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CASE REPORT

Obstructed TAPVC in a Late Preterm Neonate Misinterpreted as Pneumonia: A Reminder to Trust Our Clinical Acumen More

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ABSTRACT

Background: Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart defect (1–2% of congenital heart diseases). The obstructed form, particularly infracardiac or mixed types, often presents as a neonatal emergency with hypoxemia and rapid deterioration, mimicking pneumonia or respiratory distress syndrome (RDS).

Case Presentation: A late-preterm male neonate (35 + 5 weeks, 2.77 kg) developed early tachypnea and hypoxemia (SpO_2 93% on room air). Chest X-ray showed a left upper-zone opacity, and congenital pneumonia was suspected. Despite oxygen, surfactant, and antibiotics, the infant progressed to severe mixed acidosis (pH 7.10, pCO_2 139 mmHg, lactate 11 mmol/L), refractory shock, polycythemia (PCV 71%), and acute kidney injury. Echocardiography initially revealed only an atrial septal defect. The neonate died at 42 hours of life. Retrospective review suggested a missed obstructed TAPVC with ASD.

Conclusion: Obstructed TAPVC should be suspected in neonates with persistent hypoxemia and acidosis unresponsive to routine management. Early repeat echocardiography focusing on pulmonary venous drainage is crucial. This case emphasises that clinical vigilance and timely reassessment remain vital for detecting critical congenital heart disease.

KEYWORDS

• Obstructed TAPVC • Neonatal respiratory distress • Congenital heart disease • Late preterm • Misdiagnosis • Pulmonary venous obstruction • Clinical vigilance • Echocardiography

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INTRODUCTION

Congenital heart diseases (CHDs) represent the most common form of congenital malformations, encompassing a wide spectrum of structural cardiac anomalies that arise from abnormal cardiac development during the embryonic period. Among these, **Total Anomalous Pulmonary Venous Connection (TAPVC)** is an uncommon but clinically significant defect, accounting for approximately **1–2% of all congenital heart diseases.**[1] In TAPVC, the pulmonary veins fail to connect to the left atrium and instead drain oxygenated blood into the right atrium or systemic veins. Consequently, the systemic and pulmonary circulations are completely mixed, and **a right-to-left shunt at the atrial level** often through a patent foramen ovale or atrial septal defect is essential for systemic oxygen delivery and survival.[2] Approximately **15% of TAPVC cases are associated with pulmonary venous obstruction**, which is most frequently seen in the **infracardiac and mixed subtypes**. The presence of obstruction leads to severe pulmonary congestion, hypoxemia, and metabolic acidosis, often resulting in **rapid neonatal deterioration within the first few hours of life.**[2] Anatomically, TAPVC is classified into **supracardiac, cardiac, infracardiac, and mixed** types, each with unique surgical implications requiring accurate preoperative delineation.[3,4]

When the pulmonary venous pathway is obstructed, TAPVC becomes a **neonatal emergency,** typically manifesting with **respiratory distress, cyanosis, and severe acidosis**. However, its presentation often overlaps with more common neonatal conditions such as **congenital pneumonia, respiratory distress syndrome (RDS), or persistent pulmonary hypertension of the newborn (PPHN)** especially in **late-preterm infants** who otherwise have normal antenatal scans.[4] Such overlap may lead to **delayed or missed diagnosis**, with fatal consequences if not promptly recognized. Here, we present a **late-preterm neonate** who was initially diagnosed and managed as a case of congenital pneumonia but was later suspected to have **obstructed TAPVC with an atrial septal defect (ASD).** This case emphasizes the **diagnostic challenges** of differentiating obstructed TAPVC from pulmonary pathology and underscores the enduring importance of **clinical acumen and timely echocardiographic reassessment** in the early identification of critical congenital heart diseases.

