# Perioperative anaesthetic management of a patient with single ventricle undergoing mastoidectomy

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#### **ABSTRACT**

Patients with single ventricles undergoing non-cardiac surgery have a higher incidence of perioperative complications. Factors associated with the most significant risk are the complexity of cardiac disease and the patient's physiological status. We here describe the anaesthetic management and pathophysiological considerations of an 18-year-old male with a single ventricle undergoing elective mastoidectomy. Comprehensive and multidisciplinary perioperative care is mandatory for optimal management. The patient had a history of previous cardiac surgeries for his cardiac condition and presented for elective mastoidectomy due to chronic suppurative otitis media. Preoperative assessment revealed a satisfactory general condition with peripheral oxygen saturation (Spo2) of 90-92% on room air. Cardiovascular evaluation showed weak S1 and S2 heart sounds and a faint systolic murmur heard best at the left upper sternal border. Echocardiography indicated a common ventricle with a small left atrium and significant tricuspid regurgitation. Long term medications included carbamazepine, warfarin, and aspirin. Comprehensive perioperative care, including meticulous preoperative assessment, tailored intraoperative management, and appropriate postoperative monitoring is crucial for patients with single ventricles undergoing non-cardiac surgery. Specialized centres equipped to manage high-risk cases are recommended to optimize outcomes and minimize complications.

**Keywords:** anaesthesia, cardiac anaesthesia, congenital heart diseases, single ventricle

# Introduction

Among all congenital heart defects, the incidence of hypoplastic left heart syndrome (HLHS) is 3.8% and accounts for 23% of mortality within the first week of life.<sup>[1,2]</sup> It is characterized by varying degrees of hypoplastic left ventricle accompanied by a single right ventricle and different valvular abnormalities involving either aortic stenosis or aortic atresia and severe mitral stenosis or mitral atresia.<sup>[3]</sup>

HLHS impairs normal circulation physiology, mixing pulmonary and systemic blood in the typical single right ventricle, which directly communicates with the pulmonary circulation and the systemic circulation through the patent ductus arteriosus (PDA). The PDA must be maintained after birth with an infusion of prostaglandin E1 to maintain systemic circulation perfusion until surgical interventions are planned. [4] Maintaining a balance between pulmonary and systemic blood flow provides the optimal state to maintain adequate perfusion of both the lungs and body perfusion.

# **Case Report**

An 18-year-old male who is known to have a single ventricle heart underwent two heart surgeries in which a ventricular repair and reconstruction were done, including repair of the aortic arch and pulmonary artery bandage at three months of age and Fontan operation at age five years. He had two months history of recurrent focal convulsions secondary to his ear disease as stated by his neurologist and on treatment with carbamazepine.

In his current admission for elective surgery, the patient presented with three years history of complaints consistent with chronic suppurative otitis media (CSOM) not responding to medical treatment and was planned for elective mastoidectomy.

The preoperative assessment was thorough, with input from the cardiology team and adherence to the 2022 European Society of Cardiology guidelines for assessing cardiovascular health in patients with congenital heart disease undergoing non-cardiac procedures. It identified an intermediate risk profile. Of particular concern was right heart function, notably tricuspid regurgitation, a significant risk factor for single ventricle patients undergoing non-cardiac surgery. Additionally, findings indicated a dilated right ventricle and atrium, suggesting a pulmonary artery pressure of 30.<sup>[9, 10]</sup>

On clinical examination a satisfactory general condition was noticed, with peripheral pulse oximetry saturation (Spo2) of 90-92% on room air. Respiratory rate 18/min, breathing pattern, and bilateral breath sounds were normal.

Heart sounds S1 and S2 were weak with a faint systolic murmur heard at the left upper sternal border. Echocardiography showed satisfactory aortic flow, a common ventricle with a small left atrium, and a dilated dominant right atrium, resulting in significant tricuspid regurgitation. A pulmonary artery band was noted in situ with a pulmonary pressure of 30 mmHg; the estimated ventricular ejection fraction was 60%.

The patient's long-term medications were carbamazepine

200mg twice daily for control of seizures, in addition to aspirin 75mg and warfarin 3mg started earlier by his cardiology team. Warfarin had been replaced with low molecular weight heparin 5 days preoperatively.

Routine investigations were performed in addition to the renal profile, liver profile, bleeding profile, viral screening, and CXR. All results were within normal ranges. International Normalised Ratio (INR) of 1.5 and haemoglobin of 15.4g/dl.

