



## Perioperative Management of Tonsillectomy in Pediatrics with Tetralogy of Fallot

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### ABSTRACT

**Introduction.** Tetralogy of Fallot (TOF) is a cyanotic congenital heart disease that includes ventricular septal defect, aortic overriding, infundibular stenosis, and Right Ventricle Hypertrophy (RVH). Perioperative management includes preoperative management, intraoperative anesthesia, and post-operative and potential problem management.

**Case presentation.** We report a -8-year-old-girl with congenital heart disease TOF planned for tonsillectomy and adenoidectomy. The patient weighs 18 kilograms, has a blood pressure of 96/55 mmHg, pulse rate of 104 times per minute, respiratory rate 24 times per minute, and oxygen saturation of 74-79% supine position with room air. We find ejection systolic murmur at SIC 4/6 left parasternal and clubbing fingers-on physical examination. Laboratory preoperation shown Hb value 22.9; Hct : 70; AL : 11.2; AT : 281; AE : 8.58. We pre-medicate with ondansetron, dexamethasone, and sedation in the transit room with ketamine and midazolam. Induction by fentanyl and rocuronium to facilitate endotracheal intubation.

Postoperatively, the patient was transferred to the PICU with an ET in a tube. She is monitored for a post-operative complications such as active bleeding to hyper cyanotic and performed extubation less than 24 hours after surgery.

**Conclusion.** Anesthetic goals in this patient are to maintain or slightly increase the Systemic Vascular Resistance, minimize Pulmonary Vascular Resistance dan provide mild cardiac depression.

**Keywords:** TOF, pediatric, tonsillectomy.

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## Introduction

Tetralogy of Fallot (TOF) is a cyanotic congenital heart disease that is often found in children. TOF is characterized by the presence of a ventricular septal defect, aortic overriding, infundibular stenosis, and right ventricular hypertrophy (RVH).<sup>1-3</sup> TOF is cyanotic congenital heart disease with an incidence rate of  $\pm 16\%$  of existing congenital heart disease.<sup>1,2,4,5</sup> Clinical manifestations of TOF are caused by a decrease in pulmonary blood flow, where the degree of cyanosis of TOF is determined by the severity of obstruction to blood flow out of the right ventricle (infundibular stenotic).<sup>6</sup>

Patients with a history of TOF have a high risk of bacterial endocarditis, and noncardiac surgery is often performed to exclude sources of infection that may be associated with bacterial endocarditis. In a clinical review of 191,261 patients under the age of 18 who underwent noncardiac surgery, a history of congenital heart disease increased the risk of death in both minor and major surgery. General anesthesia procedures in patients with a history of TOF have a risk of hemodynamic disturbances that can lead to an increased risk of hypoxia both before anesthesia, during anesthesia, and after anesthesia.<sup>7</sup>

## Case Presentation

An 8-year-old girl diagnosed with chronic tonsillitis and adenoid hypertrophy having a history of cyanotic congenital heart disease tetralogy of Fallot was planned for tonsillectomy and adenoidectomy at Dr. Moewardi General Hospital Surakarta. She was preparing for TOF correction at Harapan Kita National Hospital, Jakarta. The patient came with a complaint that her throat had felt lumpy since 2 years ago. She also complained of shortness of breath when doing heavy activities. Lastly, shortness of breath and cyanotic 2 weeks before the patient came to the hospital. History of other diseases such as asthma and drug allergy is denied.

On physical examination, body weight was 18 kilograms, blood pressure was 96/55 mmHg, pulse rate was 104 times per minute, respiratory rate was 24 times per minute, and oxygen saturation was 74-79% supine position with room air. On examination of the airway, the airway is free from obstruction, mallampati II, free neck movement, and toothless (+). Tonsil hypertrophy was found with sizes T3-T4. The mass can move freely, crypt (-) color like surrounding tissue, and has no visible signs of inflammation as in Figure 1A. On physical examination, the heart sounds regular I-II, murmur (+) ejection systolic in left parasternal 4/6 SIC, warm acral, and clubbing fingers in all four extremities. Examination of other body systems within normal limits.



