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CASE REPORT

BRAIN BLEED IN A BLUE BABY: A DORV CASE UNDERGOING EMERGENCY CRANIOTOMY

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ABSTRACT

Background: Cyanotic congenital heart disease (CCHD) poses significant perioperative challenges, particularly during emergency neurosurgical procedures. Double outlet right ventricle (DORV) is a rare cyanotic congenital heart disease that complicates anesthetic management due to chronic hypoxemia and right-to-left shunting. **Case Presentation:** We report the case of a 5-year-old boy with uncorrected DORV who sustained severe traumatic brain injury after a fall. Head CT revealed a large right temporoparietooccipital epidural hematoma with subdural and subarachnoid hemorrhage and a 1.7 cm midline shift. Emergency craniotomy was performed under general anesthesia with invasive hemodynamic monitoring and cautious ventilatory management to balance cerebral and cardiac demands. Although intraoperative hemodynamics remained stable and the hematoma evacuation was technically successful, the patient developed persistent postoperative hypoxemia and multiorgan failure, resulting in death on postoperative day three.

Despite a technically successful evacuation of the hematoma and stable intraoperative parameters, the patient's postoperative course was complicated by persistent hypoxemia due to right-to-left shunting, culminating in multiorgan failure and death after three days of intensive care. This case underscores the profound anesthetic and surgical challenges posed by the coexistence of severe traumatic brain injury and uncorrected cyanotic congenital heart disease, highlighting the importance of multidisciplinary collaboration, tailored perioperative strategies, and the urgent need for early cardiac corrective interventions to improve outcomes in similar high-risk scenarios. **Conclusion:**This case demonstrates the profound anesthetic and surgical challenges of emergency neurosurgery in pediatric patients with uncorrected cyanotic congenital heart disease. Effective management requires multidisciplinary collaboration, individualized anesthetic strategies, and early cardiac correction to improve survival in such high-risk scenarios.

Keywords**:** Cyanotic congenital heart disease, Double outlet right ventricle, Traumatic brain injury, Pediatric anesthesia, Emergency craniotomy.

INTRODUCTION

Cyanotic congenital heart disease (CCHD) represents a group of complex structural heart defects characterized by the admixture of oxygenated and deoxygenated blood, leading to chronic hypoxemia. These conditions are associated with significant morbidity and mortality, particularly in children who remain uncorrected. The physiological burden of sustained hypoxemia manifests not only as impaired growth and development but also as chronic systemic adaptations such as polycythemia, hyperviscosity, and endothelial dysfunction. As a result, patients with CCHD have limited physiologic reserve, making them highly vulnerable in the context of additional systemic insults such as trauma or infection.

Among the spectrum of CCHD, double outlet right ventricle (DORV) is a relatively rare entity in which both the aorta and pulmonary artery arise predominantly from the right ventricle. The clinical presentation of DORV is heterogeneous and largely determined by the location of the ventricular septal defect (VSD), the presence of pulmonary stenosis, and the balance of systemic versus pulmonary blood flow[1]. In patients with uncorrected DORV, chronic

hypoxemia is common, and their baseline hemodynamic status is already precarious. This creates an especially high-risk environment when they present for urgent or emergent surgical procedures, as anesthetic and perioperative management must balance the competing demands of systemic and pulmonary circulation[2].

Traumatic brain injury (TBI) remains one of the leading causes of morbidity and mortality in pediatric populations worldwide[3]. Children are particularly susceptible to head trauma due to their high activity levels and relative vulnerability to falls. Severe TBI, particularly when associated with space-occupying lesions such as epidural or subdural hematomas, requires urgent neurosurgical intervention to prevent irreversible neurological damage and death. In such cases, rapid surgical decompression is the cornerstone of management, often leaving little time to optimize preoperative status. When superimposed on a background of complex congenital heart disease, the urgency of neurosurgical management must be weighed carefully against the elevated perioperative risk.

The perioperative care of children with CCHD undergoing neurosurgery is particularly challenging[4]. Anesthetic management must consider the delicate balance of systemic vascular resistance, pulmonary vascular resistance, oxygen delivery, and cerebral perfusion. Even small perturbations in ventilation, oxygenation, or circulation may result in profound hemodynamic instability. In addition, coagulation abnormalities frequently observed in cyanotic heart disease further complicate surgical planning, as patients may have an increased risk of both bleeding and thrombosis[5]. These challenges require careful planning, invasive monitoring, and a multidisciplinary approach involving cardiology, anesthesiology, and neurosurgery.

The interaction between uncorrected cyanotic heart disease and severe TBI represents a rarely encountered but highly complex clinical scenario[6]. While each condition independently poses substantial management difficulties, their coexistence creates a compounding effect in which the risks of hemodynamic collapse, poor cerebral oxygenation, and perioperative mortality are markedly increased. The literature addressing the simultaneous management of these conditions remains sparse, with only a limited number of case reports describing outcomes in similar patients[7]. As such, each report adds valuable insight into the principles and pitfalls of perioperative care in this vulnerable population.

