CASE REPORT

MANAGING EMERGENCY NON-CARDIAC SURGERY IN A PEDIATRIC PATIENT WITH MOST SEVERE CYANOTIC HEART DISEASE: A CASE STUDY

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ABSTRACT

Anesthesia for congenital heart disease patients undergoing noncardiac emergency surgery presents significant challenges due to complex anatomical and physiological abnormalities, requiring tailored interventions to maintain hemodynamic stability. Anesthetists need to be knowledgeable not only about the normal series cardiac circulation but also the parallel (or balanced) and single-ventricle circulations. Providing anesthesia to pediatric patients with congenital heart anomalies during procedures unrelated to the heart involves a range of specialized considerations and remains a complex aspect of perioperative care. Multiple factors contribute to the complexity of anesthetic management in children with congenital heart disease, such as the patient's age, nature and extent of the cardiac abnormality, hemodynamic compensation, surgical urgency, and any coexisting health issues. This case discusses the anesthetic management of an 8-year-old boy with congenital heart disease and severe pulmonary hypertension undergoing emergency laparotomy. It addresses perioperative challenges and provides an overview of pediatric pulmonary hypertension physiology, risk stratification, and intraoperative considerations.

Keywords:

Cyanotic Heart Disease, Non-Cardiac Surgery, Pediatric Surgery

INTRODUCTION

Congenital heart disease (CHD) occurs in approximately 0.6% of newborns, with its incidence remaining stable over time.[1] Improvements in surgical and medical treatments have primarily shifted mortality to adulthood. The population of adults with CHD is steadily growing, with the exception of those with Eisenmenger syndrome and unrepaired cyanotic defects.[2] Pediatric pulmonary hypertension is a multifaceted condition with a range of causes, affecting everyone from premature infants to young adults. The most commonly addressed form is pediatric pulmonary arterial hypertension, whether idiopathic or associated with congenital heart disease, due to its progressive and often fatal course.[3] In individuals with complex congenital heart defects, such as pulmonary atresia-ventricular septal defect and single-ventricle anomalies, pulmonary hypertension may affect up to 49.7% by the age of 40.[4] Pulmonary hypertension (PH) in congenital heart disease (CHD) with biventricular physiology is diagnosed when the mean pulmonary artery pressure exceeds 25 mmHg and pulmonary vascular resistance exceeds 3 Wood units. Anesthesia for CHD patients undergoing non-cardiac surgery should be individualized, with induction and maintenance techniques adapted to the child's unique condition and the specifics of the procedure.

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Case Report

An 8-year-old boy (weight: 20 kg, height: 114 cm, BMI: 15.39 kg/m²) presented with a 3-day history of abdominal pain and bilious vomiting (8 episodes in one day). His medical history included cyanotic congenital heart disease (CHD) with bilateral Glenn shunts inserted 2.5 years ago, which remained functional as per recent echocardiography findings. The echo showed a single functioning right ventricle (RV), single atrium, moderate tricuspid regurgitation (TR), a non-functional compressed left ventricle (LV), pulmonary artery pressure (PAP) of 74 mmHg, and a double outlet RV. His medications included sildenafil (50 mg twice daily) and bosentan (62.5 mg twice daily).

The patient's baseline peripheral oxygen saturation

(SpO_2) was 56% on room air. He had clubbed fingers and secondary erythrocytosis (hemoglobin: 192 g/L, hematocrit: 0.68) with mild leukocytosis (10.65×10^9/L) and elevated C-reactive protein (42.9 mg/L). Abdominal ultrasound revealed a heterogeneous lesion adjacent to the small bowel, cystitis, and bilateral hydroureter. A contrast-enhanced CT scan confirmed small bowel torsion, suggesting malrotation and volvulus, necessitating emergency surgical intervention.

