

## Case report of a caesarean delivery in a primigravida with congenitally corrected transposition of the great arteries

PS Odendaal,  JL Burke 

Department of Anaesthesiology and Critical Care, Stellenbosch University, South Africa

Corresponding author, email: [psodendaal91@gmail.com](mailto:psodendaal91@gmail.com)

We present a case of a 27-year-old primigravida with complex congenital heart disease who underwent an elective caesarean delivery at a tertiary institution in South Africa. Pregnancy has profound implications for this subset of patients. Vaginal delivery with epidural analgesia is safe in parturients with congenital heart disease. In this case, however, operative delivery was indicated, and we conducted a general anaesthetic under intraoperative transoesophageal echocardiography monitoring.

**Keywords:** congenital heart disease, transposition of great arteries, pregnancy, transoesophageal echocardiography, caesarean delivery

### Case report

A 27-year-old primigravida at 25 weeks gestation presented for follow-up at the obstetric high-risk clinic. She was known to the division of Cardiology to have complex congenital heart disease (CHD) consisting of congenitally corrected transposition of the great arteries (L-TGA or CCTGA), severe pulmonary stenosis and a large non-restrictive ventricular septal defect (VSD). Palliative Blalock-Taussig shunts were performed at the ages of 2 months and 2 years. Obstetric and perioperative risks were explained to the patient prior to conception via several multidisciplinary team (MDT) sessions.

History revealed no chronic medication, a baseline New York Heart Association (NYHA) dyspnoea grading of 1, with no orthopnoea, paroxysmal nocturnal dyspnoea, angina pectoris, cyanotic spells, syncope or palpitations. No associated dysmorphism, syndromes or significant family history were reported.

Her functional status changed from NYHA grade 1 to grade 2 at 25 weeks gestation, and she was admitted to hospital. The MDT decided that after appropriate steroid therapy, delivery of the foetus was indicated at 28 weeks gestation. A breech presentation was diagnosed and operative delivery was planned.

At 27 weeks gestation, the patient's arterial haemoglobin saturation (SpO<sub>2</sub>) decreased from a baseline of 92% to 77%, and she was transferred to the Obstetric Critical Care Unit (OCCU). Her SpO<sub>2</sub> improved to 85% on a 40% O<sub>2</sub> Venturi mask, while she was tachypnoeic with a respiratory rate of 32 breaths per minute. In the OCCU, the patient was afebrile, not acutely distressed and able to lie supine. She had a pulse rate of 105 beats per minute, varying systolic blood pressure of 80–95 mmHg and diastolic blood pressure of 40–50 mmHg.

General examination revealed clubbing and central cyanosis. Focused cardiovascular examination showed regular pulses, no radio-femoral delay, a non-displaced apex, palpable parasternal thrill during systole, and left parasternal heave. This correlates

with a grade 5/6 holosystolic murmur, radiating from the central precordium to the rest of the anterior chest. No signs of congestive cardiac failure were present.

Laboratory investigations showed a haemoglobin level of 14.1 g/dL, leucocytosis of  $17.39 \times 10^9/L$ , and normal platelet count. Arterial blood gas analysis revealed a pH of 7.43, pCO<sub>2</sub> of 4.0 kPa, pO<sub>2</sub> of 7.1 kPa and bicarbonate of 22.1 mmol/L on FiO<sub>2</sub> of 0.4.

An early obstetric ultrasound estimated gestational age. On subsequent admission of the patient at 25 weeks gestation, the estimated foetal weight was 1.4 kg, in breech position and a fundal placenta.

