CASE REPORT

Anesthesia Management in Blalock-Taussig Shunt Procedure

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ABSTRACT

Background: The systemic to pulmonary artery shunts are done as palliative procedures for complex cyanotic congenital heart diseases. Blalock-Taussig shunt (BT shunt) provide regulated blood flow to the lungs allowing growth of pulmonary arteries until the patient reaches proper age and body weight suitable for definitive corrective repair. BT shunts are first line management in patients with critical cyanotic conditions.

Case: A 12-month-old boy diagnosed with PA-VSD subaortic, L-R shunt PDA and critical PDA stenosis experienced a recurrent spell condition with the lowest oxygen saturation 40%, underwent urgent BT shunt surgery. Oxygen saturation increases to 80-85% after shunt procedure.

Discussion: Anesthesia management includes optimizing preoperative condition and patient hydration state, providing balance anesthesia during surgery, maintaining balance of pulmonary and systemic blood flow. High oxygen fraction can be given to maintain oxygen saturation before BT shunt anastomosis. Mechanical ventilation, heart rate with sinus rhythm, preload and contractility is maintained to obtain normal cardiac output. After BT shunt anastomosis, the oxygen fraction is reduced with a saturation target of 70–85%. Postoperative management includes anticoagulant administration and monitoring postoperative complications. The patient developed complications of increased pulmonary blood flow postoperatively and was admitted to the PICU for 3 days. The patient was discharged in good condition from ward on day 7.

Conclusion: Understanding the physiology of heart defects and perioperative management determine the success of BT shunt surgery, reducing patient morbidity and mortality. Optimizing intraoperative and postoperative oxygen delivery with oxygenation targets PaO_2 40-45 mmHg and saturation 70-80% reflects the balance of pulmonary blood flow and systemic blood flow (Qp:Qs=0.7-1.5:1).

Keywords: anesthesia management; blalock-taussig shunt; PA-VSD; perioperative management; pulmonary atresia

INTRODUCTION

Congenital heart defects are found in 8 per 1000 live births and 25% of those being cyanotic. Palliative surgery is performed on patients with critical cyanotic heart defects that cannot be fully corrected in order to maintain life.[1]

Shunt from the systemic to the pulmonary artery is a palliative procedure in patients with complex cyanotic congenital heart defects. This shunt is the first line management for neonates experiencing critical cyanotic conditions.[2,3]

The purpose of a systemic to pulmonary shunt is to assist the pulmonary artery in developing and maintaining regulated blood flow to the lungs until the patient reaches appropriate age and body weight for definitive corrective repair. This procedure increases blood flow to the pulmonary artery, resulting in higher arterial oxygen saturation, reduced cyanosis, and improved physical activity tolerance.[2,4]

BT shunt is the most performed systemic to pulmonary shunt procedure. According to data from the National Congenital Heart Disease Audit in the United Kingdom, 171 neonatal patients (<30 days) underwent BT shunt surgery between 2012 and 2015. At the National Cardiovascular Center Harapan Kita in Jakarta, 73 out of 897 patients with congenital heart defects (8.13%) underwent palliative systemic-pulmonary shunt surgery in 2022.[5]

Perioperative anesthetic management in BT shunt procedure remains challenging due to a relatively high intrahospital mortality rate. This rate reaches 15% in single ventricle patients and 3% in biventricular patients. Understanding the physiology of heart abnormalities is crucial in perioperative management to ensure the success of BT shunt surgery and reduce patient morbidity and mortality.[2]

Literature on comprehensive anesthetic management in BT shunt procedure from preoperative to postoperative period is limited. This report presents a case on perioperative anesthetic management in BT shunt procedure, as literature on anesthetic management in BT shunt procedure related to the distribution of pediatric cardiac surgery services in Indonesia.

CASE

The patient is a 12-month-old boy who presented with blue lips and nail tips, which are particularly noticeable when he is crying or tired. He also exhibits rapid breathing during physical activity. There were no complaints of shortness of breath, cough, or fever during the preoperative visit. The patient underwent 7 days of treatment before surgery due to repeated spells resulting in a saturation level of 40%. Urgent BT shunt surgery was scheduled.

The heart defect has been known since he was 8 months old and takes regular medication, captopril with a gradual increase in dosage up to 12 mg every 8 hours.

Patient born in spontaneous birth with midwife assistance. Birth weight was 2500 grams. The newborn immediately cried and had blue lips and nails. Patient is the third child of 3 siblings. There is no history of congenital abnormalities in the family. The patient has no history of allergies or other medical conditions.

During the physical examination, the patient was alert and had strong crying movements. Body weight 7.3 kg, body

length 70 cm, body surface area (BSA) 0.38 m^2. Respiratory rate 32 times/ minute, SpO_2 62% without oxygen supplementation. Vesicular breath sounds are present and there no crackles or wheezing. Pulse rate is 129 times/minute, regular, strong lifting. Blood pressure is 88/67 mmHg, heart sounds are regular and there is a murmur. The acral palpable is warm, not edematous and appears cyanotic.

Preoperative laboratory examinations are presented in Table 1. Antero-Posterior (AP) chest x-ray showed an enlarged heart with a cardio thorax ratio (CTR) of 56%, accompanied by increased bronchovascular patterns (Figure 1).