Figure 1. A. Clinical patients B. Tonsil T3-T4

Blood laboratory examination revealed a hemoglobin value of 22.9 g/dL; Hematocrit 70%; leukocyte count  $11.2 \times 10^3$ , platelet count  $281 \times 10^3$ , erythrocyte count  $8.58 \times 10^6/\mu\text{L}$ ; Antistreptolysin type O (ASTO) titer is above 400. Electrocardiographic examination showed a normo sinus rhythm with a pulse rate of 103 beats per minute, with a deviation of the axis to the right, and no ST-segment changes were found. Radiological examination of the lateral AP head shows adenoid hypertrophy. Echocardiography revealed AV-VA solitus concordance site of the systemic venous estuary, normal pulmonary veins; Perimembranous outlet VSD (PMO) with a diameter of 0.8 cm; overriding aorta 50%; tricuspid regurgitation (TR) mild with peak pulmonary gradient (PG) 32.84 mmHG; severe pulmonary stenosis with Peak PG 78.37 mmHG; ejection fraction (EF) 64%, LA/Ao 1.11, E/A > 1; Aortic arch to the left of Koar (-).

The patient's physical status, according to the American Society of Anesthesiologists (ASA) 3 with a general anesthetic plan with an endotracheal tube (GA-ET), non-invasive monitoring, and post-operative care in the Pediatric Intensive Care Unit (PICU).

The patient fasted for 6 hours with the maintenance of 56 ml/hour of fluid through a venous catheter (22 gauge). The patient was given premedication on Ondansetron 2 mg and dexamethasone 2 mg. Before entering the operating room, the patient was sedated in the transit room with Ketamine 10 mg and Midazolam 1 mg, the patient was brought to the operating room, and an ECG monitor was installed, showing SpO<sub>2</sub> 74%, and Non-invasive Blood Pressure (NIBP) 96/54 mmHg, pulse rate 108 beats per minute. Preoxygenation was given with 100% O<sub>2</sub>, and the patient was induced with fentanyl 36 mcg, and the facility was intubated with 10 mg Rocuronium. Intubation using a size 6.0 endotracheal tube.

Maintenance of patients using Sevoflurane 2-2.5 vol% and O<sub>2</sub> 50% in Airbar. The surgery lasted for 1 hour with 40 ml of bleeding. 56 ml/hour crystalloid was given during the operation. The patient has been transferred to the PICU room with a size 6.0 endotracheal tube attached.



Postoperative analgesia with Paracetamol 400mg/ 8 hours and Fentanyl 0.5 mcg/kg/hour. Post-operative monitoring was performed to evaluate post-operative complications and anesthesia. The patient was extubated less than 24 hours postoperatively.

## **Discussion**

Children aged 8 years were planned for tonsillectomy and adenoidectomy surgery for indications of chronic tonsillitis and adenoid hypertrophy. The patient had a history of cyanotic congenital heart disease and Tetralogy of Fallot (TOF).

Preoperative assessment was carried out together with pediatric cardiologists, nurses, and surgeons in detail to obtain information on medical history, surgery, and anesthesia history so as to identify possible aggravating factors in the patient, according to the picture of cyanotic congenital heart disease, Tetralogy of Fallot (TOF).

The patient was being prepared for TOF correction at the Harapan Kita Heart Center Hospital, Jakarta, with the complaint that the throat has felt lumpy since 2 years ago. The patient has complaints of shortness of breath when doing strenuous activities, and lastly, shortness of breath and blues 2 weeks before the patient came to the hospital. There was no history of drug use in the last 1 month. A history of other diseases such as diabetes mellitus, asthma, and drug-allergy was denied.

Preoperative patient visits are carried out to establish good communication with the family to provide information about the surgery and anesthesia that will be carried out as well as complications that can occur in post-operative care. It is also important to do psychological preparation for the patient and the patient's family before the operation. Followed by a physical examination, the body weight was 18 kilograms, blood pressure 96/55 mmHg, pulse rate 104 times per minute, respiration rate 24 times per minute, and oxygen saturation of 74-79% supine position with room air. On examination of local status, tonsillar hypertrophy was obtained with a size of T3-T4, the mass was free to move, crypt (-) color-like surrounding tissue, and no signs of inflammation were seen (see Figure 1B). Physical examination of the heart revealed a pulse rate of 104 beats per minute, regular I-II heart sounds, and a systolic ejection murmur at SIC 4/6 left parasternal.

