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Anaesthetic Management of Congenital Cyanotic Heart Disease in a Robotic-Assisted Pheochromocytoma Excision: A Rare Case Study of Preload-Dependent Physiology

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Abstract

Background and Case: This case study examines the anaesthesia management of a 22-year-old female with congenital cyanotic heart disease (CCHD) and a hyper-vascular adrenal lesion suggestive of pheochromocytoma, who underwent robotic-assisted surgery. The patient's complex history , mixing lesion including single ventricle physiology with and reduced pulmonary blood flow, presented significant perioperative challenges such as avoiding hyperoxemia ,hypercarbia , avoiding hemodynamic pertubations, which may increase risk of overflow in shunt and thrombosis.

Conclusion: An effective anaesthetic approach was crucial, involving a multidisciplinary team to ensure hemodynamic stability and address the unique risks associated with robotic techniques. Strategies included meticulous fluid management, tailored medication regimens for blood pressure control, and advanced monitoring throughout the procedure. Postoperatively, vigilant management of analgesia and fluid was essential to prevent post operative pulmonary complications

Keywords: Anaesthesia, Congenital Heart Disease, Pheochromocytoma, Perioperative Management, Robotic Surgery, Metanephrines

Introduction

The perioperative management of patients with congenital cyanotic heart disease (CCHD) undergoing complex surgeries poses unique challenges due to their altered cardiovascular physiology [1]. When these patients require surgery for pheochromocytoma, a rare neuroendocrine tumor, the complexity increases significantly. Pheochromocytomas (PHEO) secrete excessive amounts of catecholamines, which lead to episodic or sustained hypertension, tachycardia, and arrhythmias, adding significant stress to an already compromised cardiovascular system [2]. This case study highlights the anaesthetic management of a patient with CCHD, who presents with preload-dependent physiology, during robotic-assisted pheochromocytoma excision. The combination of CCHD, hemodynamic instability induced by the pheochromocytoma, and the physiological demands of robotic surgery presents unique challenges for the anaesthetic team, necessitating meticulous planning and intraoperative management.

CCHD refers to a group of structural heart defects that result in cyanosis due to abnormal circulatory patterns. These defects, such as tetralogy of fallot, transposition of the great arteries, and tricuspid atresia, cause deoxygenated blood to bypass the lungs and enter systemic circulation, leading to chronic hypoxemia. Over time, patients with CCHD develop compensatory mechanisms, including increased hemoglobin concentration, to enhance oxygen delivery. Despite these adaptations, the cardiovascular physiology of CCHD patients remains fragile. They are often dependent on maintaining adequate preload to support cardiac output, and any fluctuation in venous return can result in cardiovascular instability. This is especially concerning in surgical settings, where multiple factors such as anaesthesia, fluid shifts, and surgical manipulation can compromise hemodynamic stability.

PHEO, though rare, further complicates this scenario. Originating from neural crest-derived chromaffin cells, PHEO typically arise in the adrenal medulla but can also occur along the sympathetic ganglia. These tumors secrete large amounts of catecholamines, mainly norepinephrine and epinephrine, which trigger significant cardiovascular effects. The unregulated release of these hormones leads to unpredictable surges in blood pressure and heart rate, increasing the risk of hypertensive crises, arrhythmias, and even myocardial ischemia. In patients with preexisting cardiac abnormalities, such as those with CCHD, managing the hemodynamic fluctuations caused by PHEO requires careful consideration of both Anaesthetic and surgical interventions [3].

Robotic-assisted surgery, while offering several advantages such as precision and minimally invasive approaches, introduces additional hemodynamic challenges. The use of pneumoperitoneum to

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insufflate the abdomen, combined with the steep trendelenburg positioning commonly used in robotic procedures, can significantly alter venous return, reducing preload and cardiac output [4]. For patients with CCHD, who are already preload-dependent, these factors can have critical consequences. Additionally, pneumoperitoneum increases intra-abdominal pressure, which may impair diaphragmatic movement and further reduce venous return to the heart. These changes must be anticipated and actively managed during the perioperative period to prevent catastrophic cardiovascular events.

