An Unusual Case of a Parturient with Uncorrected Pentalogy of Fallot Presenting for Elective Cesarean Section Delivery of Twins

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Abstract

We present a 31-year-old primigravida with uncorrected pentalogy of Fallot, pregnant with monochorionic-diamniotic twins, undergoing elective lower segment cesarean section at 36 weeks gestation. Preoperative workup included a transthoracic echocardiogram which revealed a large ventricular septal defect of 1.8 cm with bidirectional shunting, a moderate size atrial septal defect of 1.8 cm with predominant left-to-right shunting, an overriding aorta, moderate right ventricular hypertrophy, and severe pulmonary valve stenosis. Notably, the patient was acyanotic with normal effort tolerance. Preoperative preparation involved the input of cardiologists and obstetric and cardiothoracic anesthetists. Issues such as the use of extracorporeal membrane oxygenation and cardiopulmonary support in the event of cardiac failure were discussed. Autotransfusion postdelivery was also addressed, and plans made for therapeutic venesection should need to arise. Intraoperatively, the planned anesthetic technique was slow and titrated combined spinal–epidural. However, a general anesthetic technique with rapid sequence induction was used in view of extreme patient anxiety. Intravenous induction was performed with ketamine and etomidate, followed by paralysis with succinylcholine. Anesthesia was maintained with desflurane on a mixture of air and oxygen. Phenylephrine infusion was titrated according to the patient’s blood pressure and systemic vascular resistance. The uterotonic of choice was duratocin given as a slow bolus, followed by a 4-h infusion of oxytocin. The patient was put in a head-up position to prevent venous air embolism and to decrease autotransfusion to central circulation. Postoperatively, she was extubated and sent to the Intensive Care Unit for continuous monitoring with FloTrac.

Keywords: Cesarean delivery, pentalogy of Fallot, twin pregnancy

INTRODUCTION

Patients with tetralogy of Fallot (TOF) are at risk of cardiovascular complications during pregnancy and are at higher risk of fetal loss.10 TOF is a congenital heart defect which, as the name suggests, involves four anatomical abnormalities: pulmonary infundibular stenosis, overriding aorta, ventricular septal defect (VSD), and right ventricular hypertrophy.

Pentalogy of Fallot (POF) is a variant of TOF with coexisting atrial septal defect (ASD). This condition is even rarer and more complex. Adverse maternal events are related to the magnitude of right-to-left shunting, which accentuates arterial hypoxemia. Conditions which may exacerbate the right-to-left shunt include a decrease in systemic vascular resistance (SVR) as well as an increase in pulmonary vascular resistance (PVR).

Patients with multiple pregnancies have been shown to have an even more hyperdynamic circulation than singleton pregnancies.13 This makes the pregnancy for patients with congenital cyanotic heart disease all the more challenging.

Previous case reports published have described patients who already had some forms of correction of their cyanotic heart disease and are mainly singleton pregnancies. Other case reports which describe patients with uncorrected disease describe the use of a regional technique for cesarean sections.

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There have also not been any reported cases of uncorrected cyanotic heart disease with multiple pregnancies. The previous cases reviewed in the literature also did not seem to have any issues with patient anxiety and the need for a general anesthetic (GA) technique in favor of a regional anesthetic (RA) technique due to this.

Hence, this case was challenging in multiple aspects.

**Case Report**

We present a 31-year-old primigravida with an uncorrected POF, pregnant with monochorionic-diamniotic (MCDA) twins, planning to undergo elective lower segment cesarean section (LSCS) at 36 weeks gestation. She was first diagnosed with POF at the age of 8 years but subsequently defaulted follow-up with cardiology. Her MCDA twins were conceived naturally.

This patient presented late at 30 weeks gestation to our institution. As such, there was no record of antenatal care before this. During her first presentation, she was immediately referred to a cardiologist specializing in congenital heart diseases and was worked up for her heart condition. A detailed ultrasound scan was also performed at this visit. She was given an early appointment with the obstetric anesthesiologist to perform a preoperative evaluation and assessment in the preparation for delivery. She was subsequently followed up weekly by her obstetrician until the planned date of delivery.

