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CASE REPORT

Anaesthesia for caesarean section in a patient with uncorrected Tetralogy of Fallot complicated by eclampsia

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Congenital heart disease (CHD) occurs in approximately 1% of the global population. Tetralogy of Fallot (TOF) is the most common cyanotic lesion.[1] Survival until adulthood is rare without surgery.[2] Resource-constrained settings predispose patients to reaching reproductive age without diagnosis or correction, and increase perioperative morbidity. Uncorrected TOF is challenging to manage during pregnancy and delivery, with increased risk of obstetric, cardiac and fetal complications.[3] The incidence of preeclampsia in these patients is the same as for the global population.[4] We describe the emergency management and caesarean delivery for a woman with uncorrected TOF, complicated by eclampsia and postpartum haemorrhage (PPH).

Keywords: anaesthesia, eclampsia, Tetralogy of Fallot, congenital heart disease, preeclampsia

Case report

A 29-year-old woman with severe preeclampsia and uncorrected Tetralogy of Fallot (TOF) presented in labour to a resource-limited South African hospital. Gestational age was estimated at 33 weeks based on an ultrasound performed the previous week, during assessment and referral to a high-risk antenatal clinic. Notes from the 32-week visit documented uneventful pregnancy, and a successful caesarean delivery 11 years earlier. She was not receiving medications for cardiac disease. A full blood count, electrolytes and renal function were within normal limits. Her haemoglobin was 13.6 g/dL and platelet count 175 x 10^9/L. An electrocardiogram showed signs of right ventricular hypertrophy (RVH) and axis deviation. Echocardiography demonstrated all features of TOF, with right ventricular wall hypertrophy measuring 26 mm, severe pulmonary stenosis (pressure gradient 78 mmHg) and preserved ejection fraction (77%).

Unfortunately, she missed the high-risk clinic appointment, and was referred in labour with blood pressure (BP) of 157/103 mmHg, proteinuria and bilateral pitting oedema. Examination revealed a 4/6 pansystolic murmur, a respiratory rate of 18/min, and clear lung fields, with peripheral oxygen saturation (SpO_2) of 88%. She received magnesium sulphate, methyldopa, dexamethasone and nifedipine for tocolysis prior to transfer. During re-admission her clinical condition deteriorated, with BP 95/56 and SpO_2 80%. Urine production was scanty with accompanying anasarca, and she became restless. Nursing staff administered oxygen and called for help. When she suffered an eclamptic seizure, it was decided to expedite delivery by emergency caesarean section (CS).

She was postictal on arrival in theatre. After placement of an intra-arterial line, general anaesthesia (GA) was induced using alfentanil, etomidate and suxamethonium, and maintained with sevoflurane and nitrous oxide in oxygen. She required vasopressor support, initially with phenylephrine and adrenaline boluses, and later an adrenaline infusion of up to 0.4 µg/kg/min. Neither noradrenaline nor vasopressin were available. Decreases in mean arterial pressure below 80 mmHg were associated with worsening hypoxia, attributed to increased right-to-left shunt due to lower systemic vascular resistance (SVR). Despite an F_iO_2 of 1.0, an intraoperative arterial blood gas showed pH 7.22, identical P_aCO_2 and P_aO_2 of 5.46 kPa, standard bicarbonate 16.9, base excess -10.9 and lactate 4.7 mmol/L. A healthy baby with normal Apgar scores was delivered. Despite prompt administration of a 2 International Unit (IU) bolus and then infusion of oxytocin, intraoperative blood loss exceeded 1 500 ml. The patient received tranexamic acid, two units of packed red blood cell concentrate (PRBC) and four units of reconstituted lyophilised plasma during CS.

Although stable after extubation, she suddenly deteriorated awaiting transfer to the intensive care unit (ICU). Further vaginal blood loss of 2 500 ml was discovered. Repeated episodes of severe hypotension and hypoxaemia required re-intubation, and resuscitation with fluids, blood and inotropes, guided by point-of-care transthoracic echocardiography. No platelets were available, but she received four more units of PRBC, four of fresh frozen plasma, and 100 ml cryoprecipitate. An intrauterine Bakri balloon was placed. After gaining control of the haemorrhage, she was transferred to ICU. Postoperative echocardiography showed intracardiac shunt reversal. She was later referred to an academic centre, where she sadly demised three weeks later due to complications of COVID-19.

Discussion

This case highlights the conflicting requirements for afterload reduction in hypertensive disorders of pregnancy, and the necessity of maintaining SVR to prevent right-to-left shunt in TOF.