Physical examination in patients with TOF is often found to have heart murmurs, especially along the left sternal edge, caused by pulmonary valve stenosis.<sup>6</sup> Although congestive heart failure can occur in TOF patients. This event is rare because the presence of a large VSD causes a balance of pressure between the heart's right and left.<sup>8</sup>



On electrocardiographic examination, this patient obtained a normo sinus rhythm reading with a pulse rate of 103 beats per minute, with a deviation of the axis to the right, and no ST-segment changes were found. Electrocardiographic examination of patients with TOF found characteristic changes in the right axis and right ventricular enlargement. The state of congestive heart failure rarely occurs due to a large VSD. This condition is caused by the intraventricular pressure to the heart load in a balanced state.<sup>9</sup>

The results of the echocardiography examination support the description of the TOF cardiac anomaly. The AV - VA solitus site found concordance of systemic venous estuaries, normal pulmonary veins; PMO VSD with a diameter of 0.8 cm; overriding aorta 50%; TR Mild with Peak PG 32.84 mmHG; PS Severe with Peak PG 78.37 mmHG; EF 64%, LA/Ao 1.11, E/A > 1; Aortic arch to the left of Koar (-).

Lab results showed Hemoglobin 22.9 g/dL, Hematocrit 70%, thrombocytes 281.000/mm<sup>3</sup>, and leukocytes 11.200/mm<sup>3</sup>. The results of laboratory examinations showed that the hemoglobin concentration was high, so there was a problem of polycythemia vera in the patient, which was compensation for chronic hypoxia.

In TOF, chronic hypoxemia occurs so that it will stimulate erythropoiesis, and secondary erythrocytosis can occur. Secondary erythrocytosis from chronic arterial hypoxemia will cause disturbances in coagulation function, so TOF patients with erythrocytosis, especially hematocrit up to 70%, are at high risk for bleeding during surgery due to impaired hemostasis. K-dependent clotting factor in the liver and impaired platelet aggregation.<sup>2,3</sup>

The patient fasted for 6 hours with the maintenance of 56 ml/hour fluids via a venous catheter (22 gauge). Prior to surgery and anesthesia, the patient's condition must be in optimal condition. Keeping the patient in a state of euvolemia is very important. This can be maintained by ensuring oral intake before fasting and can be administered through maintenance with intravenous access. Cardiac drugs that have been given to this patient should be continued until the time before surgery except diuretics.<sup>2,3</sup>

Before leaving for the operating room, the patient was given premedication on Ondansetron 2 mg and dexamethasone 2 mg. The administration of this premedication is expected to provide an antiemetic effect and reduce the histamine release that may occur.

The patient has sedated in the transit room with Ketamine 10 mg and Midazolam 1 mg. This is done to prevent the patient from crying so that an increase in PVR can be avoided. Crying in TOF patients is avoided because it can lead to hypercyanosis. Therefore, the administration of drugs such as sedatives and anxiolysis is needed in these patients.



Intramuscular administration can be performed in patients who do not have an intravenous line installed.<sup>5</sup> However, this method sometimes causes pain and can cause the child to cry. It can be considered oral administration with midazolam at a dose of 0.75 mg/kg BW 30 minutes before being sent to the operating room. Besides that, it can be given an alternative by intranasal.<sup>2,3,5</sup> Intranasal administration with ketamine 10 mg/kg BW provides a sedative effect when the patient is separated from his parents or for facilities in terms of the installation of intravenous access.

Ketamine intravenous is used based on the effect of ketamine which can increase the dominant heart rate when compared to pulmonary vascular resistance (pulmonary vascular resistance, blood pressure, cardiac output, and causes an increase in SVR/PVR). Ketamine also has an analgesic effect without causing respiratory depression. In principle, the administration of anxiolytics and sedation in mild conditions aims to maintain respiration.<sup>2,3,5</sup> In patients who are very anxious, intramuscularly, Ketamine and Midazolam can be given but on the condition that they are only given when the patient will be brought to the operating table. During the procedure, monitoring is continuously carried out, one of which is a precordial stethoscope and pulse oximetry.

