Anesthetic management of omphalopagus twins with pentalogy of Fallot undergoing separation surgery

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ABSTRACT

The management of conjoined twins has several special aspects, mostly linked to the parents of the twins, such as communication, ethics and trust issues. A multidisciplinary team approach plus high level of vigilance at each perioperative stage is required. Conjoined omphalopagus twins, aged one year, were planned for separation surgery. They had fused liver, fused portal and hepatic veins, and partially fused intestines. Patient B appeared with complications of pentalogy of Fallot (PoF). The patient also had polycythemia (hemoglobin 14.7 g/dL, hematocrit 50%). Radiological features showed anterior abdominal wall with images of interconnected intestinal system between the two. In thoracoabdominopagus conjoined twins, the possibility of shared circulation becomes important especially associated with injection of intravenous drugs. The goal of anesthesia in PoF is to maintain cardiac output by maintaining heart rate, contractility and preload; and preventing right to left shunting via ventricular septal defect (VSD). Induction can cause vasodilation and the resultant decrease in systemic vascular resistance (SVR) can exacerbate right-to-left shunts. Monitoring end tidal CO₂ (EtCO₂) is needed. We present a report of anesthesia for such a case.

Abbreviations: ASD: Atrial Septal Defect; PoF: Pentalogy of Fallot; VSD: Ventricular Septal Defect; SVR: Systemic Vascular Resistance

Key words: Conjoined Twins; Pentalogy of Fallot; Congenital Heart Disease; Cyanosis; Anesthesia


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1. INTRODUCTION

Perioperative preparation in separation surgery of the conjoined twins needs a multidisciplinary approach and needs a high level of vigilance at each perioperative stage. Problems that can arise during surgery such as bleeding, hypothermia, hypoglycemia, hypoxia, hypercarbia, acidosis, electrolyte imbalance, and coagulopathy, should always be anticipated, and treated appropriately if these happen in the perioperative period.

The conjoined twins must be treated as two different individuals, even though there are parts of their organs that are attached or shared.¹

Pentalogy of Fallot (PoF) is a form of tetralogy of Fallot (TOF) with additional atrial septal defect (ASD) and ventricular septal defect (VSD), overriding aorta, pulmonary artery flow obstruction, and right ventricular hypertrophy. Prevalence of TOF in congenital heart disease is about 10% with an incidence of 3 in 10000 births. However, there are still no global epidemiological
reports on PoF. It’s a challenging task to treat such cases because of the hemodynamic variations that may occur perioperatively. This report presents a case of anesthetic management in one-year-old conjoined twins with PoF who underwent surgical separation.

2. CASE REPORT

A pair of conjoined babies, aged one year was planned for omphalopagus separation surgery (Figure 1). Patient B had been diagnosed to have congenital heart disease – PoF, at 3 months of age. The patient A had no comorbidities. The twins were born by cesarean section at another hospital with 8 months of gestational age, birth weight 2000 grams each. At 3 months, the child B began to experience blue discoloration of fingers, toes, and mouth; sweating; and looked tired when crying or feeding. There was no history of spells or lower respiratory tract infection. He failed to thrive till the time of his preoperative examination.

Figure 1: Pre-surgical condition of the twins. Baby A (right) and Baby B (left)

2.1. Assessment

On examination, the patient was fully conscious and active, mesocephalic head circumference with concave part on the left temporalis. Chest examination revealed no additional sounds in both lungs, symmetrical chest expansion with normal stem fremitus, and no chest wall retractions. On cardiac examination, a pansystolic murmur at IV-V right parasternal area was heard. He had warm extremities. Fingers and toes showed clubbing (Figure 2), the dorsalis pedis artery and the brachial artery pulses were regular and adequate. Abdominal and urogenital systems were within normal limits.

Blood examination showed polycythemia with hemoglobin at 14.7 g/dL and hematocrit as 50%. Babygram picture showed the anterior abdominal wall with a shadow of the interconnected intestinal system, suspected omphalopagus, and dextrocardia. No pulmonary abnormalities were seen. On abdominal ultrasound, it was found that the liver was fused between patient A and B, with fused portal and hepatic veins, partially fused intestines, and no abnormalities on other intra-abdominal organs.

Figure 2: The baby B had cyanosis finger tips and toes.

