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General anesthesia for laparotomy in a patient with uncorrected tetralogy of Fallot with pulmonary atresia (pseudotruncus arteriosus)

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Introduction

Patients with tetralogy of Fallot (TOF) accompanied by pulmonary atresia (pseudotruncus arteriosus) rarely survive beyond the third decade. Therefore, general anesthesia for noncardiac surgery on such a patient is extremely rare and challenging. We report a case of TOF with pulmonary atresia that was successfully managed under general anesthesia.

Case report

The patient was a 39-year-old woman, (height 166 cm, weight 43 kg). Total hysterectomy and bilateral salpingo-oophorectomy were planned for uterine cancer. She had cyanotic heart disease which was not clearly diagnosed during childhood and had been restricted from participating in hard sports. On cardiac catheterization at the age of 20, she was diagnosed as TOF with pulmonary atresia. No palliative shunt surgery had been performed.

Physical examination on admission showed prominent digital clubbing with marked cyanosis. She felt shortness of breath on climbing stairs. Chest roentgenogram demonstrated a typical "boot-shaped" heart with mildly diminished pulmonary flow. The hemoglobin concentration was 16.2 g·dl^{-1} and the hematocrit was 53%. Arterial blood gas analysis on room air showed a pH of 7.39, Pco_2 41 mmHg, Po_2 40 mmHg, and oxygen saturation 74%. ECG showed a heart rate of 76 beats·min^{-1}, right axis deviation, and incomplete right bundle branch block. Cardiac catheterization before surgery showed a right ventricular pressure of 100/0 mmHg, left ventricular pressure of 100/0 mmHg, and pressure in the aorta of 100/60 mmHg. The pulmonary blood flow was derived from collateral vessels arising from the descending aorta which were considered bronchial arteries. The pressure in the orifice of the collateral vessel was 100/60 mmHg, but the cardiac catheter was unable to be inserted further. The pulmonary arterial pressure was expected to be low.

The patient was premedicated with intramuscular atropine (0.5 mg) and midazolam (2.0 mg). A left radial arterial catheter was inserted before induction of anesthesia and an epidural catheter was also inserted through the L1-L2 intervertebral space for postoperative analgesia. Pulse oximetry, arterial blood pressure, and end-tidal carbon dioxide concentration were monitored throughout the anesthetic course. After 5 min of preoxygenation, arterial blood gas analysis revealed a pH of 7.43, $Paco_2$ of 34 mmHg, Pao_2 of 63 mmHg, and Sao_2 of 93%. At the same time, the pulse oximeter indicated 88% of Sao_2, probably because of a rightward shift of the oxyhemoglobin dissociation curve caused by the cyanotic heart disease. As this value was consistent with the value measured by a CO oximeter (IL 482, RIKO, Tokyo, Japan), the pulse oximeter was carefully monitored throughout the surgery to treat dangerous hypoxemia promptly. Following the induction of general anesthesia with intravenous fentanyl 0.2 mg, ketamine 40 mg, and midazolam 2 mg, the patient was intubated with a tracheal tube following 10 mg of vecuronium. Anesthesia was maintained with air, oxygen, and sevoflurane (0.3%–0.5%) with a continuous infusion of ketamine (0.5–1 mg·kg^{-1}·h^{-1}). Vecuronium was given for neuromuscular blockade. Inspiratory oxygen concentration was controlled between 70% and 100% to keep Sao_2 above 80% monitored by the pulse

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oximeter. Arterial CO_2 was maintained in normocapnia. The first episode of mild hypotension (from 140/72 to 116/68) leading to hypoxemia (Sao_2, from 84% to 76%) occurred at 55 min after the beginning of surgery. Intravenous injection of ephedrine 5 mg raised blood pressure from 116/68 mmHg to 152/80 mmHg, but Sao_2 did not increase. Then, 1 mg of methoxamine was injected, which increased Sao_2 from 76% to 84% with 10 mmHg increase of systolic blood pressure. The second episode of hypoxemia was also treated with 1 mg of methoxamine and volume replacement, which improved Sao_2 from 73% to 81% (blood pressure from 138/72 to 152/72 mmHg).