The unique combination of CCHD, catecholamine-secreting tumors, and robotic surgery creates a perfect storm of potential complications. Hemodynamic instability in these patients can arise from several factors: the baseline cyanotic heart disease, the effects of catecholamine release from the tumor, and the physiological changes induced by the surgical approach. The role of anaesthesia in this context is not only to ensure the patient's comfort but also to maintain cardiovascular stability by balancing fluid management, vasopressors, and anaesthetic agents. Thus the present case study presents the perioperative challenges and management strategies in the anaesthetic care of a patient with CCHD undergoing robotic-assisted excision of pheochromocytoma.

Case presentation

We report the case of a 22-year-old female, weighing 40 kg with a height of 150 cm and a BMI of 21, presenting with a complex medical history of congenital heart disease, specifically diagnosed on birth with tricuspid atresia with D-transposition of the great arteries (D-TGA) and double outlet right ventricle (DORV). The patient had undergone a left classical Blalock-Taussig (BT) shunt in 2001 and subsequent ductal and right pulmonary artery (RPA) angioplasty in 2002. Despite these interventions, she managed her congenital condition without significant cardiac complications up to the point of her last follow-up. She presented with shortness of breath, with cyanotic apnoea spells , drop in saturations, and paroxysmal spells of headache with elevated blood pressure and anxiety, underwent a CT cardiac angiography which showed severe BT shunt thrombosis towards the pulmonary end 80%, during the same sitting an incidental finding was noted on imaging: a 5.1 x 4.9 cm hyper-vascular heterogeneously enhancing lesion originating from the left adrenal gland. This lesion mildly displaced the adjacent left kidney and was associated with an enlarged lymph node. Diagnosis of pheochromocytoma was confirmed with lab investigations showing elevated levels of metanerphine and nor metanerphine. (Fig. 1)

Significant Laboratory Findings

The patient's laboratory evaluations revealed several critical values pertinent to her case. Her 24-hour urinary metanephrine was elevated at 337 mcg/day, and her normetanephrine was significantly higher at 2806 mcg/day, suggesting active catecholamine secretion by the adrenal mass. Echocardiographic assessment showed a large secundum atrial septal defect (ASD) with right to left shunting and a single ventricle physiology. Additionally, a non-restrictive ventricular septal defect (VSD) was noted with atresia of the tricuspid valve. RV severly hypoplastic, severe BT shunt thrombosis towards the pulmonary end 80% no MAPCA or PDA multiple tiny APCAS .The pulmonary artery branches were not visible. The mitral valves appeared normal with atrioventricular concordance, and a patent Blalock-Taussig (BT) shunt was observed, N. Electrocardiogram results confirmed Sinus tachycardia with a heart rate of 122 bpm, consistent with her hypercatecholaminergic state. Blood tests indicated polycythemia, further complicating her cardiovascular management. Serum cortisol was within normal limits at 15 mcg/dl. Renin and aldosterone levels were mildly elevated with plasma renin activity at 2.9 ng/ml/hr and aldosterone at 3.2 ng/dl. DHEA sulfate was measured at 17 mcg/dl, which was within the expected range.