Trans-thoracic echocardiography (TTE) examination showed VA pulmonary atresia, subaortic R-L shunt ventricular septal defect (VSD). There was a patent ductus arteriosus (PDA) vertical duct, L-R shunt with continuous flow. Left ventricular systolic function normal, ejection fraction (EF) 63%. Right ventricular systolic function normal, tricuspid annular plane systolic excursion (TAPSE) 1.6 cm. Aorta overriding with trivial aortic regurgitation (AR), no aortic stenosis (AS). Pulmonary atresia, confluent pulmonary arteries, right pulmonary artery (RPA) diameter 4.8 mm, left pulmonary artery (LPA) diameter 3.6 mm, descendens aorta (AoD) diameter 7.45 mm (Halfsize 6.5; McGoon 1.1). Mitral and tricuspid valves are good, aortic arch was on the left with no coarctatio.

Table 1. Preoperative laboratory examination results

Hemoglobin	17.9	PT	11.9	Bil. D	0.15	Na	135	pH	7.29
Hematocrit	63.3	aPTT	28.0	Bil. I	0.26	K	4.6	pCO_2	38.8
Leukocyte	7560	SGOT	38	Bil. T	0.41	Cl	103	pO_2	41.5
Thrombocyte	226000	SGPT	13	Ur	30.5	Ca	2.49	BE	-8.3
Anti HCV	NR	Albumin	4.2	BUN	14.3	Mg	2.3	HCO_3	18.5
HbSAg	NR	Globulin	2.4	Cr	0.38	CRP	6	SaO_2	66%

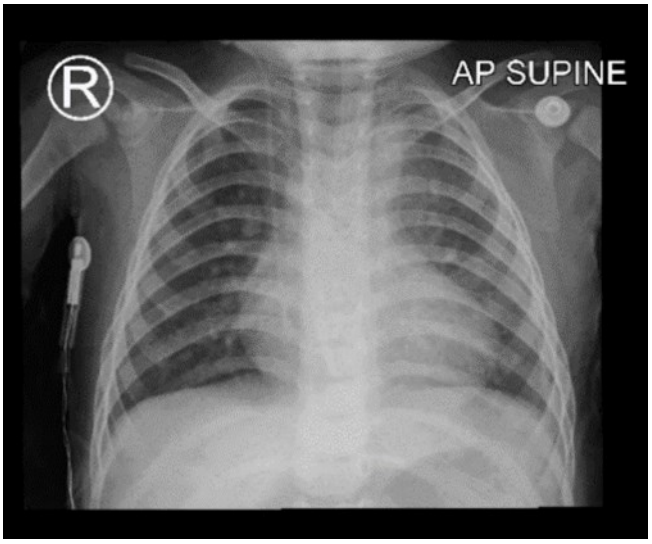

Figure 1. Preoperative chest x-ray

Cardiac catheterization revealed pulmonary atresia, subaortic VSD (fallot type), and PDA. Pulmonary arteries were confluent, with incomplete right lung arborization and complete left lung arborization. RPA diameter 5.9-6.1 mm, LPA diameter 6.4-6.4 mm, and AoD diameter 10.48 mm (Halfsize 6.5; McGoon 1.2). There was severe stenosis at distal PDA with mild stenosis at the bifurcation of the pulmonary artery and RPA. Major aorto pulmonary collateral arteries (MAPCA) of the AoD filled the right upper lung field.

Patient was assessed as physical status American Society of Anesthesiologists (ASA) 4. Prior to surgery, the patient's family was educated on the importance of fasting and blood component preparation. The anesthesia procedure was explained, including the associated risks, side effects, and complications, as well as care and administration of postoperative analgesia in the intensive care unit (ICU).

On the day of surgery, patient was taken into the operating room and underwent inhalation induction with sevoflurane. Electrocardiogram (ECG) monitoring and pulse oximetry were installed simultaneously. Peripheral intravenous access was placed in the right dorsum vein and arterial line in the right femoral artery. Pre-induction evaluation showed blood pressure 74/42 mmHg, pulse rate 122 beats/minute, respiration rate 36 times/minute, SpO_2 52%, ECG sinus rhythm. The patient received 70 ml of ringerfundin, then anesthesia was induced using 1 mg of midazolam, 20 mcg of fentanyl, 1 mg of vecuronium, and 1 volume% sevoflurane.

Once drug onset was achieved, an uncuffed endotracheal tube (ETT) 4.0 used for intubation to a depth of 12 cm, and breath sounds were evaluated. Central venous catheter was inserted into the right internal jugular vein. Mechanical ventilation was initiated using ventilator pressure control mode with a pressure control (PC) of 12, inspiration: expiration (I:E) ratio 1:2, positive end expiratory pressure (PEEP) 3 and oxygen fraction 60%. Inhalation agent sevoflurane at 1-2 volume%, intermittent fentanyl, and vecuronium were used for maintenance.

Patient was positioned in right lateral decubitus for left thoracotomy, the surgery field was disinfected and prepared. Additional analgesic fentanyl 30 mcg and vecuronium 1 mg were administered before the incision. No significant hemodynamic changes were observed during the incision. The left postero-lateral thoracotomy incision was made at intercostal space (ICS) 3, penetrating the cutis, subcutis, and latissimus dorsi muscle. Serratus anterior muscle was moved aside to the anterior. Left subclavian artery and LPA were identified. LPA appeared 6 mm in diameter. Heparin was administered at dose 100 IU intravenous (IV). After reaching activated clotting time (ACT) value, the proximal punctum of the polytetrafluoroethylene (PTFE) 5mm tube was anastomosed to the left subclavian artery, end-to-side using a C-clamp. PTFE tube was filled with heparin and clipped. Subsequently, the distal punctum of PTFE tube was anastomosed to the LPA, end-to-side using a C-clamp.