Prior to pregnancy, an electrocardiogram gated magnetic resonance image (ECG gated MRI) (Figure 1) and formal transthoracic echocardiogram both reported situs solitus, atrioventricular discordance (the right atrium emptying into the left ventricle, and the left atrium emptying into the right ventricle), transposed great vessels, a double outlet right ventricle, significant right ventricular hypertrophy, a large non-restrictive VSD, severe pulmonary stenosis, a marginally narrowed left pulmonary artery trunk, with normal aortic and other major vessel morphology. The Qp:Qs ratio (ratio of pulmonary to systemic blood flow) was decreased at 0.5 and indicated a right-to-left shunt. The ejection fraction (EF) was 70% with normal cardiac chamber dimensions, no regional wall motion abnormalities, and normal morphology of other valves. Upon the patient's admission to hospital, a formal transthoracic echocardiogram was performed at 25 weeks gestation and showed no acute changes in ventricular function.

A chest radiograph was requested when desaturation occurred. Multiple nodular infiltrates were noted in her lung fields. In conjunction with her leucocytosis and a negative COVID-19 polymerase chain reaction (COVID-19 PCR) test on a nasopharyngeal swab, a community-acquired pneumonia (CAP) was diagnosed. She improved with antibiotic treatment, and operative delivery proceeded at 28 weeks gestation.

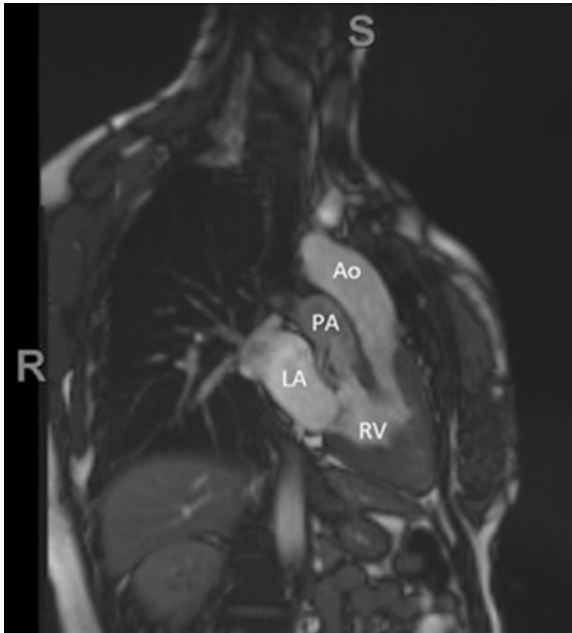


Figure 1: ECG gated cardiac MRI

Note: The presence of a hypertrophied double outlet right ventricle and transposed great vessels are seen  
LA – left atrium, RV – right ventricle, PA – pulmonary artery, Ao – aorta

The considerations for anaesthesia management were as follows:

- Understanding the complex heart disease
- Determining the impact of pregnancy physiology on the disease
- Understanding the effect of surgical delivery on the maternal physiology
- Planning optimal analgesia
- Planning postoperative care

The haemodynamic goals were to maintain adequate preload while preventing a decrease in systemic vascular resistance (SVR), which could worsen the right-to-left shunt fraction. Therefore, a decision was made to conduct general anaesthesia, including invasive arterial blood pressure and transoesophageal echocardiography (TOE) monitoring.

Preoperative optimisation consisted of fluid resuscitation, ceftriaxone and azithromycin for CAP, and thromboprophylaxis using enoxaparin. Betamethasone doses were administered for foetal lung maturation. The foetal and placental positions, as well as foetal weight, were confirmed preoperatively. A blood crossmatch was requested for possible haemorrhage.

In theatre, the patient was cooperative and able to lie supine. Aorticaval compression was alleviated by a left lateral tilt using a right-sided obstetric wedge. An 18-gauge intravenous cannula was sited peripherally, and 2 g cefazolin administered 30 minutes pre-incision. A radial arterial catheter was inserted for continuous invasive arterial pressure monitoring in addition to standard ASA monitoring. A central venous catheter (CVC) was sited in the right internal jugular vein, under ultrasound guidance.