CASE PRESENTATION

A late preterm male neonate was born at 35 + 5 weeks of gestation by spontaneous vaginal delivery with a birth weight of 2.77 kg. The baby cried immediately after birth, and no congenital anomalies were noted. Within minutes, however, he developed tachypnoea with oxygen saturation of 93% on room air and was admitted to the NICU for observation. On admission, he was hemodynamically stable, with a heart rate of 148 beats/min and a capillary refill time under three seconds. Systemic examination was otherwise normal. The mother was a primigravida, seronegative for HIV, HBsAg, HCV, and VDRL, with normal antenatal ultrasound findings and no relevant history.

The neonate's respiratory distress progressively increased, requiring oxygen supplementation via nasal prongs, later escalated to CPAP support (PEEP 5 cm H_2O, FiO_2 50%). Orogastric feeds were started and tolerated well initially. Despite these measures, respiratory effort continued to worsen. Chest radiography showed an inhomogeneous opacity in the left upper zone with normal cardiac size and diaphragmatic domes, suggestive of possible congenital pneumonia. The baby was intubated and ventilated (pressure control 12 cm H_2O above PEEP 6, FiO_2 60%), administered surfactant, and started on broad-spectrum antibiotics through a peripherally inserted central catheter.

Subsequently, the neonate developed circulatory shock, requiring fluid boluses and inotropic support with dopamine, later escalated to meropenem and colistin. Polycythaemia (PCV 71%) was detected and treated with partial exchange transfusion. Bedside ultrasound of the cranium and abdomen were normal, while echocardiography revealed a small atrial septal defect with otherwise apparently normal cardiac anatomy. The baby's condition continued to deteriorate, developing inotrope-refractory shock and oliguria, indicating acute kidney injury.

Arterial blood gas analysis showed severe mixed acidosis (pH 7.10, pCO_2 139 mmHg, HCO_3^- 41.3 mmol/L, lactate 11.1 mmol/L) with hyperkalaemia (7.6 mmol/L) and polycythaemia (Hb 22.3 g/dL), consistent with

ventilatory failure and tissue hypoxia. Colistin was discontinued, and corrective measures for hyperkalaemia were instituted. In view of persistent hypoxaemia despite 100% FiO_2 and severe mixed acidosis, differentials considered included severe congenital pneumonia or early-onset sepsis, persistent pulmonary hypertension, and critical congenital heart disease (CCHD). Among CCHD, obstructed total anomalous pulmonary venous connection (TAPVC), transposition of great arteries, hypoplastic left heart syndrome, and pulmonary atresia were considered possible. Despite an apparently normal initial echocardiogram, the refractory hypoxaemia, high lactate levels, and unilateral pulmonary opacity were highly suggestive of obstructed TAPVC with secondary pulmonary venous congestion.

The infant's condition continued to worsen despite maximal ventilatory and inotropic support, culminating in cardiac arrest and death. The final diagnosis was early-onset neonatal sepsis with congenital pneumonia leading to inotrope-refractory septic shock and multiorgan dysfunction, including acute kidney injury and polycythaemia, in a late preterm infant. Retrospective evaluation of the clinical, radiological, and biochemical features indicated a likely missed critical congenital heart disease most consistent with **obstructed total anomalous pulmonary venous connection with atrial septal defect** presenting initially as respiratory distress mimicking neonatal pneumonia.