Following anastomosis, SpO_2 levels increased to 75-80% with stable hemodynamics. Bleeding control and evaluation of surgery field are performed after anastomosis. Heparin continuous infusion was administered at dose 10 IU/kg/min. Left pleural drain was

inserted. The surgical wound closed layer by layer.

The patient was transferred to the ICU with hemodynamic heart rate 105 times/minute, arterial blood pressure 84/42 (62) mmHg, central venous pressure (CVP) 8 mmHg, SpO_2 80% with an oxygen fraction 60%, without inotropes. During the initial 24 hours of treatment in the ICU, the patient was sedated due to increased blood flow to the lungs and metabolic acidosis. Metabolic acidosis was corrected through sedation, ventilation control, and administration of inotropes to maintain hemodynamics and oxygen delivery. Metabolic acidosis condition was resolved on the first day of treatment. Patient received continuous intravenous morphine 10 µg/kg/hour for analgesia, followed by paracetamol infusion 100 mg every 8 hours on the second day of ICU care.

Patient was extubated on the second day of ICU care. Postoperative echo cardiography evaluation showed good systolic function, EF 80%; good RV contractility, TAPSE 14 mm; subaortic VSD with pressure gradient (PG) 7 mmHg; L-BT shunt flow PG 22 mmHg. Patient was transferred to the intermediate care unit on the third day of ICU care and was discharged after treatment on the seventh postoperative day.

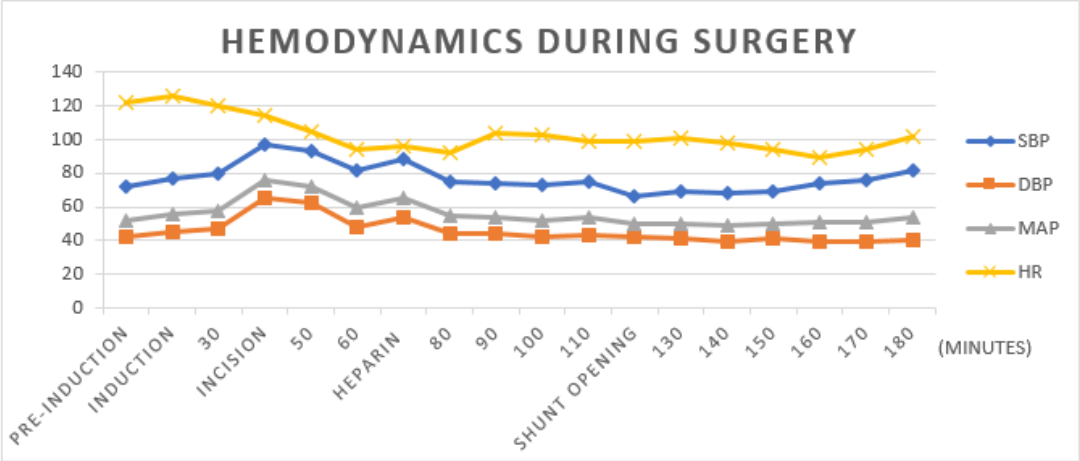

Figure 2. Hemodynamic graph during surgery

Table 2. Blood gas analysis during surgery

Post Induction				Post Anastomosis			
PH	7.29	Lac	1.1	PH	7.33	Lac	1.2
PCO_2	41.1	Hb	18.5	PCO_2	30	Hb	16.3
PO_2	46.2	Hct	57	PO_2	73.1	Hct	49
HCO_3	20.1	Na	137.4	HCO_3	15.5	Na	138
BE	-6.6	K	4.45	BE	-10.6	K	4.00
SpO_2	74.2	Cl	108	SpO_2	91.7	Cl	108
GDS	94	Ca	1.19	GDS	116	Ca	1.14
ACT	136	Mg	0.54	ACT	499	Mg	0.48

Table 3. ICU care day-0

Day-0		12:00-18:00	18:00-24:00	24:00-06:00
Awareness		Sedated	Sedated	Sedated
Hemodynamic	IBP (mmHg)	80-90/50-60	70-80/40-50	70-80/40-50
	HR (x/min)	100-140	130-160	120-140
	Ventilator	PC - FiO2 50%	PC - FiO2 30%	PC - FiO2 21%
	Sat O_2 (%)	70-80	80-90	80-90
Support	Dobutamin (µg/Kg/min)	3	3	3
	Milrinon (µg/Kg/min)		0.375	0.375
Problem		Metabolic acidosis	Acidosis Lung overflow	Acidosis Lung overflow
Pain score	BPS	3	3	3
Sedation/ Analgesic	Morphine (µg/KgBB/hour)	10	10	10

Table 4. ICU care day 1-3

		Day 1	Day 2	Day 3
Awareness		Sedated → aware	Aware	Aware
Hemodynamic	IBP (mmHg)	80-90/40-50	90-100/50-60	80-90/40-50
	BP (x/m)	120-160	100-130	90-120
	Ventilator	PSIMV FiO_2 21%	Nasal prong 2lpm	Nasal prong 1lpm
	Sat O2 (%)	80-85	80-85	70-80
Support	Dobutamin (µg/Kg/min)	3	1.5	-
	Milrinon (µg/Kg/min)	0.375	0.375	-
Problem		Acidosis	-	-
Pain score	FLACC	2	2	2
Analgesic		Morfin 10 (µg/KgBB/hour)	Paracetamol 3x100mg	Paracetamol 3x100mg
Other drugs			Captopril 3x2 mg Furosemide1x10 mg Aspilet 1x35mg	Captopril 3x2mg Furosemide1x10 mg Aspilet 1x35mg

