

Use of a Combined Anaesthetic Technique and Minimally Invasive Hemodynamic Monitoring in a Primigravid with Uncorrected Tetralogy of Fallot for Caesarean Section

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ABSTRACT

Tetralogy of Fallot (TOF) in pregnancy is a rare occurrence which poses a high risk for detrimental effects on both mother and fetus. This paper reports a 21-year-old primigravid diagnosed with uncorrected TOF who had a successful caesarean section at 32 weeks of gestation. To address the hemodynamic challenges, the anaesthetic management involved the use of a minimally invasive hemodynamic monitor, controlled mechanical ventilation and a combined technique of intravenous anaesthesia using remifentanyl and lumbar epidural anaesthesia using levobupivacaine.

Keywords: Tetralogy of Fallot, pregnancy, general anaesthesia, remifentanyl, epidural anaesthesia

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease.¹ The disease is comprised of four anatomic abnormalities including ventricular septal defect, right ventricular hypertrophy, overriding of the aorta, and pulmonary stenosis.¹ In the absence of surgical repair, only 10% of patients reach the third decade of life while only 3% reach the fourth decade and beyond. In pregnancy with uncorrected TOF, maternal mortality is increased by 12% from the normal population.² The pathophysiologic changes of TOF, on top of the physiological changes of pregnancy, elevate the risk for potential harmful outcomes by 7%.³ Complications leading to maternal hypoxemia in uncorrected TOF may result in fetal growth retardation (36%), fetal death (14%) and maternal death (10%).⁴ The complex demands of both the mother and fetus pose difficulty in anaesthetic management.

CASE SUMMARY

A 21-year-old primigravid diagnosed with Tetralogy of Fallot was scheduled for elective lower segment caesarean section at 32 weeks age of gestation (AOG). She was diagnosed with TOF at the age of 5 years old presenting with increased frequency of dyspnea and cyanotic spells on exertion. The patient was advised corrective surgery but was lost to follow up due to financial constraints. She has no other known illnesses, maintenance medications, previous hospitalizations nor surgeries.

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During pregnancy, the patient was maintained on ferrous sulfate, calcium carbonate and aspirin. At 27 5/7 weeks AOG, biophysical profile and congenital anomaly scan were done revealing fetal congenital heart disease (atrial septal defect, cardiomegaly and pericardial effusion) which then prompted close monitoring at the intensive care unit.

At 31 5/7 weeks of gestation, she had easy fatigability, exertional dyspnea and no neurologic symptoms of syncope, stroke and seizures. She was noted to be awake and not in cardiorespiratory distress, weighing 47 kilograms, with a body mass index of 20. Vital signs were BP of 110/70 mmHg, HR of 93 bpm, RR of 24 cpm, SpO₂ of 82% on room air, increasing to 87% with oxygen support at 10 liters per minute (LPM) via Hudson face mask. She presented with cyanotic lips, jugular venous distention, clubbing and bipedal edema. Cardiac exam showed an adynamic precordium, with the apex beat localized at the 5th left intercostal space within the midclavicular line, two grade 3/6 systolic murmurs on the second intercostal space, left parasternal border, and fourth intercostal space, left, radiating to the right parasternal border. Abdominal examination showed a gravid abdomen with a single live fetus in cephalic presentation, with good fetal heart tones. Airway assessment showed Mallampati Class 2 with no other airway anomalies.

Laboratory tests revealed polycythemia, normal platelet count, coagulation profile and serum electrolyte results. ABG showed compensated metabolic acidosis, SpO₂ of 78%, severe hypoxemia and increased alveolar-arterial gradient from shunting. Chest x-ray revealed cardiomegaly and no pulmonary infiltrates. Cardiac tests and imaging showed regular sinus rhythm with occasional premature ventricular contractions, ventricular septal defect with a left to right shunt, overriding of the aorta, right ventricular hypertrophy, pulmonary stenosis, dysplastic main pulmonary artery and pulmonary valve, elevated right ventricular systolic pressure, concentric remodelling of the left ventricle with adequate contractility and global systolic function. Tricuspid valve prolapse with moderate tricuspid and mild aortic regurgitation was seen with an ejection fraction of more than 50%.

A decision was reached to perform an elective caesarean section at 32 weeks AOG in consideration of the maternal disease state and fetal maturation. Prior to the procedure, aspirin and enoxaparin were stopped, and omeprazole was given for aspiration prophylaxis. In the operating room, standard non-invasive monitors were attached to the patient. Preoperative vital signs were BP of 120/70 mmHg, HR of 99 bpm, RR of 23 cpm and SpO₂ of 88% with oxygen support at 10 LPM via Hudson face mask. Midazolam 0.02 mg/kg was given prior to the insertion of a lumbar epidural catheter.

Midazolam 0.02 mg/kg and a target-controlled infusion (TCI) of remifentanyl at 0.8 ng/mL was started prior to the placement of central venous and arterial catheters which were used to continuously monitor hemodynamic indices via the EV1000 clinical monitoring platform.

