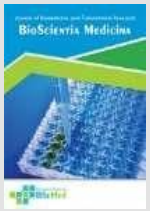
eISSN (Online): 2598-0580



Bioscientia Medicina: Journal of Biomedicine & Translational Research

Journal Homepage: www.bioscmed.com

Cochlear Implantation with Fontan Circulation: A Rare Case of Non-Cardiac Pediatric Surgery

I Putu Kurniyanta[1], I Made Bhasma Adiyana[1*]

[1]Department of Anesthesiology and Intensive Therapy, Faculty of Medicine, Universitas Udayana/Prof. Dr. I.G.N.G Ngoerah General Hospital, Denpasar, Indonesia

ARTICLE INFO

Keywords:
Anesthesia management
Cochlear implantation
Fontan circulation

***Corresponding author:**
I Made Bhasma Adiyana

E-mail address:
Bhasmaadiyana@student.unud.ac.id

All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.37275/bsm.v8i1.912

A B S T R A C T

Background: Fontan circulation is a cardiac circulation following the Fontan procedure in patients with a single ventricle congenital heart defect. This circulation depends on the difference in pressure between pulmonary capillaries and central venous pressure to achieve optimal cardiac output. This case report aims to discuss intraoperative anesthesia management in a patient with Fontan circulation undergoing a non-cardiac surgical operation. **Case presentation:** The patient is a 7-year-old male with a history of congenital heart disease who has undergone the Fontan circulation procedure and is now undergoing Cochlear implantation surgery. The patient was managed with general anesthesia - endotracheal intubation and controlled ventilation during the operation, following the principle of minimizing intrathoracic positive pressure, low PEEP, and low tidal volume with a target peripheral saturation > 95%. Cardiac output monitoring during the intraoperative period is done using intra-arterial blood pressure. After 5 hours of surgery, the patient was extubated with stable hemodynamics. **Conclusion:** The primary goal of intraoperative anesthesia management in patients with Fontan circulation is to maintain an optimal transpulmonary gradient by ensuring optimal transpulmonary blood flow, thus preserving cardiac output.

1. Introduction

Fontan circulation is a surgical approach first introduced in 1971 by Dr. Francis Fontan for patients with complex congenital heart disease involving a single ventricle such as hypoplastic left-heart syndrome (HLHS), tricuspid atresia, unbalanced atrioventricular septal defects, double-inlet left ventricle, double-outlet right ventricle, and some forms of heterotaxy syndrome.[1-4] This condition is found in 7.7% of individuals with congenital heart defects or 4 to 8 per 10,000 births.[1] The ultimate outcome of the Fontan procedure is the formation of cardiac circulation, which is different from normal circulation.[1] The estimated life expectancy up to the age of 30 in patients after undergoing the Fontan procedure exceeds 80%. It is estimated that there are over 70,000 patients living with Fontan circulation, and this number is expected to increase in the next 20 years.[1] Fontan circulation relies on the pressure difference between pulmonary capillaries and central venous pressure, where non-pulsatile blood flow from the central venous system passively moves into the pulmonary circulation due to the higher pressure in the central venous system compared to the lower pressure in the pulmonary capillaries.[2-4] Adequate cardiac output in this circulation depends on low afterload and sufficient preload.[3] The primary goal of anesthesia management in patients with Fontan circulation is to maintain an optimal transpulmonary gradient by ensuring that transpulmonary blood flow remains optimal, thereby preserving cardiac output.[2-4]

2. Case Presentation

A 7-year-old boy was diagnosed with severe bilateral sensorineural hearing loss. The patient was scheduled for Cochlear implantation surgery on the right side. The parents reported a gradual decline in his hearing over the past 2 years. Additionally, the patient had a history of congenital heart abnormalities, including double outlet right ventricle (DORV), atrial septal defect (ASD), ventricular septal defect (VSD), and pulmonary stenosis (PS). He had undergone two heart surgeries at Harapan Kita Hospital, with Bidirectional Cavo pulmonary shunt as the first stage and Fontan circulation creation as the second stage. The patient currently had a good exercise tolerance without cyanosis since the heart surgery. He regularly took warfarin with a varying dose of 1 mg - 1.5 mg every 24 hours. Physical examination revealed a blood pressure of 90/60 mmHg, regular heart rate of 108 beats per minute, normal heart sounds without murmurs, room air oxygen saturation of 92-93%, and warm extremities without cyanosis. The patient weighed 18 kg and measured 112 cm in height.