TOF accounts for 7–10% of all congenital heart disease (CHD), with a global prevalence of 3.5 per 10 000 live births.[3] It is characterised by an overriding aorta, ventricular septal defect, pulmonary stenosis and RVH.[5] Diagnosis is usually at birth, with corrective surgery recommended within the first year of life. Signs and symptoms are a consequence of the right-to-left shunt and, depending on the severity of disease, include hypoxia, cyanosis, polycythaemia, dyspnoea and palpitations.[6]

Advances in diagnosis and management have led to more patients with CHD reaching reproductive age.[7,8] However, survival to adulthood without surgical correction of TOF remains rare; only 24% of patients reach 10 years of age and 2% their fourth decade.[2,9] Prevalence in resource-constrained settings is likely underestimated due to limited diagnostic capabilities.[1,10] Therefore, patients such as ours may reach reproductive age without diagnosis or corrective surgery, with increased perioperative morbidity.

Pregnancy confers considerable risk to patients with TOF, with maternal and fetal outcomes affected by the disease severity.[7,11] Indicators of increased maternal risk include a high New York Heart Association (NYHA) grade of cardiac failure, SpO_2 < 90%, pulmonary hypertension and a degree of RV dilatation.[12] Haemodynamic changes in pregnancy provoke worsening of the condition. Changes begin in the first trimester and persist until term. Pregnancy is associated with a decrease in SVR, causing compensatory increases in cardiac output and circulatory volume. Decreased SVR also causes increased right-to-left shunt, which decreases maternal SaO_2 and worsens fetal hypoxia.[13,14] Associated increased cardiac output and plasma volume expansion can precipitate right heart failure, which predisposes to arrhythmias. Common medical and obstetric complications include thrombotic phenomena, spontaneous abortion, prematurity, infants small for gestational age, low birth weight, preterm labour and postpartum haemorrhage (PPH).[3,11,15]

There are three widely accepted risk-scoring systems validated for use in pregnancy.[3] Many tools utilise the NYHA classification to predict pregnancy outcomes, but this is problematic, as there is significant overlap in symptoms between cardiovascular disease and normal pregnancy progression.[16,17] Unrepaired CHD is High Risk according to the modified World Health Organization Pregnancy Risk classification (mWHO), requiring management at tertiary care centres.[14]

Pregnancy also has unique cardiovascular diseases: preeclampsia and eclampsia. In South Africa, hypertensive disorders are the second most common cause of maternal death, followed closely by obstetric haemorrhage.[18] The incidence of preeclampsia in Africa is higher than that of the global population,[19-21] but is equally common in patients with CHD.[3,4,11]

Ideal management of uncorrected TOF includes early diagnosis, risk stratification, education, preconception counselling, multidisciplinary specialist collaboration, planned delivery and advanced monitoring such as echocardiography.[3,16] As is common in Africa, this was not achieved for our patient.

Individualised care should be based on disease severity, with consideration of access to care, delivery facility capabilities, specialist availability, and timing and mode of delivery.[14,16] Balancing against maternal condition, allowing pregnancy to progress until 39 weeks' gestation is desirable for optimal fetal outcomes. For pregnancies at or near term, vaginal delivery is preferred, with CS reserved for obstetric indications and patients at high risk for maternal decompensation.

There is no consensus on the preferred anaesthesia technique. The most appropriate strategy should complement the clinician's expertise. Goals centre upon preserving maternal haemodynamics while considering physiological and pharmacological factors influencing the balance between pulmonary- and systemic vascular resistance (PVR and SVR).[13] This includes using analgesia to mitigate the sympathetic response, central and large-bore peripheral intravenous access, and advanced haemodynamic monitoring such as intra-arterial line and point-of-care echocardiography.[16] Physiological stressors associated with increased PVR (such as hypothermia), decreased SVR (dehydration and bleeding), and factors known to trigger infundibular spasm (hypoxia, hypercarbia, acidosis) should be avoided.[6]

The American Heart Association recommends carefully titrated neuraxial anaesthesia, although heterogeneity of maternal CHD requires an individualised approach. Gradual-onset sympathectomy from a graded epidural, accompanied by vasopressor titration to counteract hypotension, is well tolerated in patients with mWHO class 3–4 lesions. Caution should be exercised when administering a test dose in patients who could decompensate from the rapid onset of a spinal anaesthetic or intravascular injection of adrenaline. The test dose may be given as two divided aliquots, using a low-dose local anaesthetic solution while assessing motor and sensory blockade, and avoiding adrenaline. Loss-of-resistance should be performed with saline, due to the risk of paradoxical air embolism.[16]

GA is required where neuraxial anaesthesia is contraindicated; in patients at risk of cardiopulmonary decompensation, or where respiratory symptoms such as dyspnoea or hypoxia preclude the patient lying supine.[16] Disadvantages include the need for airway manipulation in a high-risk population and the physiological consequences of using positive pressure ventilation. Hypertensive disorders of pregnancy heighten the risk of complications during obstetric GA,[22] but general anaesthesia was indicated in our patient due to her recent eclamptic seizure and hypoxaemia.

