Case Report

Nepalese Heart Journal 2020; Vol 17 (2), 51-53

Anesthetic Management of a Patient with Fontan Physiology for Laparoscopic Cholecystectomy.

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Cite this article as: Parajuli SS. Anesthetic Management of a Patient with Fontan Physiology for Laparoscopic Cholecystectomy. Nepalese Heart Journal 2019; Vol 17 (2), 51-53

Submitted date: 8th August 2020
Accepted date: 27th September 2020

Abstract

With the modification in surgical techniques and advancement in medical sciences, the survival rate of patients with complex cardiac abnormalities has increased to more than 90% in about 10 years after Fontan palliation. These patients can present with some form of non-cardiac surgeries during their lifetime. Better understanding of Fontan physiology can help in proper perioperative anesthetic management of these patients. Our cases was of a 17-year-old female patient with Fontan physiology for her Ebstein's Anomaly, planned for Laparoscopic Cholecystectomy under general anesthesia. Here we describe how we successfully managed the case perioperatively.

Keywords: Anesthetic Management; Fontan Physiology.

DOI: https://doi.org/10.3126/njh.v17i2.32681

Introduction

Children with severe Ebstein's Anomaly with hypoplastic right ventricle need various staged surgeries to increase pulmonary blood flow for survival. Francis Fontan and Eugene Baudet from France in 1971 described a technique where all systemic venous blood flows were diverted into pulmonary arteries without intervening right ventricle and thereby creating surgical palliation for various complex congenital cardiac abnormalities.[1]

With the modification in surgical techniques and advancement in medical sciences, the survival rate of patients with complex cardiac abnormalities has increased to more than 90% in about 10 year after Fontan palliation.[2,3]

These patients can present with some form of non-cardiac surgeries during their lifetime. Better understanding of Fontan physiology and maintaining the passive pulmonary blood flow can help in proper perioperative anesthetic management of these patients.[4,5] Our case was of a 17-year-old female patient with Fontan physiology for her Ebstein's Anomaly, planned for Laparoscopic Cholecystectomy under general anesthesia. Here we describe how we successfully managed the case perioperatively.

Case Report

Our case was a 17-year-old woman with a height of 152 cm and weight of 45 kg scheduled for elective Laparoscopic Cholecystectomy for symptomatic calculous cholelithiasis. She was born with severe Ebistein's Anomaly with hypoplastic right ventricle. She underwent a modified right Blalock–Taussig (BT) shunt procedure at the age of 1.5 years. When she was six years old, dissection and take down of the BT shunt and bidirectional Glenn shunt by anastomosing superior venacava to right pulmonary artery was performed. When she was 11 years old the palliation was completed by redirecting the blood from inferior venacava to the pulmonary artery thereby converting the patient to single ventricular Fontan physiology.

Currently she was under Aspirin 75mg per day. She was functionally independent with good exercise tolerance and metabolic equivalents of more than 4. She did not have any signs of cardiovascular compromise and heart failure. On examination, she was well built and oriented to person, place and time. Her general and systemic examinations were unremarkable. Her heart rate was 84 beats per minute, blood pressure was 118/74 mmHg and room air

oxygen saturation was 97%. Her routine laboratory investigations including renal function test, liver function test and complete blood count were within normal limits and her hemoglobin was 13.2gm%. Her electrocardiogram showed normal sinus rhythm. Two-dimensional echocardiography done a day prior to surgery showed patent inferior venacava Fontan pathway and superior venacava Glenn's connection to pulmonary artery with low velocity flow. Left ventricle function was normal with ejection fraction of 50%. There was no any valvular regurgitation and thrombus or clot in the cavity.

The patient was kept nil per oral after 12 midnight and intravenous ringer lactate was started at the rate of 100 ml/hour to avoid dehydration. Her regular dose of Aspirin was continued till the day of surgery. Injection Ampicillin 2g intravenous was given for infective endocarditis prophylaxis 1 hour prior to surgery. Patient was taken to the operation room and Electrocardiogram (ECG) and pulse oxymeter was attached. Defibrillation pads were attached on the back of the patient. Heating blankets and bair hugger were used in order to prevent hypothermia. Invasive blood pressure was measured by cannulating left radial artery with a 20 gauge arterial cannula.

She received 1gm of intravenous paracetamol, 4mg of intravenous ondansetron and 75 micrograms of intravenous Fentanyl before induction. Anesthesia was induced with IV etomidate 12 mg and rocuronium 30mg as muscle relaxant. Orotracheal intubation was done with 7.0mm ID cuffed polyvinyl endotracheal tube. Patient was kept in mechanical ventilation with standard ventilatory settings of tidal volume of 6ml/kg, respiratory rate of 12/min and no Positive End Expiratory Pressure (PEEP) was applied. Fraction of inspired oxygen (FIO_2) was kept 100% throughout the surgery. The peak airway pressure was 18cm of H_2O and capnography showed end tidal CO_2 of 30mmHg. After induction right internal jugular central venous catheterization was done and the opening central venous pressure was 12cm of H_2O. A nasopharyngeal temperature probe was kept to maintain temperature at 36°C–37°C. Anesthesia was maintained with 1-1.5 minimum alveolar concentration (MAC) of isoflurane and intravenous (IV) rocuronium as needed. Warm intravenous ringer lactate was infused at the rate of 80 ml per hour as a maintenance fluid.