DISCUSSION

Pulmonary atresia with ventricular septal defect (PA-VSD) is a complex lesion of congenital heart defects with morphological variations in the source of pulmonary blood flow. The Baltimore Washington Infant Study reported the incidence of PA-VSD was 0.07 of 1000 live births and 1.5% of all congenital heart defects.[1,6]

The pulmonary blood supply in patients with PA-VSD is maintained by: (1) the presence of retrograde flow from the PDA; (2) major aortopulmonary collateral arteries (MAPCAs) originating from the descending aorta or brachiocephalic trunk and their branches; (3) combination of PDA and MAPCAs. Collaterals may also arise from coronary artery fistula (right or left) to pulmonary artery collaterals, although this is exceedingly rare.[7,8]

The complexity of the pulmonary blood supply determines the type of surgery required to achieve complete repair. Complete surgical repair includes: (1) occlusion and unifocalization of the MAPCAs; (2) reconstruction of the right ventricular outflow tract (RVOT) using a conduit connecting the right ventricle to the pulmonary artery (RV-PA conduit) and; (3) closure of the VSD. In patients who are not eligible for complete repair, palliative surgery is performed by creating a shunt from the systemic to the pulmonary artery.[9,10]

In a systemic to pulmonary shunt, some of the blood from the systemic arteries is oxygenated as it passes through the pulmonary bloodstream and mixes with the unoxygenated venous return bloodstream before flowing into the systemic system. The degree of systemic saturation is determined by the ratio of pulmonary to systemic blood flow and the level of oxygenation of the mixed venous.[11]

An ideal systemic to pulmonary shunt must meet several criteria: (1) it should be technically easy to perform; (2) it should guarantee adequate but not excessive pulmonary blood flow, thus minimizing the risk of congestive heart failure and pulmonary hypertension; (3) shunt patency should be long-lasting; (4) it should be closed easily during definitive correction surgery, and; (5) there are no residual cardiopulmonary abnormalities after shunt closure. Prolonged maintenance of systemic to pulmonary shunts can result alterations in pulmonary vasculature.[2,11]

There are various types of systemic to pulmonary shunts. The BT shunt was the first shunt introduced, which created an anastomosis between the subclavian (or innominate) artery and the pulmonary artery. Systemic to pulmonary shunt types are described in table 5.[11]

BT shunt procedure can be performed at various ages. However, to ensure optimal development of the pulmonary arteries, it is recommended that the BT shunt be performed before the age of 2 years. In neonates aged < 2 weeks and weighing < 3 kg, risk of thrombosis after BT shunt is higher due to the use of a small shunt. Intravenous administration of prostaglandin E1 (PGE1) 0.01-0.1 µg/kg/min aims to maintain ductal patency and can increase ductal diameter. Continuous PGE1 therapy for 2 weeks can increase ductal diameter up to 50% compared to the initial diameter. After 2 weeks of administering PGE1, BT shunt procedure can be performed with a larger shunt size.[2,8,11,12,13]

Modified Blalock-Taussig shunt (mBT shunt) is the most frequently performed systemic to pulmonary shunt procedure. Shunt is created by performing a graft tube (Gore-Tex) anastomosis between subclavian artery and pulmonary artery, through a midline sternotomy or thoracotomy incision. The advantages of mBT shunt include: the subclavian artery is not sacrificed, surgical procedure is easier, the shunt is easier to take down and patency is greater than 90% at the age of 2 years. The disadvantages of mBT shunts are high risk of thrombosis, graft tube leakage, and pseudoaneurysms which can lead to massive hemoptysis.[2]

Table 5. Systemic to pulmonary shunt types[11]

Shunt	Anastomosis	Complications
Classic BT	Right subclavian artery to right PA (with left aortic arch)	Kinking of right PA, right arm ischemia, excessive pulmonary flow
Modified BT	Tube graft from right (or left) subclavian artery to right (or left) PA	Kinking of right PA, tube graft does not allow for growth, chylothorax
Waterston	Ascending aorta to right PA	Kinking of right PA, amount of blood flow difficult to control
Potts	Descending aorta to left PA	Rarely used, difficult to control shunt size, difficult takedown at later repair
Central (aortic-pulmonary shunt)	Anastomosis between aorta and PA with tube graft	Distortion of main PA, excessive pulmonary flow

Abbreviation: BT: Blalock-Taussig; PA: Pulmonary Artery

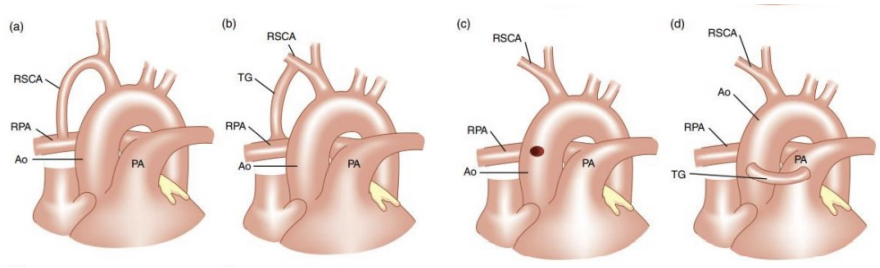

Figure 3. Systemic to Pulmonary Shunt Types (a) Classic Blalock-Taussig shunt (b) modified Blalock-Taussig shunt with Tube Graft (c) Waterston shunt (d) Central shunt with Tube Graft

