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Anesthetic management of pediatric patients following Fontan operation

Akiko Yamashita, Yukio Hayashi, Noriko Horinokuchi, Yoshihiko Ohnishi, and Masakazu Kuro

Department of Anesthesiology, National Cardiovascular Center, 5-7-1 Fujishirodai, Suita, Osaka 565, Japan

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Introduction

The recent success of Fontan surgery now provides more opportunities to anesthetize pediatric patients who have previously undergone Fontan procedure. Since the most important feature of the Fontan procedure is the surgical connection of the right atrium to the pulmonary artery, systemic venous return is directed to the pulmonary artery without a functional ventricle and the pulmonary circulation depends on the pressure gradient between central venous and left atrial pressures and pulmonary vascular resistance [1,2]. Positive pressure ventilation reduces venous return and hence decreases pulmonary blood flow. Thus, local anesthesia may be more favorable to maintain spontaneous ventilation [3,4]. However, pediatric patients cannot tolerate major surgery with regional anesthesia. We report herein the anesthetic management of two pediatric patients who previously had Fontan-type surgery. Although both patients had undergone Fontan operation, one had a complete Fontan procedure with separation of the pulmonary and systemic circulation, while the other had a considerable right-to-left shunt at the left atrium.

Case reports

Case 1

The patient was a girl aged 2 years and 9 months weighing 10.7 kg with mitral atresia, double outlet of right ventricle, and pulmonary stenosis. She had undergone Fontan surgery at 2 years and 4 months of age. Her recovery was uneventful. However, she suffered from an unknown fever and diarrhea 4 months after discharge. On admission, her body temperature was high (38.0°C), and both her leukocyte count and C-reactive protein (CRP) were elevated ($24\,600 \cdot \mu l^{-1}$ and $19.2\,mg \cdot dl^{-1}$, respectively). The diagnosis of left retroperitoneal abscess was strongly suggested by abdominal ultrasound imaging. Thus, an emergency abdominal drainage was scheduled. Since the patient already had an intravenous line, we omitted premedication, and anesthesia was induced by intravenous thiopental 25 mg, fentanyl 20 μg, midazolam 2 mg, and pancuronium 2 mg. Routine noninvasive monitoring including an ECG (lead II), a pulse oximeter (Nellcor N 2000, Hayward, CA, USA), and an automated blood pressure monitor (Dinamap, Critikon, Tampa, FL, USA) was applied before anesthesia. Anesthesia was maintained with fentanyl, isoflurane, and nitrous oxide. Inspired oxygen concentration (FiO_2) was set at 40%, and peripheral oxygen saturation (Spo_2) after the tracheal intubation was 98%. The patient was ventilated to maintain an end-tidal CO_2 between 29 and 34 mmHg. The arterial blood pressure and the central venous pressure (CVP) after induction of anesthesia were 95/63 and 13 mmHg, respectively. Heart rate was 140 bpm. Arterial blood gas analysis revealed partial pressure of arterial oxygen (Pao_2) 109 mmHg, partial pressure of arterial carbon dioxide ($Paco_2$) 28 mmHg, and pH 7.51 before the operation. During exploration of the abdomen, unexpected massive bleeding which amounted to about 200 ml in 10 min occurred, arterial blood pressure decreased to 80/48 mmHg, heart rate increased to 180 bpm, and CVP decreased to 11 mmHg. Intravenous methoxamine was administered to maintain systemic arterial pressure, while the blood loss was managed by fluid infusion followed by transfusion, resulting in restoration of blood pressures and heart rate. During this

Address correspondence to: Y. Hayashi
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accident, Spo_2 and end-tidal CO_2 concentration remained unchanged (at 99% and 31 mmHg, respectively). CVP was maintained at 13 or 14 mmHg and the hemodynamic variables were stable during the remainder of the operation. Neuromuscular blockade was reversed and the endotracheal tube was removed. Her hemodynamic variables following extubation were stable, and Pao_2, $Paco_2$, and pH were 113 mmHg, 37 mmHg, and 7.47 while breathing 4 l·min^{-1} oxygen via a face mask, respectively.