Pneumoperitoneum was created with the CO_2 flow of 3 liters/min and maximum intraabdominal pressure was kept at 8 mmHg. Patient was kept in reverse trendelenburg position during surgery. Intraoperatively, the patient was hemodynamically stable with pulse rate 72-84/min and mean arterial pressure (MAP) of 70-84 mmHg. Intraoperatively fluid was carefully administrated to maintain central venous pressure (CVP) between 12-16cm of H_2O. The respiratory rate was increased to 15 per minutes to maintain end tidal carbondioxide ($EtCO_2$) between 30-35mmHg. The peak airway pressure increased to maximum of 22cm of H_2O intraoperatively after creation of pneumoperitoneum.The total duration of surgery was 50 mins. After completion of the surgery, neuromuscular blockade was reversed with neostigmine 2.5 mg and glycopyrrolate 0.4 mg and patient was extubated successfully. Postoperatively, the vitals were stable and postoperative period was uneventful. The patient was shifted to Intensive Care Unit and O_2 was administered via face mask at 5 liters/min. Warming blankets and bair hugger were used to keep patient warm. For postoperative pain management paracetamol 1 g intravenously 6 hourly and Fentanyl 50 mcg intravenously 6 hourly were advised. Early ambulation and sips was started after 4 hours of surgery. Her hemodynamics remained stable throughout her ICU stay and the patient was shifted to the ward on the 2nd day of surgery.

Discussion

Fontan palliation was originally described by Francis Fontan and Eugene Baudet in 1971 for tricuspid atresia.[1] However now this procedure is extensively used in variety of complex congenital heart diseases in which single ventricle physiology is required for survival. Thorough understanding of Fontan physiology is essential in properly managing these patients perioperatively.

In Fontan physiology the venous return to the heart from SVC and IVC are passively directed to the pulmonary circulation bypassing the right heart. The blood then gets oxygenated in the lungs and drains into common atrium and thereby goes to the single ventricle which perfuses the systemic circulation. The pulmonary blood flow has to be maintained and the key factors are the pressure difference between the caval veins and the pulmonary artery.[6] So any factors that increase pulmonary vascular resistance like hypoxia, hypothermia, acidosis, pain, increase in intrathoracic pressure or excessive PEEP has to be avoided. Besides to maintain passive flow gradient across the cavopulmonary circuit adequate hydration and prevention of hypovolemia is necessary. In Fontan physiology CVP represents the PA pressure and has to be ideally kept between 10-16mmHg to maintain the pressure gradient for the passive flow of 5-8mmHg. Any factors that affects this gradient causes significant decrease in the ventricular filling and hence reduce cardiac output.[6,7]

In the preoperative evaluation detailed medical history focusing on health status, exercise tolerance, current medications and history of hospital admission has to be taken. Besides thorough physical examination and baseline hematological and biochemical investigations are necessary. A 12 lead ECG and preoperative echocardiography to look for any rhythm disturbances, valvular function, pulmonary vascular resistance and ventricular diastolic pressure is mandatory. Various complications of post Fontan physiology like myocardial dysfunction, Heart failure, atrial arrhythmia, venous thromboembolism, renal dysfunction and protein losing enteropathy can occur preoperatively, so patient has to be evaluated preoperatively to rule out such complications.[8, 9,10]

The perioperative goals in these patients is to maintain adequate preload, maintain cardiac contractility, avoid factors and drugs that increases pulmonary and systemic vascular resistance. These patients are at increased risk of thromboembolism due to passive venous flow so aspirin was continued till the day of surgery and early ambulation was encouraged postoperatively. Due to scarring of right heart atrial arrhythmia refractory to pharmacologic therapy needing cardioversion[9] may be present, so we used defibrillator pads in the patient.

Intravenous anesthetic agents with minimal effect in pulmonary vascular resistance, myocardial contractility and cardiac output like etomidate[11] can be used for induction as in our case and muscle relaxants without histamine releasing property can be used.[12] In our case rocuronium was used.

The creation of pneumoperitonium by CO_2 insufflations can cause increase intra-abdominal pressure reducing venous return and thereby jeopardizing Fontan circulation and cardiac output. However, by maintaining CVP of 10-16 cm of H_2O and limiting the intraabdominal pressure to less than 10 mm Hg has not shown to decrease cardiac output.[13,14] As in our case CVP was maintained between 12-16 and intraabdominal pressure was kept below 8 mmHg.

Insufflated CO_2 can be absorbed and thereby increasing the total CO_2 content so minute ventilation should be adjusted to keep $EtCO_2$ between 30-35 mmHg intraoperatively. Finally patient has to be shifted to the ICU or postoperative ward with adequate monitoring facilities and adequate analgesia, fluid management, euthermia, fluid management, thromboprophylaxis and early ambulation has to be considered.

Conclusion

Due to medical and surgical advancement anesthesiologist can frequently encounter patients with complex cardiac abnormalities for noncardiac surgeries. So thorough understanding of the Fontan physiology and maintaining adequate cavopulmonary gradient and avoidance of the factors that jeopardizes overall myocardial function is essential in proper perioperative management of these patients.

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