Abbreviation: RSCA: Right Subclavian Artery; RPA: Right Pulmonary Artery; PA: Pulmonary Artery; Ao: Aorta; TG: Tube Graft

Preoperative Management

Anamnesis regarding the history of disease must be explored in depth. This includes the symptoms, their onset, the presence of cyanosis, shortness of breath, difficulty eating or drinking milk, growth and developmental disorders, the presence of other comorbidities, as well as a history of procedures and treatment. It is important to note that patients who have had a previous BT shunt may be on aspirin, warfarin, diuretics, ACE inhibitors, and antiarrhythmic drugs. Neonatal patients with ductal dependent receive continuous intravenous PGE1 therapy 0.01-0.1 µg/kg/min to maintain ductal patency.[2,8,11,12]

PA-VSD is frequently associated with extracardiac abnormalities and other chromosomal anomalies. Most often accompanied by microdeletions at 22q11, also known as DiGeorge syndrome. This syndrome present with various symptoms, including palate abnormalities such as cleft palate. This is particular concern during the induction of anesthesia, intubation and postoperative extubation.[14]

The clinical presentation of PA-VSD depends on pulmonary blood supply. Symptoms typically appear on days 3-7 after birth in patients with PDA without MAPCAs. This is due to constriction and closure of the ductus, which causes a decrease in pulmonary blood flow resulting in cyanosis, severe hypoxia, acidosis, and shock. This condition can also occur in patients with MAPCAs stenosis, with or without PDA.[8,15]

Patients with multiple and large MAPCAs will have adequate oxygenation. Overcirculation may occur due to large pulmonary blood flow. Patients typically present with clinical symptoms of heart failure, pulmonary edema, eating/drinking disorders, and growth and developmental disorders after 4-6 weeks of age. This is due to a decrease in pulmonary vascular resistance (PVR) at age 4-6 weeks, resulting in excessive pulmonary blood flow.[8,14]

Patients with balanced Qp/Qs and stable oxygenation will persist this condition until adolescence and the third decade. They may experience mild cyanosis, pulmonary vascular disease, and potentially develop Eisenmenger syndrome.[4,16]

Pysical examination shows central cyanosis, weak crying movements, lethargy, growth and developmental disorders and eating/drinking disorders. Holodiastolic murmurs may be audible from the left parasternal to the axilla and back. Murmur associated with PDA is described as a mechanical sound in the upper chest or interscapular region. In cases of congestive heart failure, symptoms such as extremity edema, clubbing fingers, and severe cyanosis may be present.[15,17]

This case report describes a one-year-old patient who presented with symptoms of central cyanosis, growth disorders, and difficulty breastfeeding. Patient experience spells when pulmonary blood flow decreases due to constriction of the PDA, while blood flow to the lungs still occurs through the MAPCAs.

Supporting Investigation

Routine blood tests show polycythemia, which increases blood viscosity and can cause thrombus. White blood cells and other markers of infection (C-reactive protein, procalcitonin) may be increased in respiratory tract infections. Preoperative phlebotomy can be performed in patients with symptomatic

hyperviscosity and hematocrit levels above 65%. Prior to conducting phlebotomy, it is crucial to evaluate the patient hydration status and correct any dehydration condition.[2,15]

Coagulation factor abnormalities may occur due to platelet dysfunction, hypofibrinogenemia or clotting factor deficiency. Serum electrolytes should be evaluated especially in patients receiving diuretic therapy.[2,15]

Chest x-rays can be used to assess the position and size of the heart, the presence of atelectasis or lung infection, pulmonary bronchovascular patterns and hemidiaphragm enhancement.[8,15]

Echocardiography is essential for diagnosing congenital heart disease. Echocardiography can identify anatomical abnormalities in the heart, visualize blood flow using color Doppler and evaluate the presence of PDA.[6,15]

Cardiac catheterization is performed to complement the echocardiography findings. Cardiac catheterization can provide information about coronary arteries, MAPCAs (including number, size, distribution, presence of stenosis and blood pressure of each collateral vessels), confirmation the presence of PA, lung arborization, lung segments associated with PA and lung segments that have dual circulation. Computerized tomography (CT)/magnetic resonance (MR) angiography is an alternative modality for evaluating the RVOT, MPA, PA branches and MAPCAs.[8,15]

Prior to surgery, the patient's family is educated on the importance of fasting and blood component preparation. The anesthesia procedure is explained, including the associated risks, side effects, and complications, as well as care and administration of postoperative analgesia in the ICU.[2]

Preoperative fasting scheduling should be written clearly in accordance with ASA guidelines. Dehydration should be avoided in patients with cyanotic heart disease. The surgery schedule should not be postponed. If the schedule is not clear, it is necessary to establish intravenous access and provide replacement fluids for fasting.[2,18]

Intraoperative Management

Preinduction preparation

Premedication can be given to children aged > 6 months to reduce anxiety, so the children are more cooperative when separated from their parents. The drug of choice is midazolam which can be administered orally (0.5 mg/kg) or intravenously (0.1 mg/kg) if intravenous access is available. Premedication with ketamine can also be given at dose 5-10 mg/kg orally in patients with decreased heart function. In this case report, the patient was given midazolam 1 mg intravenously. The administration of premedication is carried out by an anesthesiologist and the patient must be continuously monitored.[2,3,11,18]

