifiable risk factors for RDS and surfactant was not administered. Although respiratory distress was not severe, peripheral oxygenation remained impaired, and congenital cyanotic heart disease was suspected.

Echocardiography at 3 hours of life showed dilatation of the right atrium and right ventricle. The right pulmonary veins could not be demonstrated entering the left atrium, whereas the left-sided veins were normally connected, and no other structural cyanotic defect was identified.

With a preliminary diagnosis of PAPVR, CTA was performed at 6 hours of life. It demonstrated the right superior pulmonary vein draining into the superior vena cava and the right inferior pulmonary vein draining into the right atrium, with normal connection of the left pulmonary veins to the left atrium (Figure 2). Essentially the entire right lung therefore drained anomalously, producing a shunt considerably larger than that of the single-vein PAPVR most commonly described.

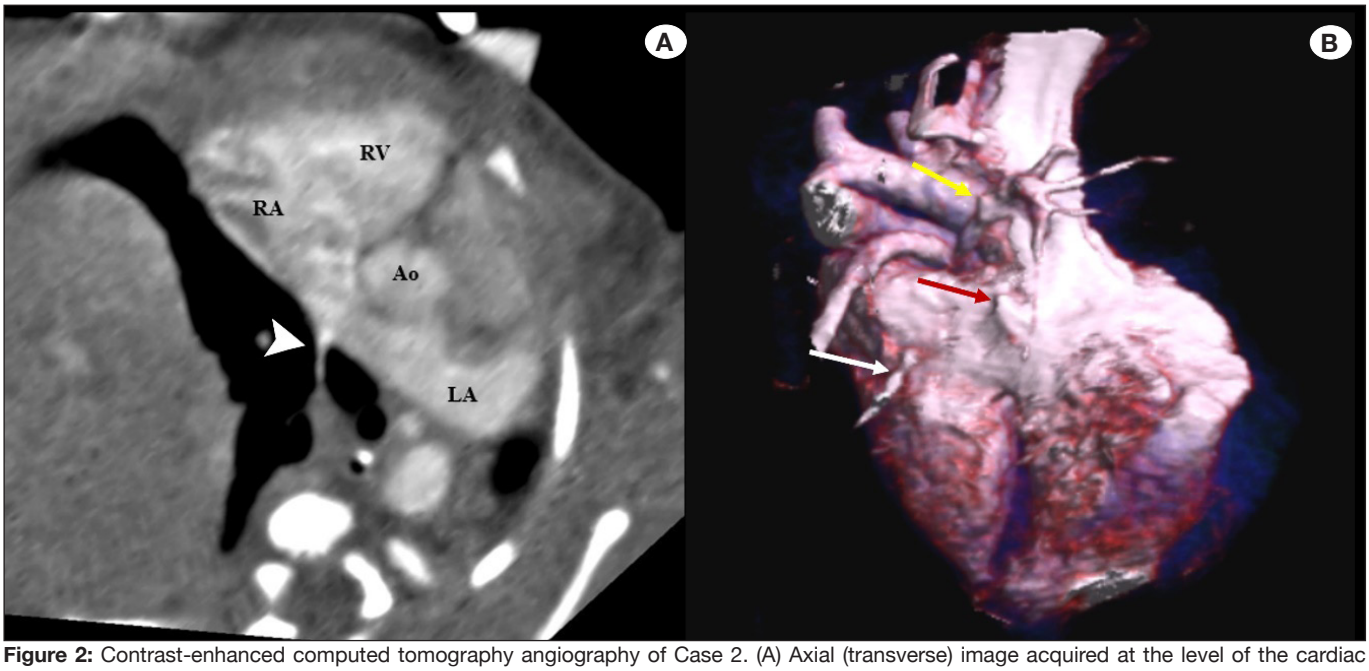


Figure 2: Contrast-enhanced computed tomography angiography of Case 2. (A) Axial (transverse) image acquired at the level of the cardiac chambers shows the right inferior pulmonary vein coursing anteromedially and draining into the right atrium (arrowhead); the interatrial septum, right atrium and left atrium are visible in the same plane. (B) Three-dimensional volume-rendered reconstruction, viewed in a posterior oblique projection, demonstrates the right inferior pulmonary vein draining into the right atrium (red arrow), the right superior pulmonary vein draining into the superior vena cava (yellow arrow), and the left inferior pulmonary vein draining normally into the left atrium (white arrow)

The infant was intubated for progressive respiratory distress and remained hemodynamically stable. He was referred to an external cardiovascular surgery centre at 8 hours of life, where surgical repair was performed. He survived the operation and remains alive and asymptomatic at follow-up; as for the first patient, the operative findings were not available to us.

Discussion

TAPVR is classified as supracardiac, cardiac, infracardiac or mixed according to the site of anomalous connection (2), and severity is determined largely by pulmonary venous obstruction. Obstructed forms, most often infracardiac, present with profound cyanosis, pulmonary oedema and respiratory failure, producing diffuse reticulogranular opacities closely resembling surfactant-deficient RDS (4). Our first patient shows that this mimicry may occur without obstruction, congestion from an unobstructed confluence draining through a restrictive interatrial communication being sufficient to reproduce it.

Because pulmonary blood flow, and therefore pulmonary venous return, is low in fetal life and the anomalous connection is not seen on the standard four-chamber view, TAPVR is among the defects most frequently missed prenatally; only a minority of cases are identified before birth even in specialised centres, and prenatal detection is associated with more expeditious postnatal management (5). Postnatal recognition is therefore the rule, and the burden of timely diagnosis falls on the neonatal team.

