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Anesthetic and Perioperative Challenges in Surgical Closure of Persistent Ductus Venosus with Supra-Systemic Pulmonary Hypertension in A Four-Month-Old Infant: Case Report

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Abstract | **Case Report**

Background: Persistent ductus venosus (PDV) is a rare congenital intrahepatic portosystemic shunt with an estimated incidence of 1 in 30,000 live births. It can cause severe pulmonary arterial hypertension (PAH) through persistent shunting of portal blood rich in vasoactive mediators directly into the systemic circulation, bypassing hepatic first-pass metabolism. ***Case presentation:*** A four-month-old female infant with suspected trisomy 21 presented with acute respiratory distress and supra-systemic PAH. CT angiography revealed a patent ductus venosus with a 3-mm portocaval shunt and a markedly dilated umbilical vein (11 mm). Surgical closure was performed; however, the initial clamp test triggered severe hemodynamic instability requiring epinephrine and norepinephrine. The postoperative course was complicated by failed extubation, acute pulmonary edema, and reintubation. On postoperative days three to four, hospital-acquired pneumonia precipitated severe acute respiratory distress syndrome (ARDS). The patient died despite maximal supportive care. ***Conclusions:*** This case illustrates the diagnostic complexity of PDV presenting with evolving PAH initially supra-systemic in the neonatal period and iso-systemic at admission in infancy, the significant perioperative hemodynamic risk of surgical shunt closure, and the potentially fatal vulnerability of these patients to hospital-acquired infections and secondary ARDS. Multidisciplinary management and full preoperative hemodynamic characterization are essential. Earlier diagnosis and intervention, before the establishment of fixed pulmonary vascular disease, may offer the best prospect for survival.

Keywords: Persistent ductus venosus, Portosystemic shunt, Pulmonary arterial hypertension, Congenital heart defect, Surgical closure, Acute respiratory distress syndrome.

INTRODUCTION

The ductus venosus (DV) is a fetal vascular structure that bypasses hepatic circulation by shunting oxygenated umbilical venous blood directly into the inferior vena cava (IVC). Normally, functional closure occurs within hours of birth, followed by anatomical obliteration into the ligamentum venosum within weeks. Persistent ductus venosus (PDV), resulting from failure of this closure, is a rare intrahepatic congenital portosystemic shunt (CPSS) classified as type B1 a direct intrahepatic connection between the portal vein and the inferior vena cava in the Blanc *et al.,* anatomical classification [1], with an estimated incidence of approximately 1 in 30,000 live births [2].

PDV may occur in isolation or in association with chromosomal anomalies, including trisomy 21. Clinical consequences depend on shunt magnitude and include hepatic dysfunction, hyperammonemia, portosystemic encephalopathy, and most severely pulmonary arterial hypertension (PAH) [3]. PAH arises from increased pulmonary blood flow and chronic exposure of the pulmonary vasculature to gut-derived vasoactive substances (serotonin, endothelin-1, thromboxane A_2) that escape hepatic first-pass metabolism [4]. Symptomatic PDV requires shunt closure, surgically or by percutaneous device, but perioperative management in infants with severe PAH carries significant risk [5].

We report a fatal case of PDV presenting as supra systemic PAH in a four-month-old infant who underwent surgical closure and subsequently died from hospital-acquired pneumonia complicated by severe ARDS. This report follows the CARE (Case Report) 2013 guidelines [6].

CASE PRESENTATION

A four-month-old female infant was admitted on March 19, 2026, for acute respiratory distress. Her weight at admission was not formally documented in the medical record; this represents a limitation in interpreting anesthetic dosing and ventilatory settings. She was born at 39 weeks of gestation via cesarean section (birth weight: 3,300 g; Apgar scores: 9/9/10) from a poorly monitored pregnancy complicated by oligohydramnios. Dysmorphic features raising suspicion for trisomy 21 were noted at birth; karyotype results remained pending. The neonatal period required a five-day neonatal intensive care unit (NICU) stay for respiratory distress. Neonatal investigations revealed supra systemic PAH with a 3-mm patent ductus arteriosus (PDA) on echocardiography and congenital hypothyroidism treated with levothyroxine 50 µg/day. All other investigations were normal.