Induction of anesthesia

The induction of anesthesia aims to balance flow, increasing pulmonary blood flow and maintaining systemic circulation. Pulmonary blood flow is increased by reducing PVR through the provision of high concentrations of oxygen, hyperventilation, and prevention of metabolic acidosis. SVR is maintained within normal values and avoid increasing the PVR:SVR ratio to maintain systemic circulation. Target PaO_2 40-45 mmHg and SpO_2 70-80% indicate adequate systemic oxygen delivery.[11,12,18]

Sevoflurane inhalation can be used for induction if intravenous access has not been placed. Balance anesthesia techniques, a combination of narcotics, inhalation agents and muscle relaxants are most often used. Patients with severely impaired cardiac function and using inotropes preoperative may not tolerate the vasodilatory effects of anesthetic agents. Alternative induction drug for these patients is ketamine 1-2 mg/kg and muscle relaxants.[2,3,12,18,19]

In this case report, inhalation induction with sevoflurane was performed as intravenous access had not yet been established. Once the access was established, rehydration fluid 10ml/ kgBB was administered to replace fasting. Balanced anesthesia technique was used for induction, with titration of fentanyl and vecuronium. No significant hemodynamic changes occurred during induction. Following induction, the patient's oxygen saturation increased to 60% with 100% oxygen fraction.

It must be ensured that there are no air bubbles in the infusion line when administering the drug. Air bubbles can flow into the systemic circulation through the shunt and cause embolism in patients with a right-to-left shunt.[3]

Standard cardiac surgery monitoring includes ECG, pulse oximetry, arterial line, central venous pressure, End Tidal Carbon Dioxide ($ETCO_2$), core temperature, blood gas analysis (BGA), ACT, and urine output.

An arterial line is placed in the radial artery opposite the planned shunt position to obtain correct measurement of blood pressure, because once the shunt is opened there may be a significant amount of blood flow stolen from the ipsilateral subclavian artery. An arterial line can be placed in the femoral artery and signs of ischemia in the lower extremities should be observed.[2,3,16]

Anesthesia maintenance

Maintenance anesthesia was performed using inhalation agents. The ideal inhalation anesthetic agent for patients with congenital heart defects must be able to maintain cardiac output by preserving cardiac contractility, heart rate and heart rhythm. The effect on SVR is expected to be minimal so that the mean arterial pressure (MAP) and coronary perfusion pressure (CPP) can be maintained. PVR should be reduced in patients with reduced blood flow to the lungs.[16]

This case reports described maintenance anesthesia using the inhalation agent sevoflurane 1-2 volume%, intermittent fentanyl and intermittent vecuronium. Low-dose sevoflurane has minimal cardiac depressant effects and does not decrease SVR.[20]

BT shunt surgery is performed via thoracotomy approach, using a retractor to obtain a good surgical field. Intermittent lung reinflation is necessary to prevent disruption of oxygenation and ventilation caused by lung retraction. This could be worse if the patient has experienced recurrent lung infections which cause increased airway pressure and low saturation due to pulmonary congestion or bronchospasm. Hyper-ventilation and high fraction of inspired oxygen (FiO_2) to maintain oxygen saturation and $ETCO_2$ may be necessary at this stage.[16]

Hypoxia is a potential risk during partial clamping of the MPA, due to increased PVR, pulmonary vein desaturation secondary to collapsed lung, hypoxic pulmonary vasoconstriction, and

atelectasis. Bradycardia can result from hypoxia. Close monitoring of blood pressure, heart rate and oxygen saturation is required.[12,16]

During partial clamping of MPA, the $ETCO_2$ value does not reflect the actual $PaCO_2$ value because pulmonary artery blood flow decreases. The BGA examination can be used as an evaluation during this procedure.[12,16]

Median sternotomy incision is performed if the patient cannot tolerate lung retraction or side-clamping of PA, hemodynamics during surgery is unstable and there is a possibility that cardiopulmonary bypass (CPB) machine is needed. PDA ligation is more difficult when using the sternotomy approach.[1,16]

Opening the shunt anastomosis leads to an increase in oxygen saturation. Systemic blood flow filling the shunt and pulmonary circulation can cause a decrease in blood pressure, which may require additional volume or vasopressor support. Inotropes can be administered to maintain cardiac contractility, particularly if PDA ligation is not performed.[1,16]

MAP decrease >10 mmHg indicates that the shunt is too large. Excessive pulmonary blood flow can lead to systemic hypotension, pulmonary edema, and metabolic acidosis. Ventilator weaning difficulties may also occur.[11,18]

Once the shunt anastomosis is completed and opened, the lung must be expanded and reinflated to prevent atelectasis. Optimal oxygen saturation after BT shunt is 75-80% which reflects the balance of pulmonary and systemic blood flow (Qp:Qs = 0.7-1.5:1). High oxygen saturation >85% indicates pulmonary overcirculation (Qp:Qs > 2:1), which is followed by a decrease in systemic blood pressure due to significant blood flow being stolen from the subclavian artery into the pulmonary circulation. A decrease in diastolic blood pressure will cause coronary circulation disorders. ECG evaluation is important to evaluate the presence of coronary circulation disorders which can be seen from changes in ECG waves and rhythms.[1,16,18]

Low saturation <70% indicates inadequate pulmonary blood flow, Qp:Qs < 0.7:1. This can be caused by high PVR, small shunt size or low cardiac output syndrome (LCOS). It is important to identify the cause of persistent hypoxemia that occurs after the shunt is opened and to exclude causes of hypoxemia from airway and breathing problems.[16,18]