Residual neuromuscular block was antagonized with atropine 1.0 mg and neostigmine 2.0 mg iv. Estimated blood loss during approximately 4 h of surgery was 650 g, and 3.25 l of crystalloid solution as well as 200 ml of 5% albumin were administered. Before extubation, 0.1 mg of buprenorphine with normal saline was administered through the epidural catheter for postoperative analgesia. After uneventful extubation the patient was transferred to the intensive care unit. After an uneventful 24-h period there, she was returned to the general ward. Following a course of chemotherapy the patient was discharged from the hospital.

Discussion

The term "pseudotruncus arteriosus" is applied to TOF in which obstruction to pulmonary flow is complete (pulmonary atresia). In such case, pulmonary blood flow is derived through arteries arising from the aorta such as bronchial arteries, and a decrease in systemic vascular resistance (SVR) or increase in pulmonary vascular resistance (PVR) tends to decrease pulmonary blood flow with worsening of hypoxemia. Therefore, anesthetic management should maintain SVR, minimize PVR, and avoid myocardial depression. Reducing total body oxygen consumption by adequate anesthesia depth can increase venous oxygen saturation and may be useful for raising arterial oxygen saturation [1].

As epidural administration of local anesthetic causes a decrease in SVR [2], a small dose of opioid (buprenorphine 0.1 mg) with 6 ml of normal saline was administered through an epidural catheter for postoperative analgesia. Inhalational anesthetic can also decrease SVR in high concentrations; therefore, only low concentrations of sevoflurane (0.3%–0.5%) were used to ensure amnesia for the procedure and to reduce total body oxygen consumption. This did not produce untoward hemodynamic changes. As sevoflurane has a relatively low blood/gas partition coefficient, it was useful to control quickly the depth of anesthesia and hence the circulation in this patient.

Although pulmonary artery pressure may be increased by ketamine in adults, 40 mg of ketamine was used slowly for induction and 0.5–2.0 $mg \cdot kg^{-1} \cdot h^{-1}$ for maintenance of anesthesia as it maintains SVR and provides analgesia. Fentanyl 0.1 mg was used to attenuate the pressor response to tracheal intubation and 0.1 mg was added just after skin incision. Fentanyl was used in low doses (total 0.2 mg) as a large dose may prolong emergence from anesthesia and may cause respiratory depression after tracheal extubation which worsens hypoxemia. The combination of fentanyl and midazolam occasionally causes hypotension in cardiac patients, but it was not prominent in our patient. This may have been due to the pressor action of the ketamine administered concurrently.

We did not use nitrous oxide in this case because it may increase PVR. The patient's lungs were ventilated with minimal airway pressure to maintain normocapnia, because high alveolar pressure or hypercapnia increases PVR and decreases pulmonary blood flow with worsening of hypoxemia.

Deterioration of arterial oxygen saturation occurred twice during anesthesia, but was successfully treated by intravenous administration of 1 mg of methoxamine. As the adrenergic innervation of the pulmonary vascular tree is more modest than that of the systemic arterial tree [3], α-adrenergic stimulation may constrict the systemic vasculature more than the pulmonary vasculature and effectively shift the systemic blood flow to the pulmonary vessels in patients with TOF. As the bronchial artery constricts with α-adrenergic stimulation [4], methoxamine may increase bronchial arterial tone, which can reduce pulmonary blood flow, resulting in worsening of Sao_2. However, the effect of methoxamine on systemic peripheral vasculature might be more prominent than the effect on the bronchial artery and the pulmonary artery [5]. Although the causes of the desaturation were not clear, possible causes may have been a reduction in cardiac output due to blood volume loss or decreased SVR due to deep anesthesia.

In conclusion, we present a case of general anesthesia for a patient with TOF with pulmonary atresia which was successfully managed by a balanced anesthetic technique (combination of fentanyl, ketamine, midazolam, and sevoflurane). Maintaining oxygenation is crucial in cyanotic patients. This can be accomplished by maintaining SVR and PVR properly with adequate anesthesia, volume replacement, and pharmacological agents according to the physiological abnormalities.

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