2.2. Investigations

Echocardiography showed situs inversus, atrioventricular (AV) concordance, ventriculoarterial (VA) discordance, apex located on the right, II with 0.4 cm of diameter, VSD with 50% overriding aorta, PDA, moderate pulmonary stenosis (PS) with 43.25 mmHg of pressure gradient. No aortic arch to the left of Koar. It
Figure 3-A: Echocardiography concluded Pentalogy of Fallot with PDA in this patient.

was concluded that there was a pentalogy of Fallot with PDA in this patient (Figure 3-A).

Multi-sliced CT angiography showed the liver of patient B fused with the liver of patient A, with joined vasculature, partially fused intestines with no vasculature of patient A. There was sublobar pneumonia in the posterobasal segment of the right inferior pulmonary lobe, dextrocardiac with situs inversus, gall bladder agenesis, right hypochondriacal spleen (ultrasound confirmed), and bladder dilatation. However, no intrahepatic mass was seen (Figure 3-B).

Figure 3-B: Multi-sliced CT angiography showed the conjoined twins several abdominal organs fusion

Esophagography examination showed that the intestinal cisterns of patient B didn't go into the abdominal cavity of patient A (Figure 2C).

Figure 3-C: Esophagography showed that B's intestinal cisterns didn't fuse A's abdominal cavity.

2.3. Anesthetic Management

Consent by the family was obtained, both patient A (baby with no comorbidities) and B (baby with PoF) received premedication: midazolam 0.1 mg/kg, ketamine 1 mg/kg, and atropine sulphate 0.01 mg/kg IV and were separated from the family. The standard monitors were attached (Figure 4).

2.3.1. Patient A

Patient A vital signs before induction were: blood pressure 106/78 mmHg, heart rate 134 bpm and oxygen saturation at 97%. Induction was done with 2 vol% of sevoflurane in 65% O<sub>2</sub> with hyperventilation until the EtCO<sub>2</sub> reached at 30–35 mmHg. Fentanyl 2 µg/kg was given as an analgesic for blunting responses. Soon after the induction, atracurium 0.5 mg/kg was injected IV prior to intubation with endotracheal tube No. 3.5. Arterial line was passed via No. 24 IV cannula in the right brachialis artery and CVP line in right femoral vein. Foley catheter was passed to monitor urine output.

Anesthesia for patient A was maintained with controlled ventilation of 2% sevoflurane in 65% oxygen, and infusions of 1 µg/kg/h morphine and 5 µg/kg/min atracurium. He remained stable throughout the surgery which lasted for 3 h 15 min. His pulse rate remained in the range of 120–130 beats/min, systolic BP in the range of 96–110 mmHg, SpO<sub>2</sub> 97–99%, and EtCO<sub>2</sub> of 30–35 mmHg. Intraoperative bleeding was 40 ml and was replaced with 10 ml of packed red cells. Fluid was
Intraoperative bleeding was 40 ml and replaced with 10 ml packed red cells. Fluid was infused at 32 ml/h. Sixty minutes before the end of the operation, he had a hypotensive spell. An infusion of epinephrine 0.05 µg/kg/min was started and he was improved. We continued infusions of dopamine 3 µg/kg/min, epinephrine 0.05 µg/kg/min, and morphine 10 µg/kg/min. Post-operatively he remained intubated, and was planned for weaning in the ICU.

2.4. Post-surgery

The patient A returned to full consciousness and was extubated in the first hour of stay in the ICU, while the patient B was extubated on 2nd day and transferred to the ward on day 8 postoperatively. For pain management ketamine 0.15 µg/kg/min was infused for 24 h and 20 mg/kg metamizole every 6 h postoperatively as analgesics for both patient A and B.

3. DISCUSSION

The separation of conjoined twins requires two anesthetic teams. One team for each child, with at least two anesthesiologists, four plastic surgeons, two pediatric surgeons, two cardiothoracic surgeons, five pediatricians, two intensive-consultant anesthesiologists, two anesthesia equipment units, two monitoring units, drugs, and two teams of anesthetic nurses. Plans start with the admission process in the handover room until the post-operative period in the intensive care unit (ICU).

In the case of thoracoabdominopagus conjoined twins, we must consider the possibility of shared circulation. This is important and associated to the use of induction and maintenance drugs, and also related to the difference rate of circulation between the two fetuses after birth due to differences in metabolic rates. In this case, no shared circulation was found so there was no influence of anesthetic drugs from the first infant who was induced earlier, and the second infant was not anesthetized, during the multi-slice CT angiography and then it was supported by the results of the CT angiography examination.