Briefly removing the patient from the ventilator and auscultation of the end of the endotracheal tube can clinically confirm the patency of the shunt. The shunt murmur sound is transmitted through the tracheal tube because the shunt is located close to the bronchus. TTE can be used to assess the function and flow of the shunt.[12,16]

Anticoagulant Management

Heparin bolus is administered during surgery before arterial clamping for anastomosis, at a dose of 100-150 IU/kg intravenous bolus. ACT is maintained over 250 seconds. Postoperative heparin should be administered as soon as possible. Heparin continuous infusion should be administered within 4 hours after surgery if bleeding from the drain is less than 3ml/kg/hour.[2,16,21,22]

Table 6. Anesthesia Management During Operation[14]

Clinical signs	Condition	Intervention
↓ SpO_2 Hypotension ↓ NIRS	Low CI	Inotropy Volume
↓ SpO_2 Normal BP	Normal CI with relatively ↑ PVR	↓ PVR Optimize ventilation ↑ FiO_2 Analgesia Consider iNO ↑ SVR
↑ SpO_2 Hypotension	Normal CI with relatively ↓ PVR	↑ PVR Optimize ventilation Reduce FiO_2 ↓ SVR
↑ SpO_2 Normal BP Normal NIRS	High CI	Consider deepening anesthetic and additional analgesia

Abbreviation: SpO_2: Saturation of peripheral oxygen; CI: Cardiac Index; NIRS: Near-Infrared Spectroscopy; PVR: Pulmonary Vascular Resistance; SVR: Systemic Vascular Resistance; FiO_2: Fraction of Inspired Oxygen; iNO: Inducible Nitric Oxide; BP: Blood Pressure

Heparin is given continuously with an initial dose 10 IU/kg/hour. Monitoring during heparinization includes coagulation profile activated partial thromboplastin time (aPTT), prothrombin time (PT), fibrinogen, and platelet values. The aPTT value was evaluated 4 hours after the initial administration of heparin and evaluated daily. The desired therapeutic target is aPTT 60-90 seconds. If the aPTT <50 seconds, heparin is given intravenous bolus at dose 50 IU/kg over 10 minutes.[2,22]

Administration of aspirin can be started when the intrapleural drain and pacing wire (if any) have been removed. The initial dose of aspirin is 3-5 mg/kg (maximum 75 mg) per day. Continuous administration of heparin is continued until the second dose of aspirin is given.[2,22]

The incidence of thrombosis after BT shunt procedure is 12%. Risk factors for thrombosis are neonatal patients, body weight less than 3 kg, polycythemia with preoperative hemoglobin >18 gr/dl, the use of a small shunt size, dehydration, and platelet transfusion. Shunt obstruction can be caused by strictures in the anastomosis area, neointimal proliferation, and vascular stenosis due to surgery. Anticoagulant management is an important factor in the success of BT shunt procedure. Platelet transfusions should be avoided in patients with shunts because of the risk of thrombosis.[,21,23,24]

In this case report, continuous heparin infusion was administered from the operating room after protamine was given. Heparin was start at dose 10 IU/kg/hour, considering that bleeding during surgery was minimal and could be well-controlled. Evaluation of

coagulation factors was performed postoperative in ICU.

Postoperative Management

Pediatric patients can be extubated immediately after completing the surgical procedure. However, in neonatal patients, it is recommended to maintain mechanical ventilation for one day and administer diuretics. The patient should be transferred to the ICU and kept on ventilator for 12-24 hours. Postoperative pain is managed by providing continuous narcotic analgesics.[2,11]

Gupta et al studied the incidence of failed extubation in neonates after BT shunt. They found that 27% of patients required reintubation or non-invasive ventilation within 96 hours. Changes in PVR, SVR and Qp that occur during lung reexpansion combine with emergence from anesthesia, transition to analgesic administration and the possibility of postoperative bleeding contribute to the occurrence of postoperative cardiac arrest.[12,21]

Postoperative complications must be closely monitored for early detection and appropriate treatment. There may be increased blood flow to the lungs or decreased blood flow to the lungs.[18]

A BT shunt size that is too large will cause excessive blood flow to the pulmonary circulation. Diagnosis can be seen from high oxygen saturation, systemic hypoperfusion, low blood pressure, left heart failure followed by signs of right heart failure, pulmonary edema, decreased central venous or mixed vein saturation, decreased splanchnic and cerebral perfusion (on NIRS), increased lactate and increased base deficit.[2,18]

Increased pulmonary blood flow can cause unstable hemodynamic conditions. Diastolic hypotension can lead to myocardial ischemia which requires close monitoring and prompt treatment.[16,18]

Management of overcirculation involves fluid restriction, administration of diuretics and manipulation of PVR and SVR. PVR is increased by decreasing the inspired fraction of oxygen, increasing PEEP, and hypoventilation to increase $PaCO_2$ (reducing minute ventilation). The SVR was reduced by considering gradual decrease in the vasopressor dose. Overcirculation often coincides with LCOS so inotropes may be required. Consider surgery to reduce the size of the shunt if low diastolic pressure is affecting the coronary circulation.[2,16]

In the presented case, the patient was sedated with controlled ventilation for the first 24 hours in the ICU due to metabolic acidosis and increased pulmonary blood flow. Increased pulmonary blood flow can be seen from an increase in SpO_2 >85% accompanied by a decrease in blood pressure. The oxygen fraction was reduced to 21% and afterload reduction was given.