Case 2

The patient was an 11.2-kg 88-cm 3-year-old girl with hypoplastic right ventricle, pulmonary atresia, and atrial septal defect, who had undergone Fontan operation at the age of 2 years. Although the recovery was uneventful, postoperative echocardiography showed atrial baffle leakage with a slight right-to-left shunt, and her Spo_2 in room air was about 80%. The right-to-left shunt increased remarkably (45%) 1 year after the operation, and arterial oxygen saturation (Sao_2) in room air was 66.9%. The ratio of pulmonary and systemic blood flow was 0.63. She was scheduled for repair of the atrial leakage under cardiopulmonary bypass. Premedication consisted of oral diazepam (7 mg) and intramuscular scopolamine (0.1 mg), as well as meperidine (10 mg) 2 h and 1 h prior to anesthesia, respectively. After application of routine noninvasive monitors, anesthesia was induced with halothane in 50% nitrous oxide and oxygen. Pancuronium was given to facilitate tracheal intubation. Anesthesia was maintained with moderate-dose fentanyl (440 μg) and diazepam (5 mg) in oxygen. The arterial blood pressure, CVP, heart rate, and Spo_2 after induction of anesthesia were 90/51, 12 mmHg, 112 bpm, and 88%, respectively. Although the patient was mechanically ventilated to maintain end-tidal CO_2 at 31 mmHg, arterial blood gas analysis revealed Pao_2 67 mmHg, $Paco_2$ 42 mmHg, and pH 7.32. After sternotomy, although arterial blood pressures and CVP decreased (80/51 mmHg and 8 mmHg) and heart rate increased (122 bpm), Spo_2 and end-tidal CO_2 concentration remained stable. The patient was treated with albumin to increase CVP to 12 mmHg, which was maintained until the cardiopulmonary bypass. The remainder of the surgery including weaning from the cardiopulmonary bypass was uneventful. She recovered from anesthesia without complications and the trachea was extubated 9 h after the operation.

Discussion

It is indispensable to understand the unique physiology of patients with a Fontan-type circulation for anesthetic management of this population. In the absence of a functional pulmonary ventricle, pulmonary blood flow depends on a pressure gradient between the right and left atrium and pulmonary vascular resistance. Since positive pressure ventilation may increase intrathoracic pressure, resulting in reduced pulmonary blood flow, spontaneous ventilation under local anesthesia is recommended [2–5]. However, even if the operation can be performed under regional anesthesia, we should watch the patient carefully for hypercarbia and acidosis associated with heavy sedation. In the present cases, the operations could not be performed under regional anesthesia and general anesthesia was required. Since there is still no reliable monitoring available for the direct evaluation of pulmonary blood flow in these patients, we cannot state that changes of pulmonary blood flow occurred when spontaneous ventilation was controlled following induction of anesthesia.

It is important to prevent hypovolemia for the maintenance of pulmonary blood flow. It is well established that monitoring of CVP is quite useful for assessment of volume status to prevent hypovolemia during anesthetic management of these patients. A sudden increase in pressure may indicate impaired systemic ventricular function and/or elevation of pulmonary vascular resistance. In addition, excessive elevation of CVP may impair venous return from the cerebral circulation. In the current cases, fluid therapy was performed to maintain the initial CVP, and no episode of increased central venous pressure was noted.

Pulmonary ventilation must be managed to prevent pulmonary vascular constriction. Factors increasing pulmonary vascular resistance include hypoxia, hypercapnia, acidemia, and hypothermia [6], and intensive hyperventilation combined with high concentrations of inspired oxygen has been recommended [2]. In the second case, with a discrepancy between end-tidal CO_2 and $Paco_2$ presumably due to the residual shunt, a more aggressive hyperventilation would have been necessary to decrease $Paco_2$. However, aggressive hyperventilation might have increased mean and peak airway pressure, causing an undesirable reduction of pulmonary blood flow. Thus, we did not dare facilitate hypocapnia by the ventilatory challenge, because hemodynamic variables were stable and Spo_2 was tolerable.