A thorough preoperative assessment was performed for this patient. She was a primigravida, 150 cm tall, weighing 48 kg at 36 weeks gestation (body mass index 21.3). Airway assessment was unremarkable with mouth opening >3 finger breadths; good range of neck movement, thyromental distance, and dentition. Assessment of her spine did not reveal any scoliosis, gross edema, or lesions on the overlying skin. Her heart rate was 80 beats/min, blood pressure (BP) was 106/77 mmHg, and auscultation revealed a continuous murmur at the upper left sternal edge, grade 4/6. Her jugular venous pressure was not elevated although she had bilateral lower limb pitting edema up to her mid-thighs which was attributed to dependant edema related to pregnancy. She was able to lie supine without any tachypnea or dyspnea and had normal effort tolerance and no history of Tet spells, there were neither crepitations nor rhonchi in her lungs, and baseline oxygen saturation on room air was 98%.

Blood investigations performed included a full blood count which showed a hemoglobin concentration of 12.3 g/dL. Coagulation profile (international normalized ratio 0.91) and platelet count were normal. Blood investigations done also showed normal renal function and normal serum electrolyte profile. No chest radiography was performed due to the pregnancy. Electrocardiogram showed a right bundle branch block with sinus rhythm at 85 beats/min. Further preoperative workup included a transthoracic echocardiogram performed at 31 weeks gestation that revealed a large VSD of 1.8 cm with bidirectional shunting, a moderate size ASD of 1.8 cm with predominant left-to-right shunting, an overriding aorta, moderate right ventricular hypertrophy, and severe pulmonary valve stenosis. Her ejection fraction was 71%.

A multidisciplinary approach involving cardiologists, neonatologists, and obstetric and cardiothoracic anesthesiologists was involved with facilitating the patient’s care. It was decided that delivery of the patient should ideally be performed at 36 weeks gestation. The risks of increasing oxygen demand and further stress on the heart due to the need for increased cardiac output would outweigh the benefits of further fetal maturation beyond this gestation. Ultrasound scans of the twins also showed that they were of a good weight and would have a good chance of survival even if they were delivered prematurely. Therapeutic venesection as well as cardiothoracic support with the use of extracorporeal membrane oxygenation (ECMO) will be instituted if the patient develops right ventricular failure from increased venous return from autotransfusion after delivery.

Considerations for the possibility of the patient going into labor before the elective date were given, and plans were made for the event that an emergency LSCS had to be performed. Both the on-call anesthesiology and obstetric teams were informed in the weeks, leading up to the planned date of delivery. For the planned surgery, both the obstetric and cardiac anesthesiologists were in the operating theater to conduct the anesthetic.

The options of RA and GA, if she was not able to proceed with RA, were explained to the patient. The patient was also informed of the need for invasive BP monitoring and postoperative intensive care. Other plans included the use of a flow-based continuous cardiac output monitoring device, FloTrac® (Edwards Lifesciences), which can be connected to the intra-arterial cannula and central venous catheter. The possible use of ECMO was also highlighted to the patient preoperatively.

The patient was fasted overnight and oral sodium citrate and oral ranitidine 150 mg was given on the morning of surgery. The patient was not on any other medications apart from her prenatal vitamins. Arterial and venous cannulations were performed under local anesthesia before the commencement of anesthesia. Despite the explanations and managing the operating theater settings, the patient was very anxious. Small doses of intravenous midazolam (0.25 mg boluses) had to be administered for anxiolysis. Invasive BP monitoring was achieved with a right radial 20G arterial cannula, to which the FloTrac® monitor was attached. A 3-lumen 7.5 Fr central venous pressure catheter was inserted in the right internal jugular vein along with a Swan-Ganz Sheath; the latter was inserted in the preparation for therapeutic venesection. Cardiothoracic surgeons were on standby for the insertion of lines in the event that ECMO was required. Antiembolic stockings and calf compressors were applied for thromboprophylaxis.
We were not able to carry out the initial plan for a slow and titrated combined spinal–epidural (CSE) as she was unable to tolerate or cooperate with the injection due to extreme anxiety. Hence, a GA technique with rapid sequence induction was used instead. Intravenous induction was performed with ketamine and etomidate, followed by paralysis with rocuronium. Anesthesia was maintained with desflurane on a mixture of air and oxygen, and the patient was paralyzed with rocuronium. Phenylephrine infusion was titrated according to the patient’s BP and SVR obtained from FloTrac®.