Laboratory and radiological examinations showed normal complete blood count, coagulation function, liver function, and kidney function. Chest X-ray suggested suspected pneumonia without clinical correlation. There was also a finding of dextrocardia with normal-sized heart and thoracic levoscoliosis. The latest echocardiography showed well-functioning Fontan circulation with a preserved ventricular function (ejection fraction 55%).

Anesthesia and surgical preparation followed standard procedures at our institution. In the operating room, the patient received premedication with 1 mg IV midazolam and 10 mg IV ketamine, and then sevoflurane was used for inducing anesthesia with the steal induction technique. The analgesic regimen included 50 mcg IV fentanyl, followed by 20 mg IV rocuronium for muscle relaxation. Endotracheal intubation was performed with a 5.5-sized tube and secured at 15 cm from the lip.

After induction, an arterial line was placed in the left dorsalis pedis artery, and anesthesia was maintained with O_2, compressed air, and 2% sevoflurane using a Dräger Primus® anesthesia machine in pressure control mode. Intraoperative monitoring showed stable hemodynamics, tidal volume within the range of 6-8 ml/kg body weight, and $EtCO_2$ between 33-36 mmHg. Intermittent doses of 0.5 mcg/kg IV fentanyl and 0.15 mg/kg IV rocuronium were administered. The surgery lasted 5 hours and 15 minutes with stable hemodynamics, and no vasopressors, inotropes, or chronotropes were required. The intraoperative fluid balance included 500 ml of lactated Ringer's solution. The patient had a minimal blood loss of 30 ml, and urine output was 200 ml.

Postoperatively, the patient was extubated in the operating room after obeying command, and the patient was monitored in the recovery room with stable hemodynamics (blood pressure 107/58 mmHg, heart rate 68 beats per minute, respiratory rate 22 breaths per minute, and peripheral oxygen saturation 96% with a 5 L/min face mask). The patient was alert, with round and reactive pupils (diameter 3 mm/3 mm) and a pain score of 0/10 on the Wong-Baker faces scale. Two hours after monitoring in the recovery room, the patient was deemed fit to be transferred to a regular ward. The patient was given fentanyl 100 mcg of 20 ml NaCl 0.9%, infused at a rate of 0.8 ml/hour, oral Ibuprofen 200 mg every 8 hours, and oral Paracetamol 200 mg every 8 hours for postoperative pain management. The patient stayed in the ward for 2 days without complications before being discharged.

3. Discussion

The patient is a 7-year-old child with a history of congenital heart defects, specifically DORV. The patient underwent Fontan surgery at the age of 2. From the history, physical examination, and diagnostic tests, there are no signs of Fontan circulation failure, such as dyspnea, fatigue, decreased tolerance for physical activities, weight gain, palpitations, syncope, new murmurs, hepatomegaly,

or oxygen saturation below 90%.[3] It can be concluded that the Fontan circulation is currently functioning well.

The required hemodynamic monitoring is adjusted based on the patient's physical condition and the complexity of the surgery. The use of intra-arterial blood pressure (IABP) is recommended for patients undergoing complex and prolonged surgeries to monitor organ perfusion continuously, collect blood gas analysis, and ensure the availability of oxygen.[3,6]

The primary goal of anesthesia management in Fontan circulation patients is to maintain an optimal transpulmonary gradient, ensuring that transpulmonary blood flow remains optimal to preserve cardiac output.[2,3] During anesthesia induction, there is a potential for decreased cardiac output, myocardial depression, and systemic vasodilation. Therefore, anesthesia agents with minimal myocardial depression effects, such as thiopental or inhalation agents, should be used.[4,7,8] In this case, midazolam and ketamine were used for premedication, and sevoflurane was used for induction with the aim of not increasing pulmonary vascular resistance (PVR) and further depressing myocardial function.