The obstetric, anaesthesia, nursing and paediatric teams performed the WHO Surgical Checklist to confirm the operative

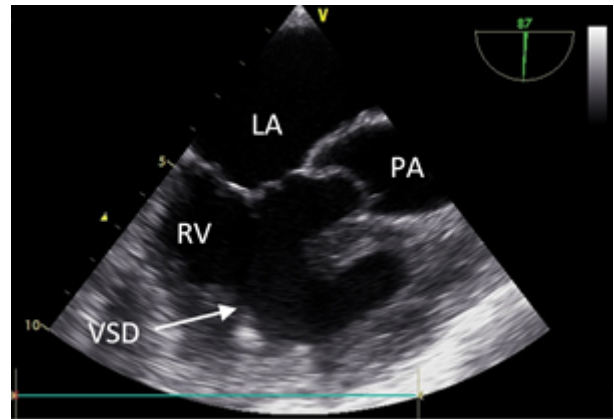


Figure 2: Intraoperative transoesophageal echocardiography image showing a large ventricular septal defect, transposed vessels, severe pulmonary stenosis, and right ventricular hypertrophy

LA – left atrium, RV – right ventricle, PA – pulmonary artery, VSD – ventricular septal defect

plan. Surgical site skin preparation was performed while the patient was preoxygenated. A cardiac-stable induction was performed. After etomidate 10 mg, fentanyl 250 µg, and rocuronium 50 mg was given, tracheal intubation was performed with a size 7.0 mm endotracheal tube, and dexamethasone 4 mg intravenous injection (IV) was administered. Transoesophageal echocardiography guided haemodynamic management (Figure 2).

The foetus was delivered via a classical incision with an induction-to-delivery time of less than 2 minutes. Blood pressure and SpO<sub>2</sub> remained constant throughout the procedure. A variable-rate phenylephrine infusion maintained the mean arterial blood pressure within 25% of baseline values. One litre Ringer's lactate crystalloid was infused intraoperatively. Blood loss was 600 ml in total.

Maintenance anaesthesia consisted of isoflurane at 0.5 minimum alveolar concentration (MAC) in oxygen and air. Multimodal analgesia consisted of fentanyl 250 µg IV on induction, a slow infusion of morphine 10 mg IV and paracetamol 1 g IV after delivery, as well as ultrasound-guided bilateral transversus abdominis plane blocks after surgery. Oral analgesics and IV morphine via a patient-controlled analgesia pump were administered in the postoperative period.

Uterotonics included oxytocin 2 international units (IU) by slow intravenous bolus followed by 20 IU/L crystalloid at a rate of 125 ml/h initiated after delivery.

After the reversal of muscle relaxation, the patient was extubated and transferred to the OCCU where she had an uneventful stay. She was discharged five days later. The premature neonate was admitted to the neonatal intensive care unit (NICU) for monitoring. Screening for congenital anomalies was negative. Both mother and baby were in good health one month after surgery, when the baby was discharged from hospital.

## Discussion

CHD is a rare pathology that poses multiple challenges to the anaesthesiologist during non-cardiac surgery. The current incidence of 9 per 1 000 births worldwide, is increasing.<sup>1</sup> Data suggest that low- and middle-income countries (LMIC) have a greater burden of disease than high-income countries (HIC), accounting for 96% of children born with congenital anomalies. CHD accounts for 57.4% of cardiac disease during pregnancy across all income groups.<sup>2</sup>

Congenitally corrected transposition of the great arteries is classified as a univentricular heart.<sup>3</sup> It predisposes the patient to complete atrioventricular block, tricuspid regurgitation and heart failure.<sup>4</sup>

Physiological changes during pregnancy, such as an increase in plasma volume, tachycardia, and a decrease in systemic vascular resistance, may precipitate cardiac failure. Risk factors for cardiac failure include the process of labour, pain associated with labour and the postpartum increase in preload.<sup>4</sup> Surgical delivery, in addition to anaesthesia techniques, confound the management of the at-risk parturient.

