Cesarean delivery in a patient with a congenitally corrected transposition of the great arteries, large ventricular septal defect, and severe pulmonary hypertension: a case-report

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Introduction

Congenitally corrected transposition of the great arteries (CCTGA), in which the ventricles are reversed from their normal anatomic position, is a rare heart defect occurring in 0.5% of the population. Pregnancies in women with CCTGA without associated anomalies, such as VSD, have been reported. [1] (1) Herein we report the management of a cesarean delivery (CD) in a 35-year-old nullipara with severe pulmonary hypertension (pHTN) due to unrepaired large VSD associated with CCTGA.

Case Report

- A 35-year-old G2P0 parturient with the diagnosis of CCTGA was referred to our hospital for peripartum care at 12w.
- Intracardiac repair during her childhood was not indicated due to pulmonary hypertension. She received furosemide 10 mg before delivery but never experienced heart failure or arrhythmia.
- Chest X-ray showed cardiomegaly and dilated pulmonary arteries (Fig 1). A preconception MRI showed good contractility of both ventricles, but the shunt fraction (Qp/Qs) was 3.8. On echocardiography, the max. right-sided mitral valve pressure gradient was 73 mmHg (Fig 2).
- She was admitted at 16 weeks' gestation for new onset proteinuria; preeclampsia was ruled out, but she was kept in hospital with bed rest, furosemide 20 mg, and heparin SQ.

Before surgery

- Right heart catheterization at 30 weeks confirmed severe pulmonary hypertension (PAP 121/43(76) mmHg), Ao 129/50(82) mmHg, CI 2.79 L/min/m2, Qp/Qs 2.24, SVRI 17.6 WU, and PVRI 10.9 WU.
- Abdominal echocardiography revealed intervillous blood pooling which might have caused fetal growth restriction (Fig 3A). Placental malformation resulted in severe intrauterine FGR, and CD was planned for week 33.
- A multidisciplinary team consisting of obstetricians, anesthesiologists, cardiologists, and neonatologists discussed the plan for her delivery.

During surgery

- Invasive monitoring, including an arterial and central venous line, was performed; a PA catheter was not placed due to concerns about accidental insertion to the left ventricle through the ventricular septal defect.

Fig 1

Fig 2

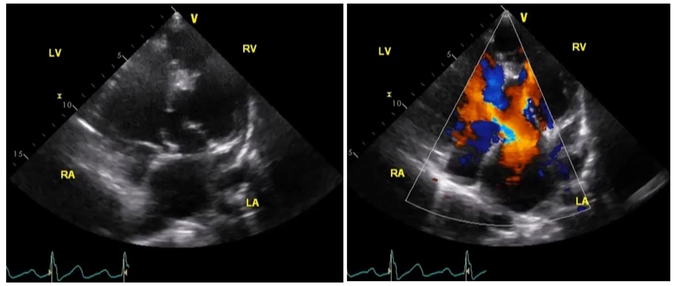


Fig 3

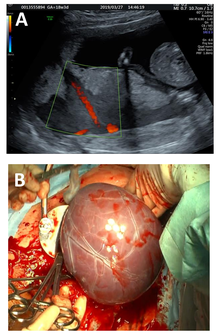
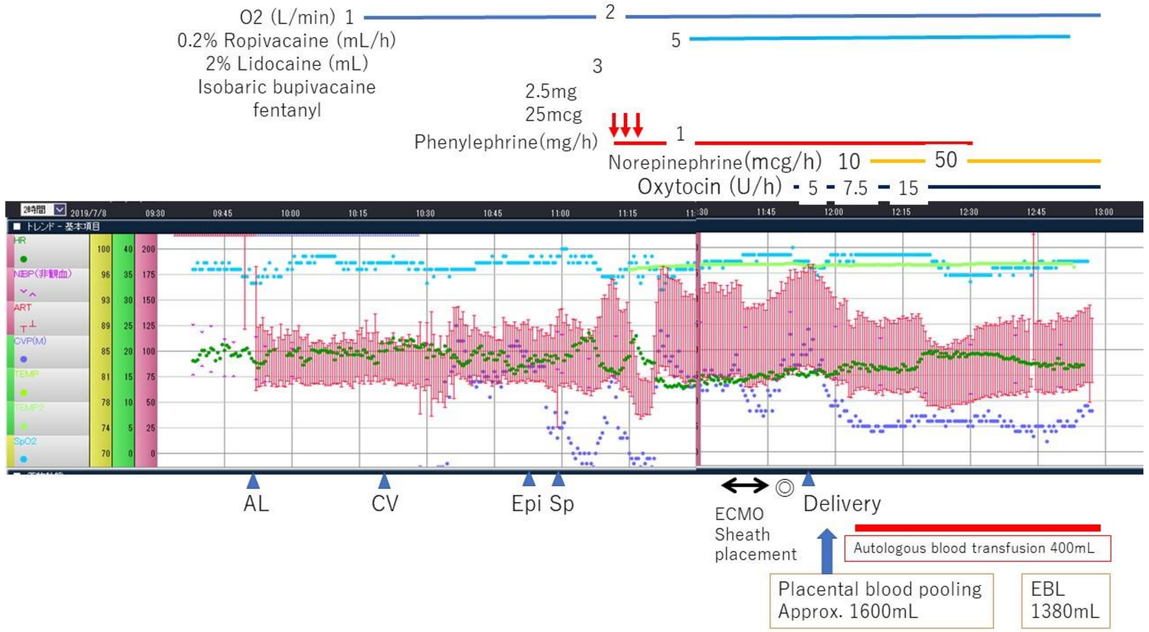


Fig4



- Spinal anesthesia was performed with isobaric bupivacaine 2.5 mg and fentanyl 20 mcg followed by a thoracic epidural placed at T10-11 with lidocaine 2% 3 ml resulting in sensory block to T4. Epidural ropivacaine 0.2% (5 mL/h) was administered during the CD.
- A right femoral vein sheath for emergency ECMO was placed in case of circulatory collapse.
- PDE3 inhibitor and nitroglycerin were prepared for possible pHTN increase. A phenylephrine infusion (16 mcg/min + 50 mcg bolus) was started for SVR maintenance.
- Delivery was uneventful (1400 g male, Apgar 8/9), with slow delivery of the placenta and oxytocin 5 units/h. The placenta was filled with approximately 1600 mL of blood (Fig 3B).
- Hemodynamic trends are shown in Fig 4. After oxytocin initiation, norepinephrine 10-50 mcg/h was started. The total estimated blood loss was 1380 mL. 400 mL of autologous blood was transfused.

After surgery

- The patient's postoperative recovery was uneventful except for pleural effusion treated with furosemide. Post-CD pain was managed with epidural analgesia (ropivacaine 0.1% with fentanyl 2 mcg/ml) for 48h and acetaminophen 720 mg q6h in the ICU. Furosemide and heparin were maintained until discharge on postop day 11.

Discussion

- To the best of our knowledge, this is the first case of a CD in a patient with CCTGA and severe pHTN.
- The aims of the anesthesia management were: 1) avoidance of stimuli exacerbating the PH; 2) maintenance of appropriate SVR to decrease the R-L shunt; and 3) avoidance of volume overloading associated with placental removal and uterine contraction.
- The overall perioperative management was successful. The blood pooling in the placenta might have contributed to stabilizing her hemodynamics in the manner of a physiologic phlebotomy at delivery.
- This case illustrates the importance of multidisciplinary team assessment and collaborative planning.

References

Arendt, Anesth Analg. 2008;107(6):1973–7.