A 48-hour history of dry cough and feeding intolerance preceded admission. Physical examination revealed: tachypnea at 70 breaths/min, perioral cyanosis, cold extremities, SpO_2 96% on 10 L/min oxygen, heart rate 155 bpm, blood pressure 80/40 mmHg, diffuse systolic murmur, hepatomegaly, lower limb edema, and generalized hypotonia with preserved consciousness. Chest radiography showed cardiomegaly. Transthoracic echocardiography (TTE) confirmed PAH iso-systemic at this evaluation (a change from the supra-systemic PAH documented in the neonatal period, likely reflecting partial pressure equalization over time) with severe right ventricular dilatation with D-shaped septal flattening, and a left-to-right PDA.

Thoraco-abdominal CT angiography identified a patent ductus venosus creating a 3-mm porto-caval shunt with marked dilatation of the umbilical vein to 11 mm (Figure 1), confirming the etiology of PAH. Liver enzymes and direct bilirubin were mildly elevated, consistent with reduced hepatic portal perfusion. Ammonia was normal. The clinical timeline is summarized in Table 1 at the end of this section.

Anesthetic management centered on suprasystemic PAH, right ventricular–left ventricular interaction, and prevention of clamp-induced hemodynamic collapse.

Given the severity of supra-systemic PAH, the patient was classified as high anesthetic risk (ASA IV). Preoperative optimization included supplemental oxygen and diuretic therapy. Inhaled nitric oxide (iNO) and preoperative sildenafil were unavailable. Given the presence of supra-systemic pulmonary arterial hypertension, the patient was considered extremely hemodynamically fragile with a high risk of pulmonary hypertensive crisis. Therefore, perioperative goals included strict maintenance of preload, optimal oxygenation, normocapnia, avoidance of acidosis, prevention of hypothermia, and adequate analgesia in order to minimize increases in pulmonary vascular resistance. The unavailability of inhaled nitric oxide (iNO) represented a major limitation in the management of this patient, particularly in the event of acute pulmonary hypertensive decompensation. Preoperative sildenafil therapy could also have been considered as part of pulmonary vasodilator optimization; however, it was not available in our setting. In addition, prostacyclin analogues were unavailable, further restricting access to advanced pulmonary vasodilator strategies.

Anesthesia was induced with sevoflurane 2%, fentanyl 6 µg, propofol 6 mg (administered slowly and in a fractionated manner to minimize the risk of systemic vasodilation and cardiovascular collapse in the context of severe supra-systemic PAH an alternative such as ketamine was considered, but propofol was chosen in low dose given the risk of tachycardia and sympathetic stimulation with ketamine in this setting), and rocuronium 2 mg. Orotracheal intubation was achieved without difficulty (Cormack–Lehane grade I). Volume-controlled mechanical ventilation was applied: FiO_2 100%, tidal volume 25 mL (~7 mL/kg), respiratory rate 40 cycles/min, PEEP 5 cmH_2O targeting mild hyperoxia and normocarbia to minimize pulmonary vascular resistance [7]. Ultrasound-guided right internal jugular central venous access was secured. Invasive arterial pressure monitoring was strongly desired given the hemodynamic complexity of this case; however, it could not be obtained due to the unavailability of an appropriately sized neonatal arterial catheter in our setting.

Intraoperatively, SpO_2 fluctuated between 79% and 100% despite FiO_2 100%. It is important to note that oxygen saturation was already labile in the preoperative period in this patient with supra-systemic PAH; the intraoperative fluctuations should therefore be interpreted in this context, rather than as solely a consequence of the surgical procedure. The first clamp test of the ductus venosus triggered severe hemodynamic instability (heart rate 80 bpm, profound hypotension), which was managed with an epinephrine 40 µg IV bolus followed by norepinephrine infusion [5]. Approximately 40 mL of blood loss required a packed red cell transfusion. Following recovery from hemodynamic collapse, a gradual and cautious approach to re-clamping was adopted. Rather than a second abrupt clamp, the surgical team proceeded with progressive, temporary occlusions of increasing duration, each time pausing to assess hemodynamic tolerance. The decision to proceed with definitive ligation was ultimately based on clinical and hemodynamic tolerance during this graded clamping, without formal portal pressure measurement

the latter representing a recognized limitation of this case. Surgical ligation was completed once sustained hemodynamic stability was demonstrated. This event highlighted the critical importance of close communication between the surgical and anesthesia teams during temporary shunt occlusion. Abrupt clampage of the ductus venosus can lead to catastrophic hemodynamic consequences, including sudden elevation of portal venous pressure, reduction in venous return to the right heart, acute right ventricular decompensation, abrupt changes in systemic and pulmonary vascular resistances, and deleterious right–left ventricular interdependence. In patients with supra-systemic pulmonary hypertension, these rapid physiological changes may precipitate severe cardiovascular collapse.