In low- and middle-income countries, where access to early corrective cardiac surgery may be limited, the number of children surviving into later childhood with uncorrected CCHD remains significant. These patients are therefore at risk of presenting with unrelated acute surgical emergencies such as trauma[8]. This reality underscores the importance of developing anesthetic and surgical strategies tailored to the needs of cyanotic heart disease patients undergoing non-cardiac surgery. Such strategies must account not only for the unique cardiovascular physiology but also for the heightened risk of complications such as hypoxemia, acidosis, arrhythmia, and circulatory failure[9].

This case report presents the perioperative course of a 5-year-old child with uncorrected DORV who sustained severe traumatic brain injury following a fall. The case highlights the anesthetic challenges, intraoperative considerations, and postoperative complications encountered in managing such a critically ill patient. Through this report, we aim to contribute to the limited body of literature on the perioperative management of uncorrected CCHD in the setting of emergency neurosurgical intervention, providing insights that may help guide clinicians faced with similarly complex scenarios[10].

CASE PRESENTATION

A 5-year-old male patient, weighing 14 kg, with a history of uncorrected cyanotic congenital heart disease (double outlet right ventricle, DORV), was admitted following a fall that resulted in head trauma[11]. Initially, the patient presented with vomiting, headache, and seizures, which were soon followed by a progressive decline in consciousness. On arrival, his level of responsiveness was markedly reduced, with a Glasgow Coma Scale (GCS) of 2-2-4[12]. He had already been intubated for airway protection. Baseline vital signs showed blood pressure of 102/64 mmHg, heart rate of 78 beats per minute, and oxygen saturation of 87%. Physical examination revealed digital clubbing, consistent with chronic cyanosis[13].

Figure 1. Non-contrast head CT scan showing a large right temporoparietal epidural hematoma (EDH)

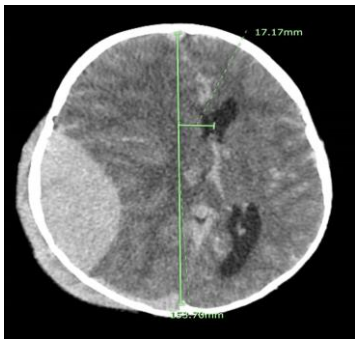

Figure 2. Axial CT scan demonstrating a significant midline shift of 1.7 cm to the left, indicating raised intracranial pressure

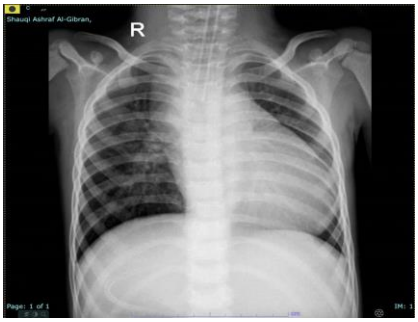

Figure 3. Chest X-ray showing cardiomegaly

Emergency craniotomy was performed under general anesthesia with sevoflurane, fentanyl, and rocuronium. Invasive arterial blood pressure monitoring was established, and serial blood gas analyses were conducted to ensure the absence of acidosis. Ventilation was carefully managed to avoid hyperventilation and the use of positive end-expiratory pressure (PEEP), with the goal of minimizing further increases in intracranial pressure[14]. The duration of surgery was 1 hour and 30 minutes. Intraoperatively, a total of 200 mL of clot and 180 mL of active bleeding were evacuated. Blood loss was replaced with 160 mL of packed red cells and 200 mL of Ringer's Lactate, while urine output was 200 mL. Throughout the procedure, no intraoperative complications occurred, and hemodynamics were supported with dopamine infusion at 5 mcg/kg/min[15].

Postoperatively, the patient remained intubated and was transferred to the intensive care unit (ICU) for continued management[16]. Initial postoperative vital signs showed blood pressure of 108/74 mmHg, heart rate of 109 beats per minute, and oxygen saturation of 70%, reflecting persistent hypoxemia due to right-to-left shunting despite stable systemic hemodynamics. The patient required mechanical ventilation for three days and ongoing dopamine infusion. Despite intensive monitoring and supportive measures, his condition progressively deteriorated. Persistent severe hypoxemia, combined with the extent of his brain injury and limited cardiopulmonary reserve, ultimately led to multiorgan failure. The patient passed away during his ICU stay.