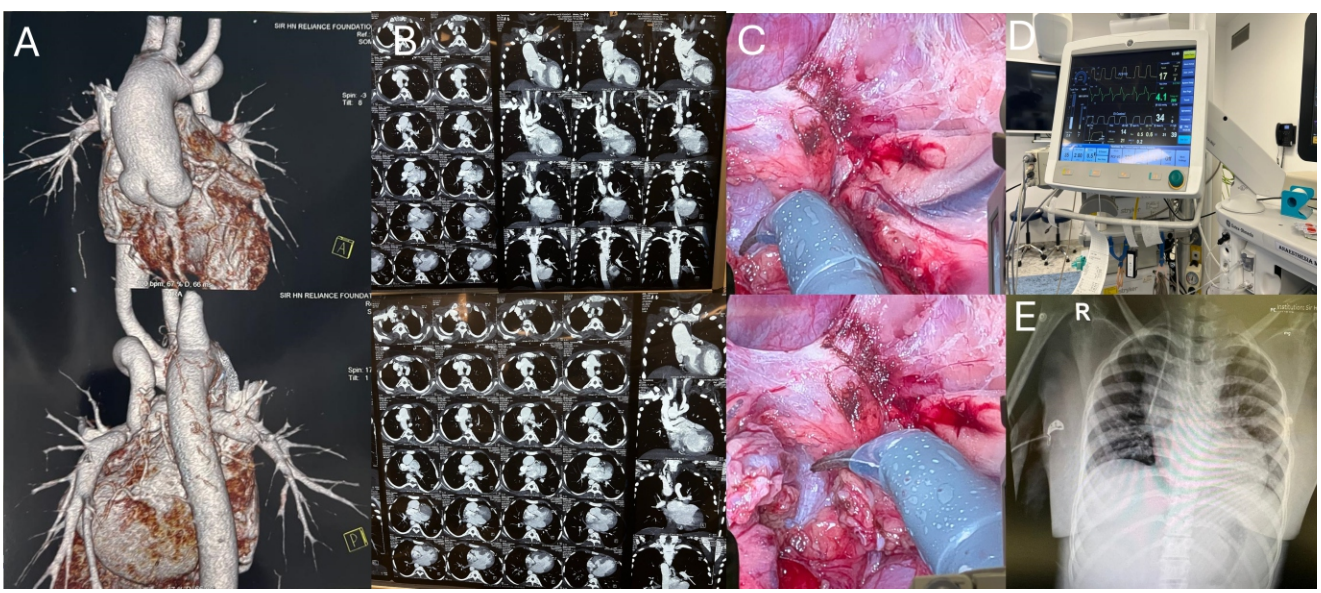

Figure 1: A. Three-dimensional computed tomography (3D CT) reconstruction of the cardiovascular system, illustrating the detailed arterial and venous structures in relation to the heart. B. Comprehensive Diagnostic Imaging: Multislice CT Scan of the Abdomen, C. Tumor, D. Intraoperating monitoring and E. Post operative chest X-ray

Liver function tests, showed total bilirubin at 0.54 mg/dL with a direct fraction of 0.27 mg/dL, ALT at 18.3 U/L, AST at 37.6 U/L, GGT at 50.4 U/L, ALP at 124 U/L, albumin at 3.65 g/dL, and globulin at 4.61 g/dL, indicating stable hepatic function. A complete blood count conducted showed a hemoglobin concentration indicative of polycythemia. Electrolyte assessment reported sodium at 134 mmol/L and potassium at 4.91 mmol/L, with coagulation studies revealing an APTT of 44.6 seconds and a PT/INR of 13.2/1.16, which were critical for assessing bleeding risks and guiding perioperative anticoagulation management.

Preoperative Management and Preoptimization

Prior to surgery in PAC OPD, the patient displayed marked respiratory distress, characterized by difficulty breathing after ascending a single flight of stairs, alongside cyanosis, pallor, tachypnea, and clubbing. Her pre-procedure oxygen saturation was critically low at 66%, highlighting severe hypoxemia. She underwent BT shunt dilatation and stenting, which improved her oxygen saturation to 80% post-procedure. To mitigate the risk of BT shunt thrombosis, a critical concern in such surgeries, anticoagulation regimen of the patient was carefully managed. She was transitioned from oral Clopidogrel to subcutaneous Enoxaparin at a dose of 0.6 mg once daily, starting one week prior to the surgery. Additionally, a 48-hour blood pressure monitoring was conducted to ensure cardiovascular stability, during which she was administered 5 mg of Propranolol three times daily and 2.5 mg of Prazosin once daily. This regimen was maintained for two weeks to manage her blood pressure, minimize surgical risks, and stabilize her condition in the perioperative period.