Anesthesia procedures for the diagnosis and management of twins are always performed by two different anesthetic teams. The operating room should be...
equipped with two anesthesia machines, two breathing equipment sets and monitors. Before the initiation of anesthesia, a physician must determine which baby will be anesthetized first (based on the condition of the babies) and what induction and intubation methods will be used. Induction should be performed on the easier and healthier baby first.

History taking in children with PoF is needed to assess the severity of abnormalities. From the history it was known that the heart condition of baby B was still compensated, characterized by normal activity, no history of cyanotic extremities except when he was crying loudly. He also suffered chronic hypoxia, characterized by clubbing, higher hemoglobin and hematocrit values than normal.

Echocardiography examination provides fairly complete information and the conclusion of this case was pentalogy of Fallot with PDA (+). Heart apex showed on the right, ASD II with a diameter of 0.4 cm, moderate PS, and overriding aorta. Important points that should be obtained by an anesthesiologist is the degree of stenosis, the presence of collateral vascularization that allows blood flow from the heart to the lungs, the presence of a patent ductus arteriosus (PDA), the severity of pressure gradient and the direction of intracardiac bypass flow. This patient had a VSD with 50% overriding aorta, moderate PS with a pressure gradient of 43.25 mm Hg, which included as moderate to severe stenosis.

The patient was planned for elective surgery, previously been hospitalized for 2 days and had been thoroughly prepared for omphalophagus separation surgery. From the preparations we carried out, his general condition was moderate, he was quite active, cried well, no shortness of breath, cough or fever. We gave intravenous fluids on ward to avoid dehydration and the more important thing was keeping this patient calm, as crying could trigger the cyanotic attack.

We chose general anesthesia for this patient, using no 3.5 cuff of endotracheal tube with consideration to avoid malposition during movement changes with pronation and supination positions, and breath control. A 0.4 mg midazolam was given as a premedication. This aimed to provide sedation with an onset of about 30–60 sec. This dose of midazolam does not depress ventilation. The induction dose of 0.1–0.2 mg/kg midazolam can make a depressant effect on the central nervous system.7

The goal of anesthesia in PoF cyanotic heart disease is to maintain cardiac output by sustaining heart rate, contractility and preload; preventing an increase of pulmonary vascular resistance: systemic vascular resistance (PVR: SVR) ratio to limit right to left shunt via VSD. Patient with PoF is prone to develop infundibulum spasm which can cause hypoxemia (tet spells). This is caused by increased sympathetic activity and cardiac contractility in conditions of crying, being agitated, feeling fear, pain, trauma, any inadequate anesthetic levels, and seizures. This can be avoided by giving premedication before separating the patient from the family.2,8,9

Anesthesia induction can cause vasodilation and the resultant of decreasing SVR can exacerbate right-to-left shunts. We induced anesthesia with ketamine to maintain SVR. Ketamine as an induction agent has been shown to increase oxygenation by decreasing right-to-left shunts as a result of increased SVR. It has advantageous effects on children with CHD and severe pulmonary hypertension by preserving SVR and ventricular performance without increasing pulmonary vascular resistance.10 The cerebral vasodilation effects of ketamine can be reduced by hyperventilation or in combination with benzodiazepines, and exacerbated by hypercapnia. Therefore, monitoring EtCO2 needs to be considered. It often goes further below the alveolar CO2 pressure value because of the higher dead space in patients with cyanotic CHD, especially in patients with multiple cardiac lesions with increased shunt flow.11

Sevoflurane is a drug of choice because it has the least effect on SVR. Isoflurane is a poor choice due to vasodilation and tachycardia effects.2,8

We used atracurium as paralytic agent for this case. Atracurium is a non-depolarizing muscle relaxant drug as its pharmacokinetics are independent of kidney and liver function.12,13 This muscle relaxant is used to facilitate intubation, provide relaxation in the operating field during surgery, and facilitate mechanical ventilation.