# Intraoperative anaesthetic management

The objectives of intraoperative management are to maintain cardiac contractility, balancing systemic and pulmonary vascular resistance, preventing dysrhythmias, and optimizing oxygen saturation. Intravenous ceftriaxone 2gm as prophylaxis for infective endocarditis was given one hour before the operation. The case was scheduled early in the morning as first case on the list. The patient was connected to standard monitoring in the form of an ECG, pulse oximeter, non-invasive blood pressure monitoring, and capnography. The optimal target was set close to his baseline reading with a mean arterial pressure of 80-100mmHg. Spo2 not less than 92%, and a pulse rate of 80-100/minute. Induction was achieved with 100micrograms of fentanyl, 80mg of 2% intravenous lidocaine, 5mg midazolam, 4mg dexamethasone, 1g paracetamol, and inhalational anaesthetic sevoflurane via mask ventilation.

Muscle relaxation was done using 40mg atracurium, a non-depolarising muscle relaxant followed by endotracheal tube placement. Maintenance of anaesthesia was achieved by using sevoflurane 0.8%, fluid therapy was restricted to a total intraoperative fluid volume of 350mls. Nitrous oxide was avoided in all stages of anaesthesia. Three litres per minute of oxygen and 5 litres of air were used throughout the operation and titrated to achieve a target Spo2.

The recovery phase was managed with extubation during deep sedation to reduce the risk of sympathetic overstimulation. The medications given were 2.5 mg neostigmine, 30 mg of 2% intravenous lidocaine, and 0.5 mg atropine. The total time of surgery was one and half hours.

Postoperatively, the patient was taken to the Intensive Care Unit for monitoring and transferred to the ward after 24 hours.

# **Discussion**

Single ventricle patients have survived to older ages

recently due to advancements in cardiac surgery, so cases do present occasionally for various non-cardiac procedures under general anaesthesia. Many studies have shown a higher rate of intra-operative and early postoperative adverse events.<sup>[4]</sup>

Assessment for perioperative risk of complications and adverse events is essential in determining the level of care required and the need for referral to specialized centres.

Many studies have identified factors associated with increased risk of complications in patients with single ventricles. The high-risk factors linked with complications are cardiac disease complexity, current physiological status, type of planned non-cardiac surgery, and age. As a result, single ventricle patients undergoing major surgery with predominant features such as decompensating heart failure, dysrhythmias, or pulmonary hypertension are at high risk of complications. Well-equipped specialized centres are required to manage elective cases.

Patients with single ventricles differ in clinical condition, severity, type of reconstructive surgery performed, and associated anomalies. As such, no one absolute anaesthetic technique or medication has been established as the best option. However, we used sevoflurane which has rapid recovery and a minimal or no effect on heart rate in induction as a desirable option. Fentanyl was the opioid agent used. However, a shorter agent such as alfentanyl would be superior when available.

Optimal intraoperative and postoperative pain management is a crucial factor that must be considered. Opioids and patient-controlled analgesia for major operations have been used as a primary intervention in major surgeries. [5]

# Recommendations

**Specialized Care:** Patients with single ventricles undergoing major surgery should be managed in well-equipped specialized centres with expertise in cardiac anaesthesia and intensive care.

**Multidisciplinary Approach:** A multidisciplinary team involving anaesthesiologists, cardiologists, cardiac surgeons, and other specialists to develop and implement comprehensive perioperative care plans tailored to the individual patient's needs.

**Preoperative Evaluation:** Thorough preoperative assessment, including detailed cardiac evaluation, optimization of medical therapy, and identification of potential risk factors, are essential for risk stratification

and optimal perioperative management.

**Intraoperative Management:** Anaesthetic techniques should focus on maintaining haemodynamic stability, optimizing oxygenation, and minimizing stress responses. Careful monitoring and titration of anaesthetic agents, fluid management, and ventilation strategies are key components.

**Postoperative Care:** Close postoperative monitoring in a critical care setting is essential to detect and promptly manage any complications. Adequate pain management and early mobilization strategies should be implemented to facilitate recovery and reduce the risk of postoperative complications.

Long-term Follow-up: Patients with single ventricles require long-term follow-up to monitor for late complications, assess cardiac function, and optimize medical management. Collaboration between cardiology specialists and primary care providers is essential for ensuring continuity of care and optimizing long-term outcomes.

# Conclusion

The successful management of patients with single ventricles undergoing non-cardiac surgery relies on comprehensive perioperative strategies tailored to individual patient needs. Specialized centres with expertise in cardiac anaesthesia and intensive care play a pivotal role in optimizing outcomes and minimizing complications.

Moreover, ongoing research and collaboration among multidisciplinary teams are crucial for advancing the field and improving patient care. By staying updated on the latest advancements in anaesthetic techniques, perioperative monitoring, and surgical interventions, healthcare providers can continue to enhance the quality of care for this complex patient population.

# Authors' contributions

MAH: Conceptualization, Writing – Review and Editing, Project administration. EM: Supervision, Conceptualization, Validation. EAM and WAM: Resources, Writing - Review and Editing.

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