Intraoperative Management

A multidisciplinary team discussion, including pediatric cardiac surgeons and cardiac anaesthesiologists, was conducted prior to the procedure to outline the plan of anaesthesia and address potential risk factors. Risks associated with the surgery and postoperative ICU care were thoroughly discussed with the patient's relatives, and a high-risk consent form was obtained. The anaesthesia plan consisted of general anaesthesia supplemented with invasive monitoring. Routine ASA graded monitors and external defibrillator pads were employed. The baseline room air oxygen saturation (SpO_2) was recorded at 78% with a blood pressure of 148/70 mmHg and a pulse rate of 110 beats per minute. Special attention was given to the left limb due to a patent Blalock-Taussig (BT) shunt monitors avoided on the left limb. A 22 G IV cannula was placed in the right upper limb, and invasive lines were secured in the right radial and internal jugular vein central line under ultrasound guidance. Preoxygenation aimed for an end-tidal oxygen concentration (EtO_2) of 50%, maintaining physiological saturation between 78-82%. Premedication included intravenous pantoprazole, magnesium, dexamethasone, and hydrocortisone. Induction of anaesthesia was achieved with titrated doses of Propofol up to 50 mg IV, followed by 5 mg of Vecuronium. An episode of hypotension accompanied by compensatory tachycardia and hypertension with a heart rate of 122 and blood pressure rising to 170/90 mmHg was managed with titrated intravenous Esmolol and 2% preservative-free Lignocaine. Once physiological saturation reached 80% with a blood pressure of 110/80 mmHg and a pulse rate of 90 beats per minute, intubation was successfully performed under videolaryngoscopy, and the endotracheal tube was secured at the 18 cm mark, confirmed via capnography. After 20 minutes of hemodynamic monitoring, the patient was positioned head-low up to 18 degrees and the robotic equipment was docked. The maintenance phase included oxygen and air mixed with Sevoflurane, alongside infusions of Dexmedomidine and Vecuronium. Inotropes and vasodilators such as Noradrenaline and Sodium Nitroprusside were prepared for immediate use to manage hemodynamic fluctuations. (Fig. 2)

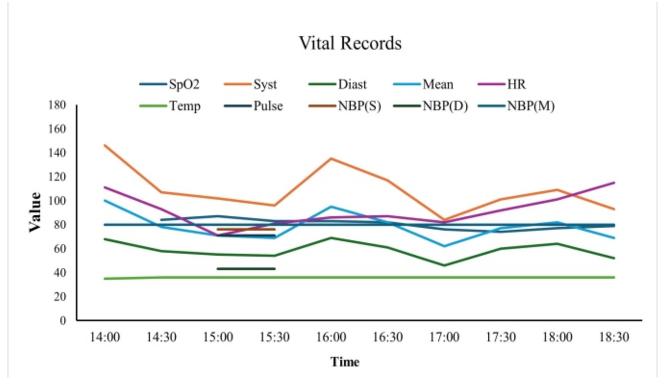

Figure 2: Intra-operative vital records of the case

Ventilation strategy involved maintaining a tidal volume of 325 ml (6 ml/kg), with a peak airway pressure of 20 mmHg and a positive end-expiratory pressure (PEEP) of 4 ml. The respiratory rate was adjusted to 14 breaths per minute with an inspiration to expiration ratio of 1:2 and a fractional inspired oxygen concentration (FIO_2) of 30%. During tumor manipulation, a 20% increase from baseline blood pressure was noted, which was effectively managed with Sodium Nitroprusside infusion and Esmolol boluses, stabilizing the hemodynamic parameters. The intraoperative fluid management accounted for 2000 ml of balanced salt solution, with a calculated blood loss of 200 ml and urine output of 1500 ml. The procedure lasted approximately 5 hours, after which the patient was gradually transitioned out of the supine position. Following a 30-minute CO_2 washout, an arterial blood gas analysis was performed to assess readiness for extubation. The extubation was carried out smoothly mild doses of esmolol were administered to avoid any hemodynamic fluctuations., and physiological saturation of 80% was maintained in the postoperative ICU with 2 liters of oxygen per minute via nasal prongs.

Discussion

Epidemiologically, both CCHD and PHEO are relatively uncommon, making their co-occurrence particularly rare. CCHD affects approximately 3 to 5 out of every 1,000 live births, while the estimated incidence of PHEO is 2 to 3 cases per million people annually [5-6]. This rare co-occurrence presents an intriguing area for clinical investigation and necessitates healthcare providers to maintain a high index of suspicion for PHEO in patients with CCHD. Early detection and management of adrenergic symptoms are crucial, despite ongoing debate regarding the potential association between these two conditions.

