

Successful one-lung ventilation in a patient with the Fontan circulation undergoing thoracotomy: a case report

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Abstract The incidence of noncardiac surgery in patients with the Fontan circulation has increased over the years due to the elongated life expectancy of these patients. In patients with the Fontan circulation, pulmonary blood flow is passive, so it is important to keep pulmonary vascular resistance low. One-lung ventilation (OLV) can have adverse effects on the Fontan circulation due to hypoxia, hypoxic pulmonary vasoconstriction, hypercarbia, and increased airway pressure. We present a case of successful OLV in a patient with the Fontan circulation and describe our perioperative management.

Keywords Total cavopulmonary connection operation · One-lung ventilation · Pulmonary vascular resistance

Introduction

The Fontan operation was first performed for the functional repair of tricuspid atresia in 1971. Total cavopulmonary connection (TCPC)—a modified Fontan operation—was established in 1988, and continues to be widely used for univentricular heart malformations. Recently, more patients with the Fontan circulation have reached adulthood and now require noncardiac surgery [1]. There are reports of cesarean sections [2], scoliosis surgery [3], laparoscopic surgery [4], pheochromocytoma [5], and craniotomies [6].

Since one-lung ventilation (OLV) can cause an increase in pulmonary vascular resistance (PVR), there is a risk that the Fontan circulation will fail. It is important to understand Fontan physiology and its interrelationship with OLV.

Case report

The patient was a 12-year-old female who was 152 cm tall and weighed 47 kg. She was diagnosed with a single atrium, single ventricle, pulmonary stenosis, and a persistent left superior vena cava at birth. She had no visceral inversion. She had previously undergone a bilateral bidirectional cavopulmonary shunt procedure when she was 1 year old and a lateral tunnel total cavopulmonary connection operation at 4 years of age. Her cyanosis worsened because there were large varices from the right superior vena cava to the right pulmonary vein (Fig. 1). Surgery was planned to treat the varices, as there was risk of rupture and thrombus. Her baseline oxygen saturation was 85%. She could tolerate short-term but not long-term exercise. Cardiac catheterization revealed that her Qp/Qs was 0.86 and her central venous pressure (CVP) was 11 mmHg. Her pulmonary arterial pressure was 11 mmHg, and her pulmonary artery index was 214. A cardiac echo revealed that the ejection fraction was 60% and the regurgitation of the common atrioventricular valve was trivial. A blood test showed that her hemoglobin was 13.9 g/dl.

The patient refused a preoperative intravascular line and took 200 ml of water 2 h before the operation to avoid dehydration. Anesthesia was induced with sevoflurane and vecuronium. She was intubated with an ID 6.5 mm cuffed tube. Anesthesia was maintained with air, oxygen, sevoflurane, fentanyl (total dose 750 μg ; 2 $\mu\text{g kg}^{-1} \text{h}^{-1}$), and vecuronium. In addition to basic monitoring, invasive

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arterial pressure, CVP of the right superior vena cava, and regional cerebral oxygen saturation (rSO₂) were monitored during the operation. Since changes in the pulmonary blood flow during OLV may decrease cardiac output, cardiopulmonary bypass was prepared for at the beginning of the operation.

The patient was placed in the left lateral position and OLV was initiated with a 7 Fr Arndt bronchial blocker. The inspiratory oxygen fraction was 1.0, the tidal volume was 320 ml, the respiratory frequency was 15 min⁻¹, and the peak

inspiratory pressure/end expiratory pressure was 34/4 cm H₂O during OLV. The end-tidal carbon dioxide was maintained at less than 40 mmHg with an oxygen saturation of greater than 83%. The CVP increased from 14 to 30 mmHg, so 0.5 μg kg⁻¹ min⁻¹ nitroglycerin was administered to decrease the PVR. Her blood pressure decreased from 98/50 to 65/39 mmHg. Therefore, 5 μg kg⁻¹ min⁻¹ dopamine and 3 μg kg⁻¹ min⁻¹ dobutamine were administered, and fluid management was regulated to maintain sufficient intravascular volume for adequate pulmonary blood flow and cardiac output. Her rSO₂ was 80% before the operation, and it was maintained at >56% during OLV. OLV lasted 3 h and 50 min (Fig. 2). There was no cardiac dysrhythmia during the operation. The total blood loss was approximately 98, and 2040 ml of Ringer's lactate was infused during the operation. The total urine output during surgery was 280 ml. The patient was extubated in the operating room, and postoperative analgesia with 1.0 μg kg⁻¹ h⁻¹ fentanyl was continued. Following the operation, oxygen saturation improved to 95% on room air.