From an anesthetic perspective, the first clamp test should be interpreted as an acute cardiopulmonary stress test. In a child with supra-systemic PAH, sudden interruption of the portocaval shunt may abruptly increase portal venous pressure while simultaneously reducing direct caval venous return and right ventricular preload. This occurs on a background of a hypertensive pulmonary circulation and a vulnerable right ventricle. The combination of decreased preload, increased right ventricular afterload, and abrupt changes in pulmonary and systemic vascular resistances may rapidly produce right ventricular dilation, septal shift, impaired left ventricular filling, and systemic hypotension. These heart-lung and right-left ventricular interactions explain why clampage can be poorly tolerated and potentially catastrophic if performed abruptly or without continuous anesthesia-surgery communication [12-15].

This mechanism supports a practical anesthetic strategy: announce each clam Perioperative PAH Management and Clamp-Induced Collapse

Editable schematic summarizing the physiopathology of clamp-induced collapse in a patient with suprasystemic pulmonary arterial hypertension.

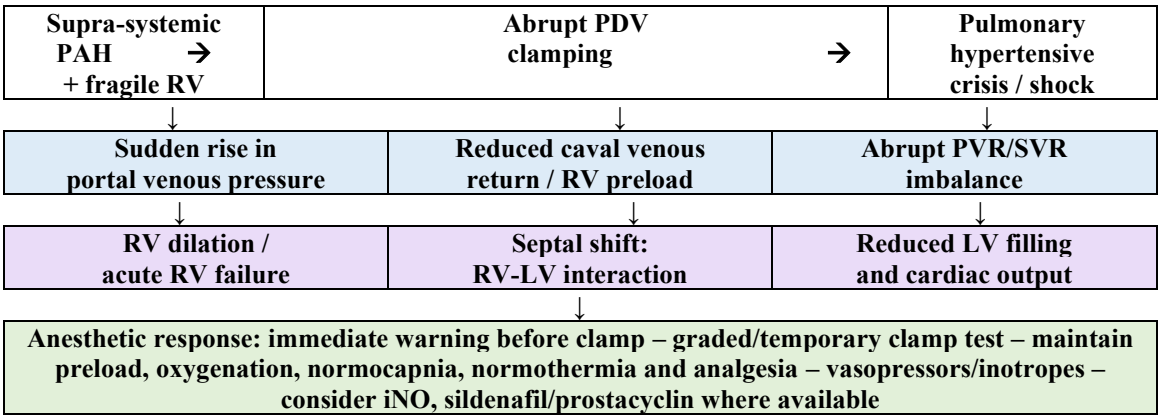

Figure 2: Proposed anesthetic pathophysiological schema of clamp-induced hemodynamic collapse during surgical closure of persistent ductus venosus in supra-systemic pulmonary arterial hypertension

In this case it's important to ensure adequate depth of anesthesia and analgesia, avoid hypoxia, hypercarbia, acidosis and hypothermia, maintain preload without fluid overload, and prepare vasoactive drugs before surgical manipulation. If available, inhaled nitric oxide should be immediately accessible as a selective pulmonary vasodilator during pulmonary hypertensive crisis. Sildenafil and prostacyclin analogues may be considered as pulmonary vasodilator therapy in specialized settings, particularly when severe PAH or right ventricular dysfunction is anticipated. In our case, the absence of iNO, sildenafil and prostacyclin analogues limited rescue options and increased the reliance on meticulous ventilation, preload management, vasopressor support and close surgeon-anesthesiologist coordination.

Postoperative Course and Outcome

The patient was transferred intubated to the pediatric intensive care unit (PICU). On postoperative day one (POD1), a pre-extubation echocardiographic assessment was performed. It demonstrated a persistent but stabilized right ventricular dilatation with moderately elevated estimated pulmonary pressures, no evidence of new ventricular dysfunction, and preserved biventricular function findings that, combined with adequate gas exchange parameters, progressive weaning of vasopressor support, biochemical stability, and full rewarming, led the team to consider extubation a reasonable clinical step at that time. Despite this, the extubation trial failed, requiring nebulized epinephrine and non-invasive ventilation. Acute pulmonary edema subsequently developed and was treated with furosemide.