Drug selection is guided by pharmacodynamics (Table I). For induction, etomidate is favourable, as it does not affect heart rate, PVR or SVR. Ketamine is controversial due to increased PVR, although SVR is maintained. Propofol causes the greatest reduction in SVR and can reduce contractility. Vasopressin may be useful due to its sparing effects on PVR.[23] Volatile agents have a dose-dependent effect on SVR, but minimal effect on PVR. Although nitrous oxide has been shown to increase PVR in

Table I: Summary of the effects of common drugs on pulmonary and systemic vascular resistance

Drug	PVR	SVR
Propofol	Unchanged	Decreased
Ketamine	Increased	Increased
Etomidate	Unchanged	Unchanged
Sevoflurane	Unchanged	Unchanged
Isoflurane	Unchanged	Decreased
Nitrous oxide	Increased	Unchanged
Oxytocin/Carbetocin	Increased	Decreased
Methylergometrine	Increased	Increased
Carboprost	Increased	Increased
Misoprostol (single dose)	Unchanged	Unchanged
Adrenaline	Increased	Increased
Noradrenaline	Increased	Increased
Phenylephrine	Increased	Increased
Vasopressin	Unchanged	Increased

PVR – pulmonary vascular resistance; SVR – systemic vascular resistance

patients with pre-existing pulmonary hypertension, it is unlikely that this results in meaningful alterations in haemodynamics,[24] and higher concentrations of volatile agents required in its absence cause uterine atony.

While the successful management of elective CS in uncorrected TOF using GA or graded epidural techniques is well described, few recommendations exist for emergency procedures with superimposed disease. Where TOF is complicated by preeclampsia, the imperative to control BP conflicts with poor tolerance of decreased SVR. Eclampsia is particularly problematic in TOF, as seizure activity is associated with hypoventilation, hypoxia, hypercarbia and acidosis.[25] Therapy is thus directed at lowering BP and preventing seizures. Importantly, all traditional first-line treatments of severe preeclampsia are associated with decreased SVR, with the exception of cardioselective β-blockers.[26] In our case, the rapid deterioration after antihypertensive therapy may have been due to decreased SVR increasing the shunt fraction. Esmolol is safe in pregnancy, can be rapidly titrated, and may be advantageous for management of severe preeclampsia with TOF.

Since patients with CHD are at increased risk of PPH and tolerate it poorly, prevention and management are critical. Increased risk of atony is attributed to anticoagulant use and restricted uterotonic administration due to cardiovascular concerns.[15] Despite the similar side effect profile and advantages of carbetocin, oxytocin is favoured in cardiac disease as it can be administered as a titrated infusion. Ergometrine causes increased pulmonary artery pressure and is contraindicated in hypertensive disorders. Carboprost also causes increased PVR, as well as possible bronchospasm. Misoprostol is safe, and a single 200 µg sublingual or 400 µg rectal dose does not cause significant side effects.[27] Uterine massage or compression sutures and early placement of a Bakri balloon should be considered.[16]

The use of point-of-care ultrasound results in less time to diagnosis and treatment, and aids in the assessment of volume status, myocardial function and shunt.[16] Lung ultrasound should not be underestimated in establishing the cause of undifferentiated hypoxaemia.

In summary, although guidelines exist for anaesthesia for isolated cardiovascular disorders, information for the combination of TOF and preeclampsia, where there are conflicting therapeutic targets, is limited. Even anaesthetists most experienced in managing maternal risk may require input from experts in cardiac anaesthesiology, ideally guided by invasive monitoring and echocardiography. Recommendations centre on preventing reductions in SVR using careful GA and/or epidural anaesthesia. In our case, neuraxial anaesthesia was contraindicated, but the patient responded well to GA with vasopressors. However, hypovolaemia, anaemia and acidosis caused by PPH precipitated profound instability.

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Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Ethics approval for this case report was provided by the Human Research Ethics Committee of the Faculty of Health Sciences of the University of Cape Town (HREC 732/2023).

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