Delivery of the twins was uneventful with Apgar scores of both twins being 3 at 1 min of birth and 9 at 5 min. Neonatologists were on standby in the operating theater at the time of delivery to allow for immediate medical attention to be given to the twins after delivery. The uterine wall was stimulated with a slow bolus of duratocin. Patient-controlled analgesia (PCA) morphine was prescribed, with a total of 1 L of Hartmann’s solution throughout the operation, with an estimated blood loss of approximately 500 ml.

Continuous FloTrac® monitoring was utilized throughout the operation, and the phenylephrine infusion was titrated according to the SVR values on the monitor to maintain an SVR of 800–1200 dynes/sec/cm². She was extubated awake and sent to the Intensive Care Unit (ICU) for continued monitoring with FloTrac®. She was admitted to the ICU overnight and returned to the floor 3 days later. Patient-controlled analgesia (PCA) morphine was prescribed for the first 2 days postoperatively. Oral analgesics were prescribed and PCA discontinued once she was able to tolerate oral feeds. Subcutaneous cleaxane injections, which were administered postoperatively for prophylaxis against thromboembolism, were discontinued after the patient was able to ambulate out of bed. The patient was discharged well on the postoperative day 3.

### Discussion

In normal pregnancy, the following physiological changes occur. Cardiac output increases from the 5th week of pregnancy and reaches maximum levels at approximately 32 weeks gestation, after which there is only a slight increase until labor, delivery, and postpartum period. By the 8th week of pregnancy, 50% of the increase in cardiac output would have occurred. This increase in cardiac output is due to an increase in stroke volume as well as in heart rate.[3] In patients with twin pregnancies, it has been shown that maternal cardiac output was greater by 20% compared to singleton pregnancies because of a greater stroke volume (up to 15%) and heart rate (3.5%).[3]

There is an initial reduction in SVR and PVR to about 70% of prepregnancy levels by 8 weeks of gestation.[4] This decrease is likely related to increased concentrations of circulating estrogen, nitric oxide, and other vasodilatory peptides.[5] This is associated with a decrease in BP, which may be attributed to a relatively under-filled vascular state due to arterial and venous dilatation. BP decreases to a nadir at approximately 20 weeks gestation and returns to prepregnancy levels by full term.[5]

Patients with uncorrected congenital heart diseases such as POF are at greater risk of cardiovascular complications during pregnancy and labor. They are also at greater risk of fetal loss and have greater chances of developing adverse outcomes such as cardiac failure, arrhythmias, and reduced contractile function of the right ventricle in addition to the risks of cyanosis and cyanotic spells. The hemodynamic load in pregnancy, together with cardiac structural changes, accounts for about 7% of adverse cardiac events.[6]

The anesthetic considerations for patients with POF include avoiding elevations in PVR, maintaining SVR, maintaining myocardial contractility, maintaining preload, and avoiding reversal of shunt. It is pertinent to avoid hypercarbia, hypoxemia, acidosis, and pain, all of which may increase PVR and contribute to reversal of shunt. The best anesthetic technique for LSCS in these patients has yet to be established and recommendations available are based on case reports and pathophysiological concepts and theories.

Both GA and RA techniques using CSE with slow-titrated doses of local anesthetic have been described;[7] single-shot spinal anesthesia is contraindicated in this group of patients due to rapid decrease in SVR postspinal, which may result in a reversal of shunt.[8]

Use of CSE may be advantageous as it avoids airway manipulation and avoids the risk of aspiration. In a case report by Solanki et al.,[9] a low-dose CSE technique was described. They used a dose of 0.5 ml of 0.5% intrathecal bupivacaine with 25 µg fentanyl and sequential epidural bupivacaine supplementations (2–3 ml of 0.5% plain bupivacaine) to