Although the patients presented here had previously undergone Fontan operation, there were important differences in their circulation: The first case preserved physiological separation of the systemic and pulmonary circulations, while the second case had a significant residual atrial right-to-left shunt (Fig. 1). The residual shunt may be helpful to preserve ventricular preload and cardiac output and to decrease central venous pressure, if it is limited [7–9]. However, the shunt in the second case was enhanced and arterial desaturation was

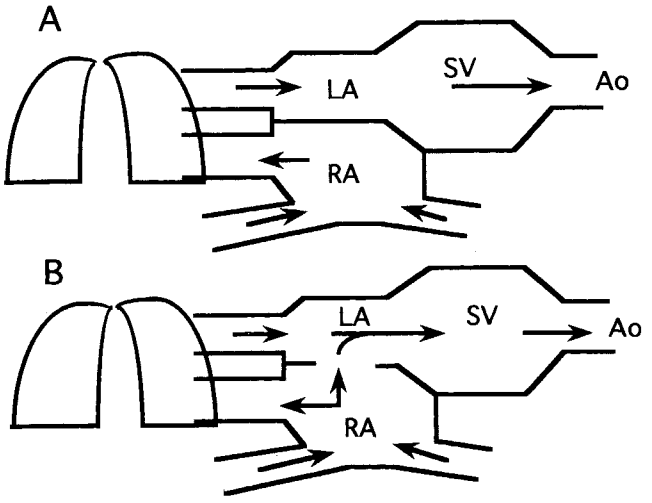

Fig. 1. Schematic illustration of the different patterns of blood flow (**A** case 1, **B** case 2). *Arrows* indicate the direction of blood flow. *SV*, systemic ventricle; *Ao*, aorta; *LA*, left atrium; *RA*, right atrium

a critical concern. Therefore, we ventilated the second patient with 100% oxygen until surgical repair. In addition, the monitoring of Spo_2 and end-tidal CO_2 may have been valuable in the second case, because these variables were affected by changes of the residual shunt.

In the first case, we administered methoxamine to manage hypotension and tachycardia induced by massive bleeding, although volume replacement including transfusion should be the first option for bleeding. In addition, the use of this kind of drug for patients with a Fontan circulation is controversial [3–5]. However, we were concerned that our patient's heart might not tolerate the short period of hypotension until the initiation of transfusion because coronary perfusion may be impaired following hypotension.

To manage the current cases, real-time monitoring of the changes in their pulmonary blood flow was required, especially in the second case. Although a pulmonary artery catheter is useful for measuring pulmonary flow in general, no data have been published about the accuracy of measurements in such patients. Hosking and Beynen [10] proposed the efficacy of superior vena cava oxygen saturation as a continuous monitor in a similar patient undergoing repair of coarctation of the aorta. Indeed, it may be helpful, because this value indirectly reflects cardiac output and is correlated with central venous pressure. However, since several factors including arterial saturation, hemoglobin, and oxygen consumption, affect this value [11], further data are be required to evaluate its sensitivity and efficacy.

We used nitrous oxide for maintenance of anesthesia in the first patient. One may claim that nitrous oxide should not have been chosen, because it facilitates subsequent increases in pulmonary vascular resistance [12]. However, this property appears to a significant extent only in the adult population with pulmonary hypertension. In infants, nitrous oxide does not produce such an elevation [13]. Since early recovery from anesthesia is favorable for patients with a Fontan circulation, the benefits of nitrous oxide must be weighed in spite of the subsequent risk of increasing pulmonary vascular resistance.

In conclusion, we report the successful anesthetic management of two pediatric patients who previously had a Fontan-type operation. General anesthesia may safely be applied to these children, and monitoring of central venous pressure is important for assessment of volume status to maintain the pulmonary blood flow.

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