On POD2, a worsening PaO_2/FiO_2 ratio necessitated reintubation and deep sedation. On POD3–4, the patient developed hospital-acquired pneumonia (HAP) with bilateral pulmonary infiltrates, fever, and rising inflammatory markers. Despite empirical broad-spectrum antibiotic therapy, HAP rapidly progressed to severe ARDS, defined per PALICC criteria (PaO_2/FiO_2 <100 mmHg, PEEP ≥5 cmH_2O, bilateral infiltrates, non-cardiogenic) [8]. Lung-protective ventilation, vasopressor support, corticosteroids were applied. Veno-venous ECMO, a recognized rescue option for refractory

pediatric ARDS [9], was unavailable. The infant died on POD4. The immediate cause of death was refractory severe ARDS secondary to hospital-acquired pneumonia; however, the pre-existing supra-systemic PAH, the perioperative hemodynamic instability, and the cascade of postoperative complications almost certainly contributed to the severity and irreversibility of the final clinical deterioration.

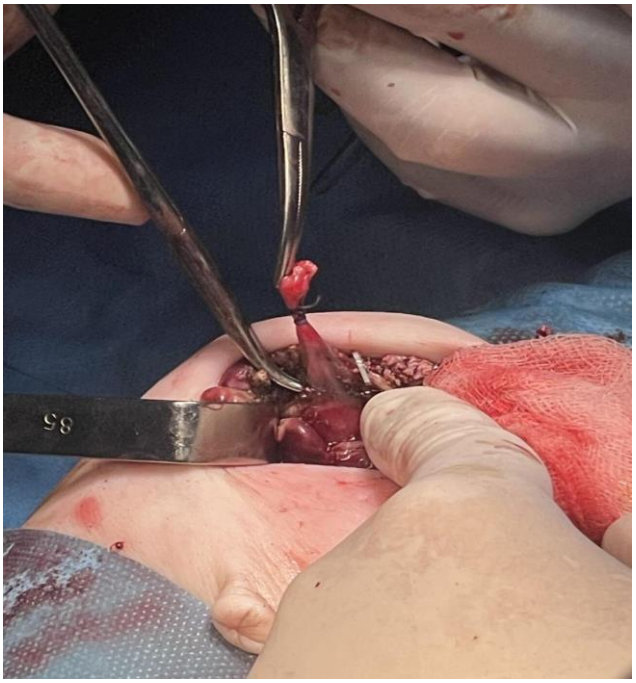

Figure 3

Table 1: Clinical timeline. D: day; POD: postoperative day; PAH: pulmonary arterial hypertension; TTE: transthoracic echocardiography; PDV: patent ductus venosus; NICU: neonatal intensive care unit; NIV: non-invasive ventilation; ARDS: acute respiratory distress syndrome

Date	Event
Birth	Term delivery; Apgar 9/9/10; dysmorphic features; trisomy 21 suspected.
Neonatal (D1–5)	NICU: supra-systemic PAH, 3-mm PDA, congenital hypothyroidism. Levothyroxine started.
March 17, 2026	Onset: dry cough, feeding intolerance, progressive respiratory distress.
March 19 (D0)	Admission: supra-systemic PAH on TTE. CT angiography: PDV, 3-mm porto-caval shunt, 11-mm umbilical vein dilatation.
March 20 (D1)	OR: Induction, intubation. 1st clamp test → severe hemodynamic collapse → epinephrine 40 µg + norepinephrine. 2nd clamp test tolerated. Surgical ligation completed. 40 mL blood transfusion.
March 20–21 (POD1)	PICU transfer intubated. Extubation trial failed. Nebulized epinephrine, NIV. Acute pulmonary edema → furosemide.
March 22 (POD2)	Worsening PaO_2/FiO_2. Reintubation + deep sedation.
March 23–24 (POD3–4)	Hospital-acquired pneumonia → severe ARDS. Maximal support. Death on POD4.

DISCUSSION

This case illustrates three clinically important dimensions of PDV: its initial presentation as supra-systemic PAH in the neonatal period, evolving to iso-systemic PAH at admission, the perioperative hemodynamic instability associated with surgical shunt closure, and the fatal susceptibility to hospital-acquired infections and ARDS.