The main principle of intraoperative respiratory management is to minimize intrathoracic positive pressure, maintain low positive end-expiratory pressure (PEEP), and low tidal volume, targeting peripheral oxygen saturation > 95%. The success criteria for mechanical ventilation during the intraoperative period were moderate alkalosis (pH 7.45, pCO_2 35 mmHg) and preventing atelectasis without compromising hemodynamics.[1,4,9] The use of inhalation anesthesia agents for anesthesia maintenance is not recommended above 1.5 MAC to avoid cardiac arrhythmias.[3] Nitric oxide (NO) is beneficial in maintaining low PVR. A high fraction of oxygen also results in vasodilation in the pulmonary capillaries, thereby increasing transpulmonary blood flow.[4,10] In this case, intraoperative respiratory management was maintained with O_2, compressed air, and 2% sevoflurane using a Dräger Primus® anesthesia machine in pressure control mode, O_2 fraction of 50%, fresh gas flow 1 liter, Pinsp 15, Psupp off, RR/Freq 25, PEEP 3, during the surgery Tidal volume is maintained between 6-8 ml/kg body weight, and $EtCO_2$ between 33-36 mmHg.

Baseline information on hemodynamics and cardiac capacity is crucial to prevent over-resuscitation or under-resuscitation.[3] Fluid administration in Fontan circulation patients should be guided by central venous pressure (CVP), transesophageal echocardiography (TEE), or esophageal Doppler.[4] Hypotension in this population can have fatal effects due to a potentially very low arteriovenous pressure gradient.[4] The intraoperative management goal is to balance the patient's volume status between preload and afterload. In this case, the surgery lasted 5 hours and 15 minutes, and total bleeding was 30 mL, and urine production was 200 mL. We only administered 500 mL of crystalloid fluid of Ringer Lactate during the intraoperative period. The patient's hemodynamics during surgery remained stable, as observed from the trends in blood pressure, pulse rate, and peripheral oxygen saturation. This patient did not require vasopressors, inotropes, or chronotropic medications during the surgery. Consequently, the patient did not require special care in the intensive care unit (ICU). ICU care would only be necessary if there were concerns about the possibility of further fluid shifts after major surgery.

4. Conclusion

The primary goal of intraoperative anesthesia management in patients with Fontan circulation is to maintain an optimal transpulmonary gradient by ensuring optimal transpulmonary blood flow, thus preserving cardiac output. This can be achieved by minimizing the increase in intrathoracic pressure during ventilation management. Additionally, maintaining the patient volume status in a state of euvolemia is crucial, as this population is highly vulnerable to changes in preload.

5. References

1. McNamara JR, McMahon A, Griffin M. Perioperative management of the Fontan patient for cardiac and noncardiac surgery. In Journal of Cardiothoracic and Vascular Anesthesia. 2022; 36(1): 275–85.
2. Nayak S, Booker PD. The Fontan circulation. continuing education in anaesthesia, Critical Care and Pain. 2008; 8(1): 26–30.
3. Jolley M, Colan SD, Rhodes J, Dinardo J. Fontan physiology revisited. Anesthesia and Analgesi. 2015; 121(1): 172–82.
4. Muñoz Y, José Sáenz M, Cifuentes R. Physiopathological approach of the Fontan patient for noncardiac surgery for the anesthesiologist. In Advances in Complex Valvular Disease. IntechOpen. 2021.
5. Kothandan H. Fontan physiology: anaesthetic implications for non-cardiac surgery: a case report. International Journal of Anesthetics and Anesthesiology. 2015; 2(1).
6. D'souza S, Satarkar B, Bharne SS. Anaesthesia for a minor procedure in a patient with Fontan physiology. Indian Journal of Anaesthesia. 2012; 56(6): 572–4.
7. Henao SMC, Uribe MM, Ospina De Los RÁ. Anesthetic management for non-cardiac surgery in a patient with Fontan palliation: Case report. Colombian Journal of Anesthesiology. 2019; 47(4): 253–6.
8. d'Udekem Y, Iyengar AJ, Galati JC. Redefining expectations of long-term survival after the Fontan procedure: twenty-five years of follow-up from the entire population of Australia and New Zealand. Circulation. 2014; 130: S32–S38.
9. Francis K, McGuire M, Celermajer D. Electrophysiology and radiofrequency ablation for the management of tachyarrhythmias in adult Fontan patients. Heart Lung Circ. 2015; 24: S429.
10. Correa R, Walsh EP, Alexander ME. Transbaffle mapping and ablation for atrial tachycardias after mustard, senning, or Fontan operations. J Am Heart Assoc. 2013; 2: e000325.