Preoperative laboratory examination revealed acidosis had occurred before the surgery. Repeated spells and low PaO_2 resulting in anaerobic metabolism in patients. Decreased haemodynamics during induction and opening of the anastomosis may exacerbate acidosis. Acidotic conditions cause an increase in PVR and reduce pulmonary circulation. Metabolic acidosis can be corrected through sedation, ventilation control, administration of inotropes and volume to maintain hemodynamics and oxygen delivery.[18]

Postoperative desaturation may occur due to increased PVR, low cardiac output state, small shunt size or shunt failure due to clotting or kinking. PVR is reduced by providing sedation and full ventilation control to reduce $PaCO_2$, increase oxygen fraction, and re-expand the lung if atelectasis occurs.[2,18]

Shunt failure may occur in the early postoperative period. The visible sign is an acute decrease in oxygen saturation and no murmur heard from the shunt. This could be caused by clotting or kinking in the shunt. This condition is an emergency and requires immediate intervention.[2,18]

Coagulation profile is evaluated to determine therapeutic targets. Non-surgical intervention by giving intravenous heparin bolus 50 IU/kg, followed by continuous infusion 20 IU/kg/hour, or increasing the rate 10% if continuous heparin has been given. Prostaglandin administration may be considered if PDA has not been ligated. Continuous intravenous PGE1 to maintain ductus patency is given at a dose of 0.01-0.1 µg/kg/min.[16]

If the oxygen levels continue to decrease despite non-surgical intervention, it may be necessary to repeat the surgery to evaluate the shunt patency and adjust its size. In this case, BGA evaluation indicates a trend of worsening acidosis, with a decrease in PaO_2 values and an increase in $PaCO_2$ levels, accompanied by an increase in lactate levels. It is important to note that these findings suggest a need for further medical attention.[2,18,25]

Other complications of BT shunt procedure are trauma or damage to the phrenic nerve and recurrent laryngeal nerve, Horner's syndrome, chylothorax and shunt thrombosis.[2,16]

CONCLUSION

BT Shunt is the first line management for patients experiencing critical cyanotic conditions. Understanding the physiology of heart defects and perioperative management determine the success of BT shunt procedure, reducing patient morbidity and mortality.

The main principle in BT-shunt management is optimizing intra operative and postoperative oxygen delivery by maintaining adequate preload, myocardial contractility, heart rate, SVR and reducing PVR. Close monitoring includes ECG, arterial blood pressure, heart rate, $ETCO_2$, peripheral oxygen saturation, core body temperature, BGA before and after BT shunt, ACT values before and after BT shunt, and postoperative haemostatic function.

Optimizing oxygen delivery with oxygenation targets PaO_2 40-45 mmHg and saturation 70-80% reflects the balance of pulmonary blood flow and systemic blood flow (Qp:Qs=0.7-1.5:1). Postoperative management includes administration of anticoagulants and close monitoring for postoperative complications.

Table 7. Management of postoperative complications[6]

Clinical presentation	Physiology	Management
SaO_2 75-80% Sa-vO_2 25-30% BP > 60/30 mmHg	Balanced flow Qp:Qs = 0.7-1.5:1	No intervention
SaO_2 >85-90% Sa-vO_2 35-40% BP < 60/30 mmHg Diastolic BP < 15-25 mmHg with mBTS; likely higher with RV-PA conduit	High pulmonary blood flow Qp:Qs > 2-3:1 Causes: Low PVR Large mBTS or RV-PA conduit Residual arch obstruction High SVR	Raise PVR: Controlled hypoventilation Mild Acidosis Low FiO_2 (0.17-0.19); compromised cerebral O_2 delivery Increase systemic O_2 delivery: Afterload reduction Hematocrit > 40% Surgical intervention: Clip mBTS or RV-PA conduit Revise arch reconstruction
SaO_2 <65-75% Sa-vO_2 25-30% but SvO_2 likely less than critical value of 30% BP > 70/40 mmHg Diastolic BP > 40 mmHg	Low pulmonary blood flow Qp:Qs < 0.7:1 Causes: High PVR Small mBTS or RV-PA conduit Pulmonary venous desaturation with underestimation of actual Qp:Qs	Lower PVR: Controlled hyperventilation Alkalosis Sedation/paralysis Aggressively treat atelectasis (pulmonary venous desaturation) Consider NO Increase systemic O_2 delivery: Inotropic support Surgical intervention: Revise mBTS or RV-PA conduit
SaO_2 <70-75% Sa-vO_2 35-40% and SvO_2 likely less than critical value of 30% BP <60/30 mmHg	Low cardiac output Causes: Ventricular dysfunction • Myocardial ischemia • Depressed contractility • Afterload mismatch (residual arch obstruction) • AV valve regurgitation	Minimize O_2 consumption: Sedation/paralysis Inotropic support/afterload reduction Surgical intervention: Repair AV valve Revise arch Consider mechanical support: • Post-cardiotomy support • Bridge-to-transplantation

Abbreviation: SaO_2: Saturation of arterial oxygen; SvO_2: Saturation of venous oxygen; Sa-vO_2: Saturation of arterial-venous oxygen difference; BP: Blood Pressure; Qp: Pulmonary blood flow; Qs: Systemic blood flow; PVR: Pulmonary Vascular Resistance; SVR: Systemic Vascular Resistance; O_2: Oxygen; FiO_2: Fraction of Inspired Oxygen; mBTS: modified Blalock-Taussig Shunt; RV-PA: Right Ventricle – Pulmonary Artery; NO: Nitric Oxide

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