The patient underwent preoperative counseling for high-risk non-cardiac abdominal emergency surgery (ASA status IV-E). High risk consent was taken from the parents. In the operating room, standard anesthesia monitoring (ECG, pulse oximetry (pre ductal & Post ductal), non-invasive BP, temperature) was initiated, and an arterial line was inserted before induction. Vital signs included a temperature of 36.5°C, heart rate of 86 beats/min, BP of 100/69 mmHg, and respiratory rate of 19 breaths/min. Pre-induction SpO_2 ranged between 56-65%. Arterial blood gas analysis revealed a pre-induction partial pressure of oxygen (PaO_2) of 48 mmHg.

Anesthesia induction included pre medication with midazolam 0.05mg/kg (1 mg), analgesia with ketamine 0.25mg/kg (5 mg) and inf Acetaminophen 15mg/kg for analgesia, induction with inj propofol 2mg/kg (40 mg), and muscle relaxant cis-atracurium 0.15mg/kg (3 mg). Intubation was performed with a size 5.5 cuffed endotracheal tube confirmed with bilateral auscultation and capnography. Intraoperative anesthesia was maintained with sevoflurane and 100% oxygen. Cis-atracurium was administered as needed by monitoring train of four via Nerve Stimulator, and fluid management included 1 liter of Ringer's lactate after calculating (NPO deficit, maintenance fluid, intra opt blood loss and 3rd space loss). The patient remained hemodynamically stable with an estimated blood loss of 200 mL, except for an arrhythmia managed with lignocaine (1 mg/kg). Intraoperative PaO_2 was 50 mmHg.

Surgical findings included splenic torsion, malrotation of the gut, and a congested spleen, necessitating splenectomy and widening of the small bowel mesentery. The procedure lasted 2.5 hours, after which the patient was shifted to the surgical ICU on spontaneous ventilation with FiO_2 100%.

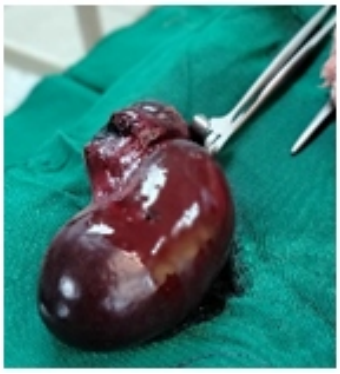

Figure 1: Showing the spleen that was removed

Table I: Intra-Op Vital Monitoring

Time	HR (bpm)	IBP(MAP) (mmhg)	SpO_2(%) FiO_2 =100	Temp °C	Urine output (ml) (0.5-1ml/kg/hr)	BSR	ABGs PaO_2 (%)
00 min	86	78	56%	36.5 °C	-		47.9%
15 min	88	77	60%	36.5 °C	-		
30 min	90	80	89%	37 °C	30ml	100mg/dl	
45 min	90	78	88%	36.5 °C	-		
60 min	101	76	88%	37.5 °C	40ml		50%
75	100	78	90%	36.5 °C	-		
90 min	104	74	90%	37 °C	-		
105	95	79	91%	36.5 °C	-		
120 min	92	65	93%	37 °C	85ml		
135 min	95	70	90%	36.5 °C	-		
150 min	110	60	89%	37.5 °C	90ml	120mg/dl	
SICU-	115	60	80%	37.5 °C		101mg/dl	

Postoperatively, sedation with dexmedetomidine, starting with a loading dose of 0.5 µg/kg followed by a maintenance infusion of 0.1 µg/kg/hr, titrated to maintain a MAP > 60 mmHg, pain management via caudal analgesia in immediate post opt period, and antibiotics (meropenem 400 mg and vancomycin 400 mg every 8 hours) were provided. Pulmonary hypertension medications (sildenafil and bosentan) were continued via nasogastric tube. The patient was successfully extubated after 24 hours with successful weaning trial, monitored for 72 hours, and discharged from the ICU in stable condition.

DISCUSSION

Advances in surgical techniques have enhanced survival rates for patients with univentricular physiology. As life expectancy increases, these patients may now present for non-cardiac surgeries.

The Glenn shunt is surgical interventions primarily used to treat congenital heart defects in individuals with single ventricle physiology. This procedure creates a shunt that connects the superior vena cava (SVC) to the pulmonary arteries, enhancing blood flow to the lungs and managing conditions where the heart's pumping ability is compromised. This arrangement allows deoxygenated blood from the upper body to reach the lungs for oxygenation directly, bypassing the heart.

