

Anaesthesia management for cleft lip in a child with unrepaired Tetralogy of Fallot in Malawi: a case report

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Abstract

Background

Children with clefts lips often present with cardiac abnormalities, among them the tetralogy of Fallot. Anaesthesia for patients with unrepaired Tetralogy of Fallot coming for a non-cardiac surgery represents an additional risk of increased perioperative morbidity and mortality.

Case presentation

We present a case of a 8 years old boy with unrepaired Tetralogy of Fallot scheduled for cleft lip repair. The Child was referred to Mercy James Centre for Paediatric Surgery and Intensive Care from an Operation Smile Mission campaign