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Anesthetic management for cesarean section in a mother after the Fontan procedure

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Introduction

The Fontan procedure, a method of right atrial-pulmonary arterial connection, was first described in 1971 [1]. Since then a number of studies have been published on the use of this procedure, which have generally presented favorable long-term outcomes [2–5]. However, there is little information on the effects of the Fontan procedure on pregnancy and delivery. Obstetric and anesthetic literature [6,7] have described a few cases in which spinal or epidural block was successfully used to provide anesthesia for cesarean delivery or analgesia for vaginal birth in patients with Fontan repair. To our knowledge, there has been no report on general anesthetic management for cesarean delivery in a mother with Fontan circulation. Accordingly, we report a successful cesarean delivery in a patient who had undergone the Fontan procedure 13 years before her pregnancy.

Case report

A 30-year-old woman, gravida 1, was referred to our hospital at 12 weeks of gestation. She had been born with complete transposition of the great arteries and a crisscross atrioventricular valve, ventricular septal defect (VSD), and pulmonary stenosis. At the age of 5, she had undergone a palliative Blalock-Hanlon procedure. At the age of 14, she experienced recurrent episodes of symptomatic supraventricular tachycardia. At 17 years of age, a Fontan procedure was carried out, in which the right atrium was connected to the main pulmonary artery with closures of the VSD and the Blalock-Hanlon shunt. She recovered uneventfully from the operation. Since the Fontan repair, and throughout her pregnancy, she has had a normal activity level (New York Heart Association Class I) and has taken no medication. At 37 weeks of gestation she was admitted to our hospital for delivery. Cesarean delivery was planned at 38 weeks of gestation.

On preoperative maternal physical examination, no cyanosis, clubbing, or edema was noted. Her hematocrit was 36%, hemoglobin oxygen saturation was 93% while breathing room air, and an electrocardiogram revealed sinus rhythm with a rate of 80 bpm, occasional supraventricular premature beats, right axis deviation, and first-degree atrioventricular block. Maternal echocardiography showed that the atrioventricular relationship was complex, with concordant crisscross heart, and the left ventricular size was relatively small. Therefore, echocardiographic measurements of left and right ventricular dimensions, left ventricular wall thickness, and left atrial size were not possible. However, two-dimensional echocardiographic estimation of her ventricular function suggested that, throughout her pregnancy, her ventricular contraction was reasonably good for a post-Fontan patient. Mild regurgitation of the mitral valve was noted, but the degree did not change during pregnancy. The pattern of blood flow in the pulmonary artery also did not change significantly during pregnancy, and the peak velocity in the pulmonary artery, which was observed during ventricular diastole, was 0.6–0.8 $m \cdot s^{-1}$ throughout her pregnancy.

On the patient's arrival in the operating room, monitoring by electrocardiogram, as well as automated arterial blood pressure and pulse oximetry monitoring, was begun. The arterial blood pressure was 120/

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75 mmHg, the heart rate was 120 bpm with sinus rhythm, and the oxygen saturation (SpO_2) on room air was 88%. Oxygen supplementation of 6 l·min^{-1} was started using a mask. In addition to the routine monitors, catheters were placed in the left radial artery for systemic blood pressure monitoring and in the right external jugular vein for central venous pressure (CVP) monitoring. General anesthesia was induced with thiopental (250 mg) and fentanyl (100 μg). After the induction, an increase in heart rate from 120 to 175 bpm was noted. Thereafter, fentanyl (100 μg) was administered intravenously. Succinylcholine chloride (80 mg) was administered to facilitate the endotracheal intubation.

The patient was ventilated with 100% oxygen using a volume-preset ventilator with a tidal volume of 450–550 ml and a rate set at 10–15 min^{-1} in order to maintain an end-tidal CO_2 level of 25–35 mmHg. Peak airway pressure was kept below 15 cmH_2O. Arterial blood gas tensions after induction of general anesthesia showed a pH of 7.38, $PaCO_2$ of 33.6 mmHg, and PaO_2 of 126 mmHg. After the fetus was delivered and the umbilical cord was clamped, 10 mg midazolam and 6 mg vecuronium were administered.

Five minutes after delivery, the maternal blood pressure decreased to 90/60 mmHg, and phenylephrine was administered intravenously. The maternal blood pressure then increased to 130/70 mmHg. The heart rate varied from 100 to 120 bpm, and SpO_2 always exceeded 96%. End-tidal CO_2 varied from 25 to 30 mmHg. CVP after induction of anesthesia was 14 mmHg. Measured blood loss was 1450 ml, including an approximately normal volume of amniotic fluid. Fluid given during the period of anesthesia consisted of 900 ml of Ringer's lactate solution. The baby weighed 2355 g and had Apgar scores of 8 at 1 min and 9 at 5 min. After delivery, uterine contraction was augmented with oxytocin 0.1 U·min^{-1} i.v. At the conclusion of surgery, the neuromuscular blockade was reversed and the trachea was extubated.