Anesthetic management in patients with single-chamber hearts is particularly complex due to the risks associated with altered hemodynamics. The Glenn procedure facilitates drainage of the superior vena cava directly into the pulmonary arteries. This surgery significantly changes physiological dynamics and require thorough preoperative assessment and planning.

It is commonly accepted that patients with congenital heart disease undergoing noncardiac procedures are at a higher risk of perioperative cardiac arrest, major complications, and mortality compared to those without congenital heart disease. Children and adults with complex congenital heart disease undergoing noncardiac surgery are at an increased risk of complications and mortality during the perioperative period. Key factors influencing this risk include the type and urgency of the procedure, as well as any underlying comorbidities.[5]

Patients with complex congenital heart defects undergoing noncardiac surgery are more vulnerable to perioperative complications, particularly when risk factors such as baseline cyanosis, signs of congestive heart failure, and poor overall health are present.[6] Consequently, these patients necessitate detailed preoperative assessments, a deep understanding of their intricate physiology, and a collaborative care team to ensure optimal perioperative management and outcomes.

In cases like this, it is crucial to monitor cardiac output and ensure stable hemodynamics. Hemodynamic monitoring and appropriate fluid management are vital in optimizing outcomes for patients with single ventricle physiology. We employed continuous invasive monitoring to evaluate cardiac function during the procedure, allowing for the quick identification and correction of any deviations from baseline hemodynamics.

Considering the likelihood of pulmonary complications in individuals with congenital heart defects, we chose controlled ventilation to reduce intrathoracic pressure changes that could hinder venous return. Recent literature suggests that a balance of risk applies when targeting higher minute ventilation for carbon dioxide (CO_2) clearance, aiming to achieve normal pCO_2 and pH values, despite the necessary increase in the intensity of mechanical ventilation.[7] Utilizing low tidal volumes and suitable respiratory rates facilitated optimal oxygenation while reducing the risk of increase intrathoracic pressure and decrease CO.

Insufficient anesthesia or poor pain management can increase systemic vascular resistance (SVR), worsening left-to-right shunting as blood is diverted from the systemic circulation, which may ultimately impair cardiac output. Likewise, reduced cardiac output can result from decreases in venous return due to systemic hypotension or increased intrathoracic pressures.[8] Therefore, in our case, we administered Acetaminophen and ketamine for intraoperative pain relief to minimize the severe side effects linked to opioids, such as respiratory depression, sedation, nausea, vomiting, and delayed mobilization. A caudal epidural was given postoperatively to decrease opioid consumption and prevent respiratory depression as a side effect. Regional analgesia may also serve as a respiratory stimulant and is linked to a decreased need for mechanical ventilation.[9]

Postoperatively, our patient exhibited stable vital signs and satisfactory urine output, suggesting preserved renal function - an important factor, particularly in individuals with compromised cardiac output. Literature emphasizes that effective postoperative monitoring and

management can significantly reduce morbidity in this population.[10] Our patient was closely monitored in a Surgical intensive care unit (SICU) to manage any potential complications, such as arrhythmias or fluid overload.

CONCLUSION

In conclusion, anesthetic management for children with congenital heart defects, especially those with single-chamber physiology, demands careful planning and execution. The positive outcome of this case highlights the significance of personalized anesthetic strategies and collaborative teamwork. Future research should aim to explore long-term outcomes for this patient population as our understanding of congenital heart disease management advances.

CONFLICT OF INTERESTS

None

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ETHICAL STATEMENT

Ethical approval was obtained from Fauji Foundation Hospital Rawalpindi wide letter no. 877/RC/FFH/RWP dated 01/10/2024).

Authors' contributions:

Liaquat Ali: Conception of study/Designing/Planning, Critical Review, Material Analysis

Saima Zia: Experimentation/Study Conduction, Analysis/Interpretation/Discussion, Manuscript Writing

Huma Hanif: Experimentation/Study Conduction, Analysis/Interpretation/Discussion, Manuscript Writing

Umer Ali: Critical Review

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