The Modified World Health Organization Classification of Cardiovascular Disease in Pregnancy and the Maternal Levels of Care Consensus Statement together assist in appropriate triage of parturients with cardiovascular diseases. In vaginal delivery for L-TGA, regional anaesthesia is advised, with assisted delivery to facilitate the second stage of labour, unless an indication for operative delivery exists.<sup>4</sup> A 'pregnancy heart team' consisting of a cardiologist, obstetrician, anaesthesiologist and perinatologist is recommended to manage complex cardiac disease during pregnancy. Delivery should take place at a centre with adequate expertise and resources.<sup>5</sup>

### Airway and breathing

Hypoxaemia and cyanosis are caused by poor ventricular function, shunting via cavopulmonary conduits, pulmonary hypertension, abnormal pulmonary arterial venous connections, and coexisting pulmonary disease.

The decrease in SVR during pregnancy may increase right-to-left shunting and lead to further hypoxaemia. Dyspnoea or hypoxaemia when supine, may warrant a prudent general anaesthetic in anticipation of cardiac failure after delivery.<sup>5</sup>

Maintenance of spontaneous breathing is ideal, but would impair the intra-abdominal surgical access. The detrimental effect of positive pressure ventilation on cardiac output can be reduced by minimising mean airway pressure.<sup>6</sup> Low tidal volumes with low positive end-expiratory pressure (PEEP) were used, and modifiable triggers for pulmonary hypertension were avoided (pain, hypovolaemia, hypoxaemia, hypercarbia, acidosis and hypothermia).

## Circulation

Maintenance of preload and preservation of inotropy, lowering pulmonary vascular resistance (PVR) and maintaining afterload, are important haemodynamic goals in right-to-left shunt cardiac lesions.<sup>5</sup> The patient with a univentricular heart is sensitive to changes in preload and PVR.<sup>6</sup> Passive leg raising during induction of anaesthesia can limit the associated decrease in cardiac output.<sup>3</sup>

The risk of acute cardiac complications necessitates advanced haemodynamic monitoring. Invasive arterial and TOE monitoring of preload, biventricular function and cardiac output assisted with clinical decisions regarding this patient under general anaesthesia.

Negative inotropic agents should be avoided, and SVR should be maintained with an alpha agonist such as phenylephrine, to maintain coronary perfusion and prevent the worsening of a right-to-left shunt. It is important to maintain a sinus rhythm and have appropriate resuscitation equipment available in theatre. In patients who undergo surgery for univentricular heart physiology, central venous catheterisation can be safely performed.<sup>7</sup>

## Drugs

General anaesthesia can influence neonatal outcomes in emergency cases.<sup>8</sup> Good theatre preparation and team communication will result in short induction-to-delivery time. This contributes to favourable outcomes for both mother and child.

Both the American Heart Association and European Society of Cardiology do not recommend antibiotic prophylaxis for infective endocarditis during elective vaginal or caesarean delivery.<sup>5</sup> In HIC, the risk for infective endocarditis is low in patients with CHD. However, in LMIC, appropriate antibiotic administration 30 minutes preoperatively must be considered for high-risk patients or those known to have rheumatic heart disease.<sup>9</sup>

A titrated oxytocin infusion is generally well tolerated. Ergot alkaloids may severely increase PVR and should be used with caution in intracardiac shunt lesions.<sup>5</sup>

## Surgery

In our patient, the breech presentation necessitated an operative delivery via classical incision, with greater potential for haemorrhage. Omission of uterine exteriorisation minimised the risk of air embolism in this high-risk patient, and right-to-left shunting necessitated air filters on all IV lines.

## Conclusion

This case demonstrates the benefit of a 'pregnancy heart team', the safe conduct of general anaesthesia and the value of TOE as continuous monitoring in complex cardiac disease during caesarean delivery.

## Conflict of interest

The authors declare no conflict of interest.

### Funding source

No funding was required.

### Ethical approval

Written informed consent for the publication of the patient's clinical details and images was obtained from the patient. Identifiable information of the patient is omitted on published images. Ethical approval to report this case has been obtained from the Health and Research Ethics Committee (HREC), Stellenbosch University, Western Cape (reference number C21/08/030).

### ORCID

PS Odendaal  <https://orcid.org/0000-0002-1567-736X>

JL Burke  <https://orcid.org/0000-0003-2805-9584>

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