One potential mechanism linking CCHD and PHEO is chronic hypoxia, often observed in CCHD due to impaired oxygen delivery. Historical studies have shown a significantly higher prevalence of paragangliomas (PGL) in high-altitude populations, with a tenfold increase in younger individuals compared to those at sea level, suggesting that prolonged hypoxic exposure may stimulate adaptive growth responses, potentially leading to tumor development [7]. This is supported by a case reported by Arias-Stella and Valcarcel, revealing that bilateral carotid body PGLs experienced tumor regression after improved oxygenation, indicating that PGLs may arise as a reactive hyperplastic response to hypoxic stress rather than a primary neoplastic process [8]. In CCHD patients, chronic hypoxemia may activate compensatory mechanisms in the adrenal glands, leading to adrenal hyperplasia and increased catecholamine production, which can contribute to the development of PHEO. Sustained low oxygen levels can trigger hypoxia-inducible factors (HIFs), which are involved in regulating erythropoiesis and angiogenesis but may also affect the growth of neuroendocrine tissues. Understanding these pathways is crucial for determining if chronic hypoxia may promote tumor development, though more research is needed to establish a clear causal link to PHEO in these patients.

Another important aspect to consider is the role of genetic mutations, particularly in patients with neurofibromatosis type 1 (NF1) and RET proto-oncogene mutations. Individuals with these genetic conditions often exhibit adrenergic symptoms associated with PHEO and PGL, including palpitations, headaches, and excessive sweating—manifestations that are closely tied to catecholamine secretion [3, 9]. This variability in clinical presentation reflects the need for careful monitoring and individualized management strategies in patients with these mutations, particularly those also affected by CCHD.

Patients with single-ventricle physiology present unique challenges in perioperative management due to their preload-dependent cardiac function. Maintaining low pulmonary and left atrial pressures is essential for sustaining adequate cardiac output during surgery. The goals of perioperative Anaesthetic management in this population include maintaining normovolemia, preserving baseline central venous pressure (CVP), supporting sinus rhythm, and optimizing contractility of the systemic ventricle [10]. Additionally, care must be taken to prevent air from entering intravenous lines and to provide effective postoperative pain control while continuously monitoring oxygen saturation.

A critical consideration in this patient population is the avoidance of postoperative hypoventilation, as complications such as atelectasis and pneumonia pose significant risks that can lead to hypoxia and hypercarbia. These conditions may further elevate pulmonary pressures and diminish the transpulmonary gradient, ultimately compromising cardiac output; therefore, meticulous attention to respiratory function after surgery is essential to mitigate these risks. Additionally, the safe use of epidural analgesia in patients undergoing resection of pheochromocytoma has not been well documented. Concerns regarding epidural analgesia in this population include the unpredictable hemodynamic effects of vasodilation and bradycardia, the risk of intravenous catheter migration due to epidural collaterals, and the potential for epidural hematoma resulting from elevated venous pressures and coagulation abnormalities.

Early identification of adrenergic symptoms such as hypertension and tachycardia can lead to timely imaging and biochemical assessments. Incorporating routine screening for pheochromocytoma in preoperative evaluations is vital for improving surgical outcomes and preventing complications. Future research should focus on the genetic and molecular links between CCHD and pheochromocytoma, as well as clinical trials to establish optimal management strategies, emphasizing the need for larger cohort studies to clarify their relationship and refine clinical practices.

Conclusion

Effective perioperative fluid management is crucial for CCHD patients with PHEO, as maintaining adequate intravascular volume is essential for ensuring sufficient CVP to facilitate blood flow through the pulmonary circulation. Decreased preload can lead to reduced pulmonary blood flow and cardiac output, while increased preload can elevate afterload, intensifying the workload on the systemic ventricle. Patients with PHEO are particularly vulnerable to significant cardiac complications during the perioperative period, necessitating careful preoperative optimization, meticulous intraoperative planning, and diligent hemodynamic management. Vigilant postoperative monitoring is essential to mitigate the high risk of complications. Ultimately, the difference between CVP and systemic ventricular end-diastolic pressure serves as a primary force driving pulmonary blood flow and cardiac output, suggesting the critical need for maintaining intravascular volume, as hypovolemia is poorly tolerated in this patient population.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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