Significant histamine-releasing effects of atracurium may occur in sensitive patients. With an initial dose of 0.5 mg/kg the increase in plasma histamine levels was around 15% but hemodynamics remained stable. This initial dose should be lowered from 0.4 to 0.3 mg/kg and given steadily or given in divided doses especially on patients with a history of significant heart disease, given the possibility of a significant reduction in blood pressure in such patients. Adilah Miraj et al. reported bradycardia followed by cardiac arrest shortly after atracurium administration.14

Management of possible right ventricular dysfunction is with fluid to increase preload, additional inotropes, and reduction of right ventricular afterload. Dosage of dopamine in the range of 3–10 µg/kg/min has adequate inotropic effect for patients with TOF. Doses > 15 µg/kg/min should be avoided because they can cause vasoconstriction and chronotropic effects. Low dose adrenaline (0.01–0.03 µg/kg/min) gives effect of beta-adrenergic receptor agonists domination. Higher dose up
to 0.1 g/kg/min can cause predominant vasoconstrictive effect on alpha-adrenergic receptors. Dose of 3 \( \mu g/kg/min \) dopamine and 0.03 \( \mu g/kg/min \) adrenaline were given to assist right ventricular function and reduce PVR by maintaining BP in range of 90/40 mmHg to 100/52 mmHg. We did not increase the dose of adrenaline and dopamine to avoid unwanted tachycardia and vasoconstriction.

This patient had polycythemia due to hypoxemia which could cause high viscosity and risk of thrombosis. High blood viscosity results in decreased cerebral blood flow. This patient had low platelets so he had a risk of bleeding during surgery. Adequate rehydration before induction and blood replacement were important to prevent worsening of the right-to-left shunt and to maintain adequate cerebral perfusion pressure. In general, preoperative hematocrit value is maintained to ensure adequate oxygen delivery.

Blood loss estimation during surgery in the case of conjoined twins can be so difficult. This is because when they are still attached, the amount and origin of bleeding are not clearly known. The easiest way to do is calculating blood loss is to measure the hemoglobin levels of each baby at the time of separation and then after being separated. Measurement of blood loss was based on the amount of blood aspiration at the surgical site, by suction machine, amount and weight of the gauzes used, and the values of hemodynamic parameters: hematocrit, and hemoglobin in both infants. The amount of blood transfused in each child was half of the total estimated blood loss that occurred during surgery.

In conjoined twins with abdominal attachment, surgeon must consider the intrathoracic pressure changes at the time of peritoneum and skin closure. The increase in intrathoracic pressure can occur because the volume of abdominal cavity in the newly-separated baby is not as large as when they were conjoined. As a result, lung compliance will decrease and cause ventilation problems.

After successfully extubating, adequate oxygenation and ventilation should be closely observed in the ICU to avoid the hemodynamic consequences of hypoxia and hypercarbia. Even mild hypoventilation will produce hypoxic vasoconstriction and increase PVR. Children with TOF have a low tolerance even just mild hypoventilation. In this case, the patient was not extubated after the surgery completion so he was admitted to ICU and programmed to gradually wean from ventilatory support. Supplemental oxygen requirement after extubation is based on the child's physiology and the SaO\(_2\) obtained in room air after extubation.

The important points of postoperative period are preventing an SVR decrease and sympathetic tone increase, by avoiding any acidosis, hypoxia, hypercarbia and pain. Pain must be addressed immediately because it can trigger infundibulum spasm which causes a hypercyanotic spell. FLACC (Face, Leg, Activity Cry, Consolability) scale has been used to assess pain in children from infancy to adolescence under varying circumstances. FLACC score for this patient at the first 24 h was 2 (mild pain). Postoperative analgesia with 20 \( mg/kg \) metamizole every 8 h and 0.15 \( mg/kg/h \) ketamine were adequate in managing postoperative pain.

4. CONCLUSION

A multidisciplinary approach and teamwork, accompanied by high vigilance at each perioperative stage, are the keys to success. Management in conjoined twins needs to be done by treating them as two different individuals, even though there are attached parts of organs. In the case of thoracoabdominopagus conjoined twins, an anesthesiologist should consider the possibility of shared circulation. This becomes important to the use of induction and maintenance drugs via the intravenous route. The anesthesia team, surgeons, pediatricians and parents should know the complications and anticipation of both anesthesia and surgery and the management during the postoperative period.

5. Conflict of interest

The authors declare no conflict of interests.

6. Informed Consent

Informed consent of the parents of the twins was obtained to document this case report and use the pictures of the children.

7. Authors’ contribution

P: Peri-operative anesthesia management, conceptualization. SBS and BN: Peri-operative anesthesia management, literature search, reviewing and editing. HG: Pre-peri-post operative anesthesia management, writing initial and final manuscript. All authors read and approved the final manuscript.

8. REFERENCES