Initial evaluation of a cyanotic neonate should include physical examination, preductal and postductal saturation measurement, blood gas analysis and chest radiography (3). A saturation gradient may indicate ductal shunting, pulmonary hypertension or congenital heart disease, but is not specific to anomalous venous return. In our second patient the 2/6 systolic murmur in the pulmonary area is best explained by increased flow across a structurally normal pulmonary valve, a functional consequence of the left-to-right shunt rather than a stenotic lesion; supportive rather than diagnostic, it nevertheless prompted early echocardiography together with the cardiomegaly and the disproportionate hypoxaemia.

Echocardiography remains the first-line modality and diagnoses isolated TAPVR with high accuracy, but its performance falls appreciably with mixed drainage, heterotaxy, associated defects or poor acoustic windows (6). In our first patient two studies performed under mechanical ventilation failed to trace the pulmonary veins to the left atrium; in the second it detected only the absence of right pulmonary venous return to the left atrium, without defining the drainage sites.

CTA depicts the confluence, the individual veins, their systemic connections and any obstruction at high spatial resolution, and is particularly valuable when echocardiography is inconclusive or detailed preoperative anatomy is required (7). Contrast-enhanced magnetic resonance angiography is a comparable alternative that also allows flow quantification and avoids ionising radiation, at the cost of longer acquisition and greater sedation requirements, which limited its feasibility in our hemodynamically fragile patients. No single technique is sufficient, and a multimodality approach improves diagnostic accuracy by covering the blind spots of each (7).

Prostaglandin E1 was started in our first patient while a duct-dependent lesion was excluded, an appropriate empirical measure in a severely hypoxaemic neonate of unknown cause (8). Its benefit in TAPVR is nevertheless limited, because the obstacle lies upstream of the ductus and systemic output depends on the interatrial communication rather than on ductal flow; in obstructed TAPVR, increasing pulmonary blood flow may even aggravate venous congestion. The persistence of the saturation gradient despite improved overall saturation is most consistent with the ductus acting as a right-to-left outlet for suprasystemic pulmonary artery pressure. Prostaglandin is therefore a temporising measure pending anatomical diagnosis, not a treatment.

PAPVR usually produces a modest left-to-right shunt and is asymptomatic or minimally symptomatic, most often found incidentally in later childhood or adulthood; cyanosis is not expected, since anomalous drainage of a few veins does not create right-to-left mixing (9). Marked cyanosis or respiratory compromise in a neonate with PAPVR should therefore prompt a search for another explanation. In our second patient both right pulmonary veins drained anomalously, so essentially an entire lung was involved, approaching a subtotal anomalous return and generating a far larger shunt than isolated single-vein PAPVR. Cyanosis with a preductal to postductal gradient can only arise from right-to-left ductal shunting, implying pulmonary artery pressure at or above systemic level in the first hours of life. The picture is therefore attributable to the transitional circulation acting upon an unusually large anomalous shunt, not to the anomalous connection alone.

Preoperative anatomy strongly determines outcome after repair. Pulmonary venous obstruction is consistently the dominant adverse factor; in a recent ten-year pediatric series survival was 40% with preoperative obstruction versus 79% without (10). Heterotaxy syndrome, single-ventricle physiology, mixed-type drainage, preoperative acidosis and prolonged cardiopulmonary bypass have likewise been associated with increased mortality, and obstructed TAPVR is a surgical emergency (10,11). Our first patient had none of these anatomical high-risk features yet died in the early postoperative period, showing that neonatal TAPVR repair carries substantial mortality even without obstruction, and that diagnostic delay is not benign.

Two features distinguish these patients from those usually reported. TAPVR that escapes detection on two separate echocardiograms is uncommon, and is more often described in mixed-type or heterotaxy-associated disease than in the cardiac type; here the false-negative studies are best explained by the acoustic limitations of a ventilated neonate with hyperinflated lungs. PAPVR presenting in the first hours of life with cyanosis, a saturation gradient and a need for respiratory support is likewise rarely documented, most reports concerning incidental discovery in older children or adults.

Both infants were transferred to another institution for surgery, so the operative findings, the technique used and the immediate cause of the first patient's death were not available to us, and follow-up of the second is limited to clinical status. Abdominal imaging to exclude heterotaxy formally was not performed, and neither cardiac catheterisation nor magnetic resonance imaging was undertaken.

Conclusion

Anomalous pulmonary venous return should be considered early in term neonates with unexplained hypoxaemia, differential oxygen saturation and RDS-like radiographic findings, particularly when typical risk factors for RDS are absent. Because the presentation reflects the transitional pulmonary circulation rather than the anatomical extent of the anomaly, a limited lesion does not exclude a severe presentation, and a non-diagnostic echocardiogram does not exclude the diagnosis. When echocardiography fails to delineate the pulmonary venous anatomy, CTA or magnetic resonance angiography should be obtained without delay, since this interval determines the timing of surgical referral.

Contribution of the authors
Concept: OFK, Design: OFK, EY, DÖY, Data Collection: HA, AFB, DÖY, Analysis/Interpretation: OFK, Literature Review :OFK, FY, Manuscript Writing:OFK, FY, Critical Review :EY

Source of funding
The authors declare the study received no funding.

Conflict of interest
The authors declare that there is no conflict of interest.

Patient confidentiality: Written informed consent was obtained from the patient's parent/legal guardian for participation in the study and publication of the patient's clinical information.

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