### Table 1: FloTrac® values during the various stages of surgery

| Events            | MBP (mmHg) | CVP (mmHg) | CI (L/min/m²) | SVR (dynes/sec/cm²) |
|-------------------|------------|------------|---------------|--------------------|
| Pre-induction     | 107        | 8          | 3.9           | 1484               |
| Post-induction    | 62         | 13         | 3.8           | 727                |
| Post-delivery     | 111        | 18         | 4.0           | 1284               |
| Post-Operative    | 109        | 12         | 5.0           | 1071               |
achieve a block height of T4 for LSCS. The patient in this case report also had an uncorrected TOF and she was of similar height and weight to our patient. It is noted that their patient had a short neck, was Mallampati III, and had a receding chin. Hence, a central neuraxial technique may be preferable to avoid airway manipulation and circumvent the problems of a potentially difficult airway.

With GA, the risks of hemodynamic alterations due to laryngoscopy, as well as problems associated with manipulation of airway, have to be considered. However, due to our patient’s high level of anxiety and inability to cooperate with a regional technique, GA was chosen. The benefits of GA include controlling ventilation and hence providing better oxygenation. Keeping in mind the problems associated with GA, we planned our anesthetic management to maintain hemodynamic stability. We chose to combine etomidate and ketamine for induction of anesthesia to avoid marked reductions in SVR. Ketamine also has analgesic properties. Intravenous lignocaine 50 mg was also given at induction to blunt the hemodynamic reflex to laryngoscopy. Nitrous oxide was avoided due to its propensity to cause elevations in PVR. The slow administration of duratocin followed by an infusion of oxytocin avoided a sudden decrease in SVR. After delivery, the babies were attended immediately by the neonatologists who were on standby in the operating theater. Care was taken to administer the narcotics only after the babies were delivered to minimize the effect of these drugs on the newborns.

Intermittent positive pressure ventilation may result in decreased venous return, compression of pulmonary vessels, hypoxemia, hypercarbia, and acidemia. Hence, our ventilation strategy for this patient was targeted at maintaining oxygenation with tidal volumes of 6–7 ml/kg, minimal positive end-expiratory pressure and controlling the respiratory rate to prevent hypercarbia. Arterial blood gas was performed to monitor the pH, pO₂, and pCO₂ of the patient to ensure that we were ventilating the patient adequately. The cardiothoracic surgeons and perfusionists were on standby for femoral–femoral bypass and ECMO in the event that there was shunt reversal or sudden increase in PVR resulting in inadequate oxygenation.

Intraoperatively, we used FloTrac® to achieve our anesthetic management goals which were mainly hemodynamic and cardiovascular stability. The use of FloTrac® allowed us beat-to-beat monitoring and real-time monitoring of cardiac index, cardiac output, SVR, and stroke volume variation. Of special importance was the monitoring of SVR which was maintained between 800 and 1200 dynes/sec/cm⁵. A phenylephrine infusion to prevent reversal of shunt. FloTrac® was chosen as our cardiac output monitor as it can easily be attached to the existing arterial cannula and central venous catheter. We did not insert a pulmonary artery catheter as the presence of a large VSD renders the pulmonary wedge pressure measurement inaccurate in reflecting left ventricular filling pressures.

This case was challenging in many aspects. One of the areas we had to consider was that of the ethics surrounding this case. This was a high-risk pregnancy and a high-risk operation considering her cardiac condition and multiple gestation. Had there been any crisis during the surgery, our duty would first be to the patient while simultaneously trying our best to give the twins a fighting chance. As such, contingency plans such as the use of ECMO and therapeutic venessection had to be made in advance to reduce the reaction time should trouble arise during the surgery. The other consideration was that of future pregnancies for this patient given the complexity of her condition. We did advice and counsel the patient regarding further pregnancies, and it was her wish not to perform any permanent procedures to prevent further pregnancies. We had to respect her autonomy and gave her advice to seek medical attention promptly should she become pregnant again in the future.

**Conclusions**

Interprofessional collaboration through a multidisciplinary team-based approach and comprehensive preoperative planning for different eventualities underlined the smooth perioperative management of a parturient with uncorrected POF for elective cesarean delivery of twin newborns.

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**Conflicts of interest**

There are no conflicts of interest.

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