Monitoring includes ECG pulse rate 108 times per minute, SpO<sub>2</sub> 74%, and Non-invasive Blood Pressure (NIBP) 96/54 mmHg. Preoxygenation with 100% O<sub>2</sub> and the patient has induced with Fentanyl 36 mcg, and the facility was intubated with Rocuronium 10 mg. Intubation using a size 6.0 endotracheal tube. Maintenance of patients using Sevoflurane 2-2.5 vol% and O<sub>2</sub> 50% in Airbar.

Intravenous induction in TOF patients has a more rapid onset but in contrast to slower inhalation induction. Sevoflurane was chosen as an inhalation agent because it did not significantly affect the ratio of SVR to PVR because sevoflurane decreased not only SVR but also PVR. The use of Halothane provides a relatively more stable SVR with a stronger negative inotropic effect, thereby reducing the effect of infundibular spasm, but Halothane is more likely to cause arrhythmias.

The use of opiates in anesthesia for children with congenital heart disease can provide good hemodynamic stability. High-dose opiates can be used in children with major surgery, although the use of a ventilator will be required to support post-operative ventilation. Low-dose opiates may be considered to reduce concentrations with inhaled agents.<sup>7</sup>

In this case, rocuronium was given as a facility for intubation because it did not cause the release of histamine, which would cause systemic vasodilation resulting in a decrease in SVR.



Fluid administration needs to be considered wise to keep the patient in a state of euvolemia. This patient was given 56 ml/hour of crystalloid fluid during surgery for fluid maintenance. The operation lasted for 1 hour with bleeding of 40 ml.

One of the complications that need to be considered in patients with TOF is cyanosis. Where this attack is better known as the Hypercyanotic Spell, which indicates an increase in right-to-left shunts due to a higher PVR than SVR. When a hypercyanotic spell occurs, the systemic oxygen saturation level drops rapidly. Make sure the patient is immediately positioned with both legs bent towards the chest to reduce preload, then given 100% oxygen, intravenous fluids, and give Noradrenaline 0.04 mcg intravenously. This is an effort to increase SVR so that it is higher than PVR.

After the operation, the patient was admitted to the PICU for post-operative monitoring. While in the PICU, the patient was monitored for post-operative complications and extubated 24 hours postoperatively. The Cuffleak Test can be performed before extubation to assess for laryngeal edema.

The postoperative analgesics used were Paracetamol 400mg/8 hours and Fentanyl 0,5mcg/kg/hour. Optimal analgesia administration is needed to control pain in patients so as to reduce the risk of changes in the physiological condition of the cardiovascular system.

Anesthesia management in patients with TOF is very important to understand the pathophysiology and pharmacology of anesthetic drugs that will affect right-to-left shunting of VSDs. When the shunting is actually increased, there will be a decrease in pulmonary blood flow and a decrease in PaO<sub>2</sub>. Right-to-left shunting through the VSD can increase in a state of decreased systemic vascular resistance, increased pulmonary vascular resistance, and increased myocardial contractility caused by sympathetic tone, which will lead to infundibulum spasm which will cause an increase in right ventricular outflow obstruction.<sup>1,3,4</sup> In general, the size of the right-to-left shunt is influenced by: 1. Decreased systemic vascular resistance, 2. Increased pulmonary vascularity, 3. Increased cardiac contractility is related to obstruction of the infundibulum of blood flow from the right ventricle.<sup>8</sup>

Pharmacologically the use of drugs that induce hypotension due to decreased SVR, such as inhalation agents, muscle relaxants, ganglionic blockers, and alpha-blockers, will increase the magnitude of the right-to-left shunting. Giving positive pressure to the lungs will also be able to increase intrathoracic pressure and will encourage an increase in PVR. Increasing PVR will be able to reduce blood flow in the pulmonary circulation. Therefore, giving positive pressure when controlling breathing also needs to be done carefully.<sup>4,5</sup>



### **Conclusion**

Anesthetic goals in this patient are to maintain or slightly increase the Systemic Vascular Resistance, minimize Pulmonary Vascular Resistance dan provide mild cardiac depression.

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