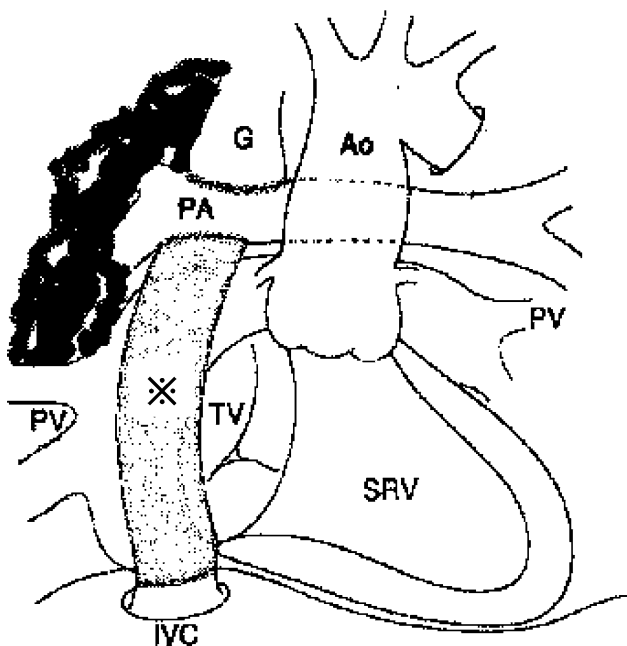
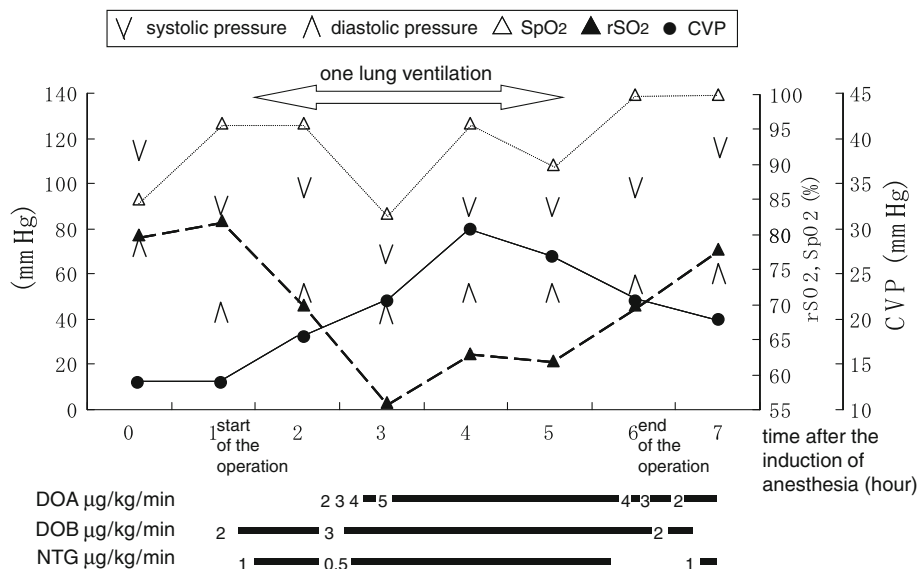


Fig. 1 Giant varices from the right superior vena cava to the right pulmonary vein (diameter: about 10–20 cm). SRV single right ventricle, G Glenn shunt, * indicates an artificial blood vessel

Discussion

It is important to maintain adequate intravascular volume and low PVR during OLV in patients with the Fontan circulation. Hypoxia, hypoxic pulmonary vasoconstriction (HPV), hypercarbia, and increases in airway pressure may result in an increase of PVR during OLV. Therefore, we were careful to maintain adequate lung volumes and gas exchanges, and used nitroglycerin to decrease PVR, although inhaled nitric oxide (NO) and intravenous milrinone are other options for reducing PVR. Yoshimura et al. [7] reported that CVP > 15 mmHg after Fontan-type

Fig. 2 Intraoperative course



operations are appropriate indications for inhaled NO therapy. We did not administer NO during the operation because we planned early extubation suitable for the Fontan circulation, and repeated the deterioration of PVR after its withdrawal remained a concern. Cai et al. [8] reported that the combined use of NO and milrinone optimally stabilized the pulmonary hemodynamics after a Fontan operation.

Generally, the ratio of the pulmonary flow from the nondependent side to the dependent side is 0.2 during OLV with the patient in the lateral position because of the influence of gravity and HPV [9]. Further study is needed to clarify how HPV affects the Fontan circulation. Although a central venous catheter carries a risk of thrombosis, the information it provides may be extremely valuable, since CVP increases and cerebral ischemia may occur during OLV. To assess the adequacy of cerebral perfusion, rSO₂ should be monitored with an INVOS[®] (Edwards Lifesciences) device and maintained at more than 75% of the baseline value to reduce ischemia-induced brain damage [10].

We considered three perioperative complications associated with the Fontan circulation. First, tachycardia can compromise the Fontan hemodynamics. Therefore, we controlled body temperature and avoided increasing the sympathetic tone due to pain by providing postoperative analgesia. Second, anticoagulation therapy is recommended to prevent thromboembolic events. In this case, epidural anesthesia was avoided because the patient had received anticoagulation therapy. Finally, antibiotic prophylaxis is necessary to prevent infective endocarditis.

Long-term complications associated with the Fontan circulation are arrhythmias, heart failure, sudden death, thromboembolism, and protein-losing enteropathy. Symptomatic bradycardia is one of the most serious complications. Although the implantation of a pacemaker is necessary in such a situation, it is difficult for the leads to reach the heart transvenously [10]. Therefore, a thoracotomy with a high possibility of OLV is required. It is

necessary to fully evaluate the patient's hemodynamics and physical condition and to carefully manage the factors during the surgery.

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