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Anaesthetic Management in a Patient with Fontan Circulation Posted for Elective Tibial Nailing

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ABSTRACT

The fatality rate for pediatric population with complex congenital cardiac disease has decreased as a result of improvements in surgical techniques and conservative management [1]. Therefore, more patients are presenting for non-cardiac surgeries as adults with congenital cardiac history [2]. For individuals with complex congenital cardiac disease and single ventricle physiology, the fontan procedure is a palliative operation that reroutes the systemic venous return to the pulmonary artery [3]. Despite improvements in outcomes, this aberrant circulation has a number of co-morbidities, including cardiac dysfunction, arrhythmias, shunt abnormalities, limited exercise ability, and other systemic manifestations [4]. These patients raise various anaesthetic concerns, such as understanding underlying cardiac morphology and mechanics and being conversant with the complexities of fontan physiology for efficient intra-operative management. In this report, we present a case of a 17-year-old male, with complex congenital heart disease with fontan physiology, posted electively for left tibia nailing under sequential combined spinal epidural anaesthesia.

KEYWORDS: Congenital cardiac disease, Fontan circulation, Combined spinal-epidural anaesthesia

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INTRODUCTION

Modern surgical methods and advancements in conservative therapy have reduced the fatalities for individuals born with complex congenital heart disease as a result more individuals with a history of congenital heart disease are presenting for non-cardiac treatments as adults[1]. Right ventricular hypoplasia is a rare congenital heart disease with an underdeveloped ventricle generally associated with pulmonary or tricuspid atresia causing right to left shunting of blood through a defect in the interatrial septa. The fontan procedure first described in 1971 is a palliative treatment for patients with a single functioning ventricle that diverts the systemic venous blood from the right atrium to the pulmonary artery bypassing the right ventricle [1]. Since then many modifications for the procedure have been described to increase chance of survival and reduce the long-term complications that come with it. the most popular being the extracardiac conduit fontan modification with or without fenestration [2]. Fenestration allows for the shunting of the deoxygenated blood to systemic circulation with a small right-to-left shunt thus reducing systemic vascular resistance and improving cardiac output [3]. The overall life expectancy with this procedure is more than 80%, but this non-physiological circuit risks long-term complications. It becomes crucial as anaesthesiologists to be well versed with the fontan physiology, intraoperative concerns, and complications associated with it.

We report the anaesthetic management of a teenage male with history of palliated complex congenital cardiac disease who came to the hospital with a left distal tibia shaft fracture posted for elective tibial nailing

CASE REPORT

A 17-year-old male, presented in the pre-anesthetic check-up for fracture left tibia surgery sustained after a road traffic accident. He gave history of having some heart disease since

birth for which we underwent multiple surgeries. On going through his past medical records, it was found that he was born with complex congenital heart disease and the 2D Echo revealed right ventricular hypoplasia, hypoplastic tricuspid valve, severe pulmonary stenosis, small ASD and a small VSD. The patient underwent Modified extracardiac fontan's procedure with fenestration at the age of 10 years. 2 years ago he started complaining of progressive dyspnea for which he underwent closure of Fontan fenestration and was doing well since then. At present he had no symptoms of cardiovascular compromise and could carry out his daily physical activities up to a maximal effort tolerance of 4 (metabolic equivalents) METS. He was on treatment with tablet aspirin 75mg once a day.

On physical examination, he was well built, weighing 52kgs and height of 170cms. He was alert, and afebrile, with a regular heart rate of 90 beats per minute, blood pressure of 110/70 mmHg in supine position, and maintained oxygen saturation of 96% on room air. Cardiac auscultation revealed a loud S1 with a single S2. Other systemic examination was normal. Preoperative blood tests were normal and ECG showed p-wave inversion in leads II, III, and chest leads V2 to V6. Echocardiography was advised, it showed status post extracardiac Fontan procedure with a fenestration closure device in situ. A patent Fontan circuit with laminar flows was observed with a normal left ventricular ejection fraction of 60%.