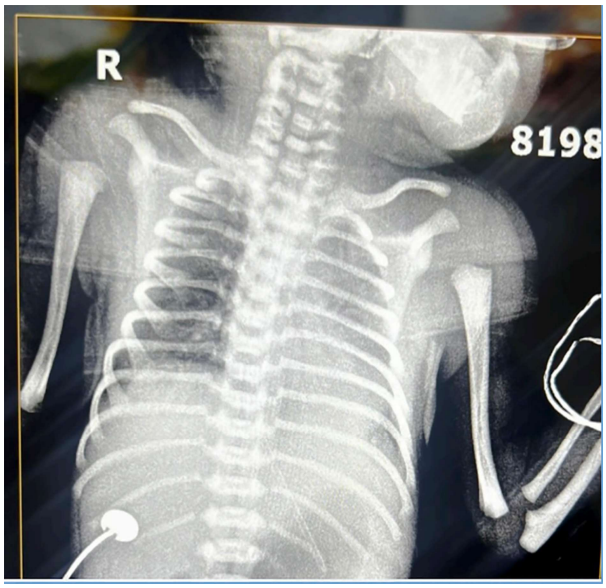

Figure 1: X-ray image showing a left upper-zone opacity

DISCUSSION

The present case of a late-preterm neonate with rapid-onset respiratory distress, refractory shock, severe acidosis, polycythaemia and eventual diagnosis of obstructed TAPVC underscores multiple important clinical and diagnostic considerations.Our case aligns with patterns described in obstructed TAPVC: in a series by M AlMutairi *et al*.,[4] obstructed infracardiac TAPVC presented in the first hours of life with severe respiratory distress and cyanosis.[4] Another study by Xiaoying Xue *et al*.[5] evaluated 48 children with TAPVC and found that the presence of pre-operative obstruction was associated with worse outcomes.[5] A further study by E Ji *et al*.[6] indicated that pre-operative acidosis and pulmonary venous obstruction (PVO) are significant risk factors for mortality following neonatal TAPVC repair.[6]

In our case, neonate had rapid deterioration, with severe mixed acidosis (pH 7.10, pCO_2 139 mmHg, lactate ~11 mmol/L), which is consistent with the physiologic derangement expected from pulmonary venous obstruction and low-cardiac output state as described in literature. The diagnosis was delayed/missed initially (echocardiogram identified only an ASD, not the TAPVC), which explains the challenge of early detection of TAPVC especially when the initial presentation mimics more common neonatal problems (e.g., pneumonia, RDS). In the Al-Mutairi study the emphasis was on distinguishing obstructed TAPVC from neonatal parenchymal lung disease[4]. The Xue *et al*.[5] 10-year retrospective study found that infants with pre-operative PVO had significantly worse outcomes. Our case had features highly suggestive of obstruction (rapid decline, shock, high lactate) and therefore suggestive of the higher-risk category.[5]

Misleading Initial Presentation in our patient, the chest X-ray showed a left upper-zone inhomogeneous opacity, interpreted as congenital pneumonia, and the neonate was treated accordingly. However, obstructed TAPVC can mimic lung disease because of pulmonary venous congestion and oedema, leading to opacities on imaging. The mis-diagnosis toward pneumonia is well described: neonates with TAPVC may present with signs of pulmonary hypertension, oedema, or respiratory failure and receive treatment

for RDS/sepsis rather than structural cardiac disease.[4]

Several physiological red flags in this case should have raised early suspicion of obstructed TAPVC. The neonate showed refractory hypoxaemia and severe hypercarbia (pCO_2 139 mmHg) despite maximal ventilatory support, suggesting a circulatory rather than purely pulmonary cause. The presence of mixed metabolic and respiratory acidosis with markedly elevated lactate (11 mmol/L) indicated profound tissue hypoperfusion and systemic circulatory failure, inconsistent with isolated pneumonia. Additionally, inotrope-dependent shock, oliguria, and acute kidney injury reflected multisystem involvement secondary to low cardiac output. The associated polycythaemia (Hb 22.3 g/dL, PCV 71%) likely further impaired microcirculatory flow and exacerbated hypoxia. These findings pointed toward a central obstructive cardiac pathology rather than a primary pulmonary process.Our initial echo found only an ASD, no detailed delineation of pulmonary venous drainage. The literature emphasizes that even with modern imaging, TAPVC especially when obstructed and in neonates is easy to miss unless one specifically traces the pulmonary venous confluence, vertical vein (if present), and looks for indirect signs such as small left atrium, right-to-left shunt at atrial level, dilated right heart. However, Muntean *et al*.[7] reported a case of prenatally diagnosed obstructed supracardiac TAPVC and discussed the importance of careful 4-chamber, 3-vessel trachea views.[7]