Table 1. FloTrac® values during the course of the procedure

Event	CO L/min	CI (L/min/m ²)	SVR (dyne-s/cm ⁵)	SVV (%)
Normal value ⁵	4-8	2.5-4.0	800-1200	<10-15
Pre-Induction	5.7	4.2	1264	11
Post-Induction	5.2	3.8	1462	3
Post-Delivery	5.0	3.6	1266	7
Post-Operative	5.1	3.7	1465	10

CO - cardiac output, CI - cardiac index, SVR - systemic vascular resistance, SVV - stroke volume variation

She was then preoxygenated and oropharynx was sprayed with lidocaine. Remifentanyl infusion target was increased to 2 ng/mL and ketamine 50 mg (1 mg/kg) and rocuronium 30 mg (0.6 mg/kg) were given. Intubation led to minimal changes in the vital signs except the SpO₂ which ranged between 92 to 95% with FiO₂: 1.0. Intravenous anaesthesia was maintained with remifentanyl infusion at 2 ng/mL and boluses of ketamine 1 mg/kg and midazolam 0.05 mg/kg every 30 minutes. A loading dose of 10 mL of 0.5% levobupivacaine was given via the epidural catheter and an infusion of phenylephrine 0.2 mcg/kg/min was started. Normal systemic vascular resistance (SVR), cardiac output (CO) and a stroke volume variation (SVV) below 13% were maintained throughout the surgery. Hemodynamic values from the EV1000 monitor during the different phases of the procedure is shown in Table 1.

Plateau pressures were maintained at 11-12 cm H₂O, ETCO₂ at 32-36 mmHg and SpO₂ at 92-95%. A live baby with an APGAR score of 9, 9 was delivered 3 minutes from intubation and was immediately brought to the neonatal intensive care unit for further care. The procedure lasting 51 minutes was uneventful. Prior to discontinuing the remifentanyl infusion, 10 mL of 0.03% morphine sulfate was given via the epidural catheter. Neuromuscular blockade was reversed with sugammadex 3 mg/kg and the patient was extubated fully awake with stable vital signs. Postoperative pain medications included 10 mL of 0.03% morphine sulfate via epidural every 12 hours for 3 doses, IV paracetamol 12 mg/kg every 6 hours for 4 doses and IV tramadol 0.1 mg/kg every 8 hours for breakthrough pain. Postoperative course was unremarkable. Compared to preoperative values, acid-base status improved and the patient was discharged after 15 days.

DISCUSSION

Without corrective surgery, the average life expectancy of patients with Tetralogy of Fallot reaching adulthood is 12 years.⁶ Patients usually have signs and symptoms of hypoxia, right-sided heart failure and thromboembolism. In this case, the patient presented with hypoxia with a baseline SpO₂ of 82%, an increased alveolar-arterial gradient and had symptoms of right-sided heart failure such as distended

neck veins, easy fatigability, exertional dyspnea and bipedal edema. Several factors may contribute to increased survival in uncorrected TOF such as left ventricular hypertrophy and extracardiac shunting. In this patient, left ventricular hypertrophy was noted with a left ventricular diastolic thickness of 11 mm, left ventricular mass index of 90 g/m² and a relative wall thickness of 0.62. Prognosis is worsened by the following risk factors: pre-pregnancy hematocrit more than 65%, history of cardiac failure or syncope, cardiomegaly, right ventricular pressure exceeding 120 mmHg, strain pattern in ECG and SpO₂ less than 80%.¹ Among these risk factors, only cardiomegaly was present in the patient based on echocardiography results. The patient's current condition demands certain hemodynamic considerations during surgery. Right to left shunting, decrease in pulmonary blood flow and worsening of right ventricular outflow tract (RVOT) obstruction should be avoided, however, the surgery and the expected changes during pregnancy complicates achieving these goals.

During pregnancy, the maternal cardiovascular system adapts to address the increased oxygen consumption and metabolic demands of a growing fetus. Endothelium dependent factors, estrogen and nitric oxide mediate peripheral vasodilatation producing a concomitant 25 to 30% decrease in SVR.⁷ As a result, stroke volume increases leading to an increase in cardiac output by 30 to 50%.^{7,8} The physiologic decrease in SVR accentuates the right to left intracardiac shunt leading to a greater risk for severe maternal hypoxemia and cyanosis. Hypoxemia increases pulmonary vascular resistance (PVR), which, in turn, worsens right to left shunting. The patient is at an increased risk for adverse cardiac events since her baseline SpO₂ is below 85%.^{9,10} Polycythemia is a systemic compensation, as evidenced by the patient's hematocrit of 53%. In the presence of an already compromised arterial oxygen content, maintaining adequate cardiac output is essential in ensuring sufficient oxygen delivery. This makes the impact of pregnancy and caesarean delivery on the RVOT obstruction an alarming concern. The physiologic decrease in SVR and the sympathetic stimulation caused by surgical pain leading to increased contractility worsen the RVOT obstruction. The hemodynamic dilemma imposed by the uncorrected TOF, pregnancy and surgery were all considered in the decision to control the patient's ventilation, to use a combined anaesthetic technique and to use minimally invasive hemodynamic monitoring.