DISCUSSION

The coexistence of severe traumatic brain injury and uncorrected cyanotic congenital heart disease (CCHD) presents one of the most complex challenges in perioperative and critical care medicine. In this case, the child with double outlet right ventricle (DORV) presented after a fall resulting in extensive intracranial hemorrhage and significant midline shift[17]. The urgent need for surgical decompression was evident to prevent further neurological deterioration, yet the underlying cardiac condition created an extremely narrow margin for safe anesthetic and surgical intervention. Chronic hypoxemia, right-to-left shunting, and limited cardiopulmonary reserve placed the patient at inherently higher risk, complicating efforts to maintain adequate oxygen delivery and hemodynamic stability during and after the neurosurgical procedure. From an anesthetic perspective, the management of such a case required careful balancing of cerebral and cardiac goals. The primary objectives included preserving cerebral perfusion pressure, minimizing intracranial pressure, and avoiding increases in pulmonary vascular resistance (PVR) that could worsen right-to-left shunting [18]. Strategies such as avoiding positive end-expiratory pressure (PEEP) and preventing hyperventilation were crucial to prevent intracranial and cardiopulmonary compromise. Intraoperatively, the team successfully maintained stable hemodynamics with invasive arterial monitoring and careful fluid and transfusion management, while also ensuring the absence of metabolic acidosis through frequent arterial blood gas analysis. The procedure was technically successful, with hematoma evacuation and no immediate intraoperative complications reported.

Despite these meticulous efforts, the postoperative course was dominated by persistent hypoxemia, with oxygen saturations remaining critically low despite stable systemic blood pressure and cardiac output supported by dopamine infusion. This reflected the physiological limitations imposed by the child's congenital heart disease. In cyanotic CHD, right-to-left shunting allows deoxygenated blood to bypass pulmonary circulation, rendering mechanical ventilation and supplemental oxygen less effective[19]. This physiological barrier highlights why

perioperative management in such cases is not only technically complex but also fundamentally constrained by the underlying cardiac pathology, regardless of optimal anesthetic and surgical strategies.

The clinical trajectory in this case also illustrates the cumulative effect of chronic hypoxemia on systemic resilience. Children with uncorrected CCHD often exhibit polycythemia, increased blood viscosity, and reduced organ reserve, factors that magnify vulnerability to secondary insults such as traumatic brain injury[20]. Prolonged hypoxemia and impaired oxygen delivery exacerbate neuronal injury, and even with timely neurosurgical intervention, the potential for meaningful recovery remains limited. In this case, despite three days of intensive ventilatory and hemodynamic support, the persistent imbalance between oxygen demand and delivery led to progressive multiorgan dysfunction, culminating in the patient's death[21].

This case underscores several critical lessons. First, it emphasizes the need for multidisciplinary collaboration between neurosurgeons, anesthesiologists, cardiologists, and intensivists in managing patients with complex cardiac physiology requiring emergency surgery. Second, it highlights the importance of preventive strategies, including earlier corrective or palliative interventions for congenital heart disease, to reduce vulnerability to catastrophic outcomes. Finally, it contributes to the growing but still scarce literature on the management of traumatic neurosurgical emergencies in the setting of uncorrected cyanotic CHD [22] . By reporting and analyzing such rare but challenging cases, clinicians can better understand perioperative priorities, refine management strategies, and ultimately work toward improving survival and outcomes in this highly vulnerable patient population. To our knowledge, very few reports in the literature have described the perioperative management of emergency craniotomy in pediatric patients with uncorrected cyanotic congenital heart disease[23]. Most existing publications focus on elective procedures or palliative cardiac interventions, whereas the intersection of acute neurosurgical emergencies and complex congenital heart physiology remains sparsely documented. This case, therefore, provides a unique contribution by highlighting the physiological constraints that limit oxygen delivery despite technically successful neurosurgical intervention and advanced perioperative monitoring[24]. The unfavorable outcome, despite optimal intraoperative and intensive care measures, underscores the profound challenges inherent in this clinical scenario and emphasizes the importance of early corrective cardiac interventions and preventive strategies. By adding to the limited pool of reported experiences, this case reinforces the need for heightened awareness, meticulous multidisciplinary planning, and development of tailored guidelines for managing neurosurgical emergencies in patients with cyanotic congenital heart disease.

CONCLUSION

This case highlights the extreme challenges of managing emergency neurosurgical intervention in a pediatric patient with uncorrected cyanotic congenital heart disease. The combination of severe traumatic brain injury and complex cardiac physiology created a narrow therapeutic window, where interventions to optimize cerebral protection could exacerbate hypoxemia, and measures to improve oxygenation risked worsening intracranial hypertension. Despite careful anesthetic and surgical management, the patient's outcome was ultimately unfavorable, underscoring the importance of early recognition, multidisciplinary coordination, and the need for tailored perioperative strategies in this high-risk population. Documentation of such cases adds to the limited body of literature and may help guide future clinical decision-making for similar complex scenarios.

DECLARATIONS

Ethical approval

Ethical approval for publication of this case was obtained in accordance with institutional policy.

Patient consent

Written informed consent was obtained from the patient's guardian for publication of this case report and any accompanying images. All reasonable efforts were made to protect patient anonymity.

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Conflict of interest

The authors declare no conflicts of interest related to this work.

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