As highlighted by Ji *et al*.[6] pre-operative acidosis and pulmonary venous obstruction significantly increase mortality in neonates with TAPVC, emphasizing that early recognition and timely surgical intervention are critical for survival. In our case, the diagnosis was established late, and no corrective surgery could be attempted, underscoring the tragic consequences of delayed identification. This infant was a late pre-term (35 + 5 weeks), a category not often represented in major TAPVC series, which predominantly include term neonates. Prematurity introduces confounding factors such as relative lung immaturity and increased risk of respiratory distress syndrome or sepsis, which can easily shift the clinician's focus away from congenital heart disease. The coexistence of polycythaemia, although not a classical feature of TAPVC, may have compounded micro-circulatory compromise, exacerbating shock and tissue hypoxia. Furthermore, the severe mixed metabolic respiratory acidosis (pH 7.10, pCO_2 139 mmHg) despite maximal ventilatory support demonstrated the rapid and catastrophic physiological decline typical of obstructed TAPVC, far exceeding the course of pneumonia or RDS.

The initial misdiagnosis as congenital pneumonia represents a well-recognised clinical pitfall; tachypnoea, hypoxaemia, and chest X-ray opacities can easily mimic pulmonary infection. However, when a neonate fails to improve with appropriate therapy antibiotics, surfactant, and ventilation clinicians must actively consider a structural cardiac etiology. Chest radiographs showing upper-zone opacities or pulmonary oedema without a clear infectious focus should raise suspicion of pulmonary venous congestion. An early echocardiographic assessment by a paediatric cardiologist, with careful delineation of the pulmonary venous confluence and drainage pattern, is essential. This case thus illustrates that maintaining high clinical vigilance and timely escalation remain crucial to improving neonatal outcomes in obstructed TAPVC.

CONCLUSION

This case underscores the importance of considering congenital heart disease in neonates with refractory respiratory distress and hypoxemia unresponsive to standard therapy. The overlapping features of prematurity, pneumonia, and sepsis masked the underlying obstructed TAPVC, leading to delayed recognition. Even a normal early echocardiogram does not exclude critical congenital heart disease. Persistent hypoxemia, acidosis, or clinical deterioration should prompt repeat echocardiography with focused assessment of pulmonary venous connections. Our case reports further confirms that while investigations are essential, clinical judgment and vigilance remain irreplaceable.

Table 1: Comparative parameter between our case with other published case

Feature	Our Case	Korkut et al., 2018)[8]	Güzeltaş et al., 2016)[9]
Gestation / Birth weight	Late preterm (35 + 5 weeks), 2.77 kg	Term, 38 weeks, ~3.1 kg at admission[8]	Neonate day 3, ~2.4 kg at referral (low birth weight)[9]
Initial presenting problem	Tachypnea, oxygen saturation 93%, left upper-zone opacity on chest X-ray, presumed congenital pneumonia	Persistent respiratory distress unresponsive to usual therapy (mechanical ventilation + antibiotics)[8]	Gasping, central cyanosis, respiratory distress, severe metabolic acidosis at 3 days old[9]
Radiologic / Chest X-ray findings	Inhomogeneous opacity in left upper zone; normal cardiac silhouette initially	Severe pulmonary venous congestion on chest X-ray (infradiaphragmatic drainage)[8]	Severe pulmonary venous congestion; vertical vein narrowing behind left pulmonary artery on echo/angiography[9]
Diagnostic timing & echo findings	Echocardiogram detected ASD only; underlying TAPVC identified retrospectively	Diagnosis of infracardiac TAPVC made after referral (late)[8]	Obstructed supracardiac TAPVC confirmed on catheterization/ echocardiography (early)[9]
Obstruction present	Yes (inferred) — obstructed TAPVC led to refractory hypoxemia, acidosis, shock	Yes — infracardiac type – obstruction common in this subtype[8]	Yes — supracardiac type with vertical vein obstruction treated with stent[9]
Clinical course	Rapid deterioration: refractory hypoxaemia, mixed acidosis (pH 7.10, pCO_2 139 mmHg, lactate ~11), shock, AKI, polycythaemia → death at ~43 hrs of life	Treatment-resistant distress, delayed diagnosis, emphasised need for early cardiologic evaluation[8]	Emergency palliative stenting of vertical vein reported to improve outcome in obstructed TAPVC[9]

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