Pathophysiology of PAH in PDV:

results from two mechanisms: increased pulmonary blood flow due to porto-systemic volume overload, and chronic delivery of gut-derived vasoconstrictors (serotonin, endothelin-1, thromboxane A_2) to the pulmonary vasculature [4]. In our patient, a concurrent PDA amplified pulmonary blood flow independently predisposes to pulmonary vascular disease through abnormal vascular development and altered nitric oxide signaling [3]. This convergence of multiple insults may explain the severity of supra-systemic PAH at four months of age, though the relative contribution of each mechanism remains difficult to quantify with certainty.

Diagnosis:

The PDV was not detected on neonatal abdominal ultrasound, highlighting the diagnostic delay characteristic of this condition [2]. CT angiography, prompted by unexplained supra-systemic PAH, provided definitive anatomical characterization. Clinicians should include congenital portosystemic shunting in the differential diagnosis of unexplained PAH in infancy, particularly when associated with mild transaminase elevation or direct hyperbilirubinemia. Formal cardiac catheterization to measure pulmonary vascular resistance and assess vasoreactivity are recommended before surgical closure in severe PAH [5] was not performed, representing a limitation in preoperative risk stratification.

Perioperative hemodynamic instability. The major anesthetic lesson of this case is that ductus venosus clampage is not a simple surgical step but a high-risk hemodynamic intervention. Acute clamp occlusion may simultaneously increase portal pressure, suppress direct caval venous return, reduce right ventricular preload, and expose an already failing right ventricle to a sudden afterload-preload mismatch. In supra-systemic PAH, the right ventricle operates on a narrow reserve: any increase in pulmonary vascular resistance or any reduction in coronary perfusion pressure may trigger RV ischemia, RV dilation and septal displacement. The left ventricle then becomes underfilled because of both reduced venous return and ventricular interdependence, resulting in profound systemic hypotension. The bradycardia and collapse observed during the first clamp test are consistent with the proposed mechanism of acute RV decompensation and abrupt RV-LV interaction though isolated hypovolemia cannot be fully excluded as a contributing factor. In our patient, this physiopathological understanding directly informed the subsequent clinical strategy: after the first failed clamp test, a progressive re-clamping approach was adopted, with iterative pauses to assess hemodynamic tolerance, and without portal pressure measurement. This graded approach, combined with vasopressor titration and close surgeon-anesthesiologist communication, ultimately allowed surgical closure to be completed. These observations support the general principles of graded temporary occlusion, direct verbal coordination before each clamp, immediate release in case of intolerance, invasive monitoring whenever feasible, and pre-prepared vasoactive and pulmonary vasodilator rescue therapy [12-16].

Hospital-acquired pneumonia and ARDS:

HAP developing after 48 hours of hospitalization represents one of the most common and lethal healthcare-associated infections in critically ill pediatric patients [11]. In this infant, multiple risk factors coexisted: prolonged intubation, failed extubation with aspiration risk, trisomy 21-associated immune dysregulation, and severe underlying cardiopulmonary compromise. The resulting ARDS, meeting PALICC criteria [8], was refractory to lung-protective ventilation. ECMO could have served as a bridge to recovery [9], but was unavailable a preventable factor in the fatal outcome. This underscores the need for ECMO access at centers that manage high-risk pediatric surgical patients.

CONCLUSIONS

PDV is a rare but potentially fatal congenital vascular anomaly that can present in infancy with life-threatening supra-systemic PAH. Surgical closure carries a high risk of acute hemodynamic instability and must be supported by comprehensive preoperative hemodynamic characterization, invasive monitoring, and ready access to vasoactive drugs. Postoperative vulnerability to hospital-acquired infections and secondary ARDS is high in these patients, particularly in those with trisomy 21 or other immune-compromising comorbidities. Access to ECMO as a rescue strategy should be planned preoperatively. Earlier diagnosis and intervention, before the establishment of fixed pulmonary vascular disease, may improve long-term outcomes.

Ethics statement:

Written informed parental consent was obtained for publication of this case report. No formal ethics committee approval is required at our institution for single case reports of standard clinical care.

Conflicts of interest: The authors declare no conflicts of interest.

Funding: No funding was received.

Reporting guideline: This case report follows the CARE 2013 Checklist.[6]

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