The patient was transferred to an intensive care unit (ICU), where her heart rate increased to 140 bpm and her blood pressure decreased to 70/40 mmHg. At this time, her CVP was 12 mmHg. The patient was treated with volume replacement with albumin 100–200 ml·h^{-1} and 2 units of concentrated red blood cells and was given digoxin 0.25 mg, resulting in restoration of blood pressure and heart rate. One day after the operation, the patient became hemodynamically stable.

Discussion

We have previously reported a case of epidural anesthesia for cesarean delivery after a maternal Glenn operation [8]. The post-Glenn circulation is similar to the Fontan circulation. In the present case, however, the pregnant patient was extremely nervous, and after repeated discussion among anesthesiologists, obstetricians, and cardiologists, general anesthesia was planned for cesarean delivery with the patient's consent.

In the present case, the Fontan repair procedure consisted of connecting the right atrium directly to the main pulmonary artery, so that the pulmonary blood flow was dependent on the pressure difference between the right and left atria. It has been reported that a right/left atrial pressure difference of 7–10 mmHg is important to maintain pulmonary blood flow and to overcome pulmonary vascular resistance (PVR) [9]. Therefore, an understanding of the unique physiology of the post-Fontan circulation is of primary importance for safe anesthetic management. Prevention of hypovolemia and maintenance of low PVR are the keys to hemodynamic stability in the patient after Fontan repair. General anesthesia with positive pressure ventilation may also be detrimental because of the possibility of positive intrathoracic pressure causing a reduction in venous return to the pulmonary circulation. During general anesthesia, skillful airway management is extremely important in order to avoid an increase in PVR. Therefore, care must be taken to avoid high peak airway pressure. Another precaution is to adjust the ventilator settings to avoid prolonged increases in duration of inspiration or increases in mean airway pressure. In our patient, the peak airway pressure was held to less than 15 cmH_2O, and the ratio of inspiratory to expiratory phases (I:E) was 1:3–4.

Pulmonary ventilation must be managed to prevent pulmonary vascular constriction. An increase in PVR, especially if caused by hypoxia, may result in decreasing venous return to the ventricle and thus a decrease in cardiac output [10,11]. Therefore, the inspired oxygen concentration was set at 100%, so that SpO_2 could be maintained at greater than 96% during the surgery. Other factors that may increase PVR were avoided, including fighting with the ventilator, hypothermia, hypercarbia, and acidosis [10]. Therefore, the tidal volume and rate were set to avoid hypercarbia, and end-tidal CO_2 remained between 25 and 30 mmHg. These respiratory management steps are unnecessary under epidural anesthesia, in which endotracheal intubation or positive pressure ventilation is not required. Generally, pulmonary blood flow is better maintained during spontaneous ventilation than during positive pressure ventilation. Early extubation and spontaneous ventilation are desirable to maintain better pulmonary blood flow. We used a small dose of narcotics in order to achieve early recovery of normal spontaneous breathing.

We administered phenylephrine to treat hypotension and tachycardia induced by bleeding, although volume

replacement, including transfusion, should be the first option for hypovolemia. The α-agonists increase PVR [12]. When a relatively long time is required for fluid therapy, phenylephrine must be used to restore blood pressure, despite the resulting pulmonary vasoconstriction and decrease in cardiac output.

In practical terms, it is difficult to use a pulmonary artery catheter because of the atypical anatomy after the Fontan procedure. We chose CVP to replace fluid in our patient. In the Fontan patient a decrease in CVP directly reflects a reduction in pulmonary blood flow and a systemic ventricular preload. During the surgery, CVP decreased to 12 mmHg as a result of unexpected massive bleeding. The blood loss was compensated by fluid infusion. However, in the ICU, the heart rate increased and blood pressure decreased. The patient was treated by volume loading 100–200 $ml \cdot h^{-1}$ with transfusion to increase CVP. Prior to the operation, we should have managed fluid replacement in our patient with measurement of CVP.

Epidural anesthesia might result in better intraoperative conditions and postoperative analgesia for cesarean delivery after the Fontan procedure. However, when the patient refuses regional anesthesia, general anesthesia can be successfully performed with careful respiratory management.

In conclusion, it is important to prevent hypovolemia and tachycardia for the maintenance of pulmonary blood flow and cardiac output in the patient who has received Fontan repair and undergoes cesarean delivery.

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