Prior to induction of anaesthesia and intubation, lidocaine spray was used and remifentanyl infusion was increased to 2 ng/mL to address the pain of laryngoscopy. Pain causes sympathetic stimulation which may lead to increased contractility and worsening of RVOT obstruction. Most agents for induction of general anaesthesia produce a decrease in SVR, which may in turn increase right to left shunting. In this case, ketamine was used because of its property of having a minimal effect on SVR and PVR, therefore decreasing the magnitude of a right to left shunt.²

Ketamine's issue of hypertension and cardiovascular stimulation was attenuated with the use of midazolam.

The patient's airway was intubated and ventilation was controlled to minimize hypoxia and hypercarbia which can increase pulmonary vascular resistance and worsen right to left shunting. Although positive pressure ventilation decreases blood flow to the pulmonary vasculature, mechanical ventilation was set to deliver adequate minute ventilation while minimizing the plateau and driving pressures, ensuring normal SpO₂ and ET-CO₂. No PEEP was used and anaesthesia was maintained with TCI remifentanyl and epidural levobupivacaine.

Remifentanyl is a titratable, short-acting mu opioid receptor agonist which provides rapid onset and offset analgesia with a potency at least one hundred times that of morphine. It provides hemodynamic stability, reducing maternal hypertension and tachycardia. Sympathetic stimulation is diminished, thereby preventing the worsening of RVOT obstruction. Remifentanyl has a very minimal vasodilating effect on the pulmonary vasculature, has an onset of action at 1 minute, peak effect at 2 minutes and duration of action of 20 minutes.¹¹ Its short context-sensitive half-life of 3 minutes makes it independent from its duration of infusion. This drug is metabolized by nonspecific plasma esterases allowing degradation regardless of organ function. Remifentanyl exhibits transplacental passage as confirmed by an umbilical vein/maternal artery ratio of 0.88. Drugs and metabolites that cross the placenta consequently have effects on the fetus, however, remifentanyl demonstrates rapid fetal metabolism and redistribution resulting in less fetal adverse effects compared to other opioids. Superior neonatal APGAR scores were seen and the need for endotracheal intubation due to neonatal respiratory depression was less with remifentanyl compared to fentanyl.¹² In this case, a 32-week-old neonate was delivered and had an APGAR score of 9, 9, which is in congruence with the mentioned findings.

Epidural levobupivacaine was used for supplemental analgesia to avoid further sympathetic stimulation. Unlike spinal anaesthesia, epidural anaesthesia allows a gradual decrease in systemic vascular resistance which may be offset by fluid loading and vasopressor use. Levobupivacaine was preferred because it causes less vasodilation than bupivacaine and is less associated with cardiac and central nervous system toxicity.^{13,14} In this case, pure levobupivacaine was used at a minimum volume required for caesarean section. A study showed that the use of a higher concentration prolongs the duration of sensory and motor blocks without increasing the incidence of adverse side effects. On the other hand, using both a higher volume and concentration of levobupivacaine is associated with an increased risk for hypotension.^{14,15} The use of an epidural catheter is also of importance post-operatively allowing the delivery of epidural morphine to provide adequate pain control after surgery.

A combined technique was chosen which allowed the use of a lower dose of remifentanyl and a lower volume

of levobupivacaine while preserving adequate depth of anaesthesia. This technique decreased the vasodilating effects of both drugs which helped maintain hemodynamic stability.

Phenylephrine infusion was used to counteract the expected decrease in systemic vascular resistance that ensued from the use of remifentanyl and epidural levobupivacaine. In patients with TOF, the resulting increase in blood pressure augments pulmonary blood flow and enhances oxygenation.^{4,16}

The use of a minimally invasive hemodynamic monitor allowed targeted management of the hemodynamic variables affected by the consequences of uncorrected TOF, pregnancy and surgery. To reduce RVOT obstruction, SVV was maintained lower than 13% with judicious hydration to ensure adequate intravascular volume. CO was maintained within the normal range of 4-6 L/min to limit unnecessary increases in contractility. Aggravation of hypoxemia was prevented by ensuring adequate CO and maintaining SVR above 800 dyne-s/cm⁵ to prevent right to left shunting.

Over all, the patient had an uneventful perioperative course. Postoperative ABG showed no acid-base derangement which may be attributed to the proper perioperative management of ventilation, volume and hemodynamic status.

CONCLUSION

This is the first known report of successful use of a combined technique using remifentanyl TCI and epidural levobupivacaine in a patient with uncorrected TOF for caesarean section. The various components of the management including the use of combined anaesthetic technique, controlled ventilation, phenylephrine infusion and continuous invasive hemodynamic monitoring complemented each other making it a safe and effective approach in such patients.

Statement of Authorship

Dr. Lauren L. Laforteza participated in writing both original and final manuscript while Dr. Maria Teresita B. Aspi reviewed and approved the final submitted report.

Author Disclosure

Both authors declared no conflicts of interest in preparing this article.

Funding Source

No specific grant from any funding agency in the public, commercial or not-for-profit sectors was received in writing this paper.

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