A night prior to the surgery, he was given tablet alprazolam 0.25mg and was advised to stay nil per oral for 12 hours. On the day of the procedure, patient was re-examined and was taken inside the operating room. Sequential combined spinal epidural was planned as the anesthetic technique of choice. Standard monitoring according to American society of Anesthesiologists was started. An intravenous access was secured with an 18G cannula and Ringer lactate was started. Patient was premedicated with IV Ondansetron 4mg, IV ranitidine 25mg and IV Cefuroxime 1.5mg IV 30 minutes before incision for surgical prophylaxis. Infective endocarditis prophylaxis was given with IV ampicillin 2g. An 18G Touhy's needle was inserted at L2-3 interspace using loss of resistance technique in sitting position, epidural catheter was advanced through it and fixed at 9cm. Subarachnoid block was performed with a 25G Quincke's needle at L3-4 level, and 1ml of 0.5% hyperbaric bupivacaine with 25mcg of fentanyl was injected slowly after CSF aspiration. Epidural top up with Injection lignocaine 2% and adrenaline 3ml was given. A sensory level of T8 and a complete motor blockage according to modified bromage scale was achieved within 10 minutes. Blood pressure was maintained within 20% of the baseline value and a total of 1L of Ringer Lactate was administered. Oxygen was administered via oxygen mask at 4L/minute. Warmer was used to main a normal body temperature and prevent hypothermia. The procedure lasted for a duration of 70 minutes without any complications. Following surgery, the patient was monitored in the postanesthesia care unit. Post-op pain control was provided with injection paracetamol 1gm BD and injection Tramadol 100mg BD.

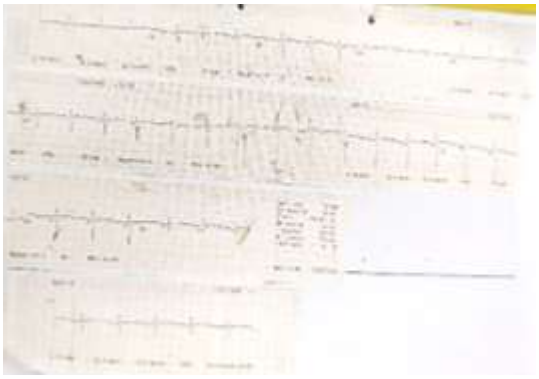
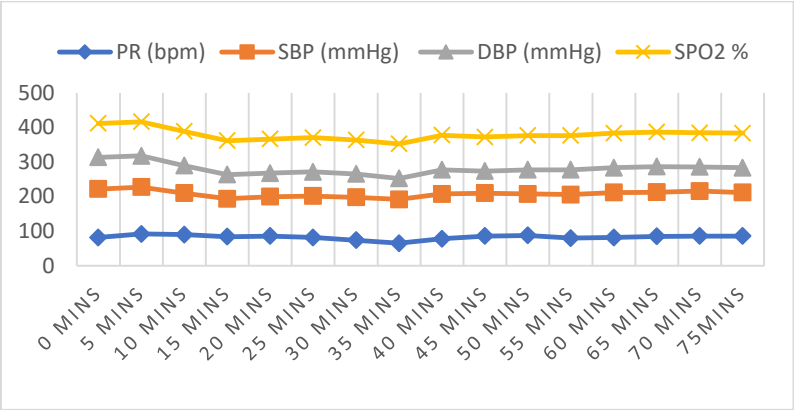


Figure 1: ECG of the patient

Figure 2: Monitor displaying intraoperative vitals



Graph 1: demonstrating Intraoperative hemodynamic variations; PR- pulse rate, SBP- systolic blood pressure, DBP- Diastolic Blood Pressure, SPO2- Oxygen saturation

DISCUSSION

In those with complicated congenital cardiac disease with only one functional ventricle the fontan procedure can prolong life and improves the quality of life [1]. It bypasses the right ventricle, directly connecting the systemic veins and the pulmonary arteries. Deoxygenated blood flows directly to the lungs for oxygenation, bypassing the right side of the heart. Over time, several modifications have been made to the conventional Fontan procedure. In the extra-cardiac conduit Fontan approach inferior vena cava is connected to the pulmonary artery via an artificial conduit composed of a biocompatible material, which totally eliminates the right atrium and simplifies the process [2]. The connection between the vena cava and pulmonary artery offers low resistance and there is only a slight pressure difference between the two blood vessels, which results in consistently low cardiac output (CO) and higher-than-normal systemic venous pressure (SVP) [4,5,6]. The anomalous Fontan physiology may lead to significant consequences such as heart failure, arrhythmias, limited exercise ability, protein-losing enteropathy, hepatic dysfunction, and plastic bronchitis [4] A side-to-side shunt between the atria and the systemic veins is surgically created, called the fenestration, increasing the CO and decreasing SVP and pulmonary artery pressure [3]. But the blood flowing through it bypasses the lungs and is therefore not re-oxygenated, thus, reducing the oxygen saturation. Our patient's fenestration closure was performed 2 years after the first procedure as he developed new onset dyspnea. Closing the fenestration typically results in improved oxygen saturation both at rest and during exercise resulting in improved exercise ability. End-organ damage may arise as a result of low CO and consistently high venous pressure like renal dysfunction due to less renal perfusion. Patients are at an increased risk of thromboembolism and are taking antiplatelet agents because of low flow states, arrhythmias, and hypercoagulability [4]. It is important to consider the congenital disease and the level of palliation attained at the time of preoperative examination. It must include thorough medical history and physical examination, the patient's functional ability, recent changes in health status, long-term effects of Fontan physiology, and current medications. Perform preliminary biochemical and hematological tests including coagulation profile. A 12-lead ECG and echocardiography for assessment of rhythm, ventricular and valvular function, PVR, and ventricular end-diastolic pressure. Review of further cardiac tests performed in the past, including echocardiography, ECG, MRI, etc.

Intraoperative care aims to maintain optimal CO. With their weakened compensating systems, even a minor CO compromise in these individuals can be harmful. Precaution should be taken to maintain a sufficient preload, excellent ventricular filling, and contractility while preventing an increase in afterload. Because the blood flow from the systemic veins to the pulmonary circulation is passive, any increase in PVR can limit ventricular filling and CO. Therefore, any increase in PVR should be avoided that could be caused by hypoxia, hypercarbia, acidosis, hypothermia, insufficient analgesia, or anaesthesia, use of vasoactive drugs, excessive mean airway pressure, and compression of the lung from pleural effusion or PEEP. Because of the diminished venous capacitance patients are extremely vulnerable to a sudden decrease in intravascular volume and to acute increases in venous capacitance, such as those caused by the direct effects of vasodilator agents or by diminished central sympathetic output. The baseline veno-constriction to maximally augment preload and therefore anaesthetics that cause veno-dilatation can be detrimental to CO and lead to cardiac instability. Such factors add to the difficulty of perioperative anesthetic management in these patients

Severe hypoxia may cause inadequate myocardial function and lack of compensation in the presence of mixed venous blood, which could result in circulatory collapse [4]. In a patient with Fontan circulation, controlling the pressure gradient between the vena cava and left atrium is essential for preventing fatal hypoxia and stabilising hemodynamic state [7]. General anaesthesia creates a secure airway but necessitates mechanical ventilation, which raises the mean intrathoracic pressure and pulmonary vascular resistance, lowers venous return and pulmonary blood flow. Regional anaesthesia permits spontaneous breathing and results in a

low intrathoracic pressure while providing excellent analgesia. However, due to the sympathetic nervous system blocking, a single spinal anaesthesia injection is usually accompanied by rapid vasodilation and reduced venous return. Therefore, combined spinal-epidural anaesthesia enables progressive attainment of sufficient analgesia without significantly altering systemic vascular resistance [8] .We performed a sequential combined spinal epidural block in our patient to ensure steady pulmonary vascular resistance and oxygenation with adequate intraoperative volume load and peripheral vascular resistance.

CONCLUSION

More adult individuals with fontan physiology are presenting for non-cardiac surgery as a result of improved surgical and medical therapy that has increased patient survival for those with complex congenital cardiac defects. Effective anaesthesia administration, necessitates an in-depth knowledge of fontan physiology and associated complications. Careful preoperative evaluation, plan of anaesthetic technique while maintaining normovolemia, avoiding myocardial depression, avoiding factors that will increase PVR, infective endocarditis prophylaxis, thromboembolism prophylaxis, and good postoperative care are crucial for a positive outcome. In conclusion, combined spinal-epidural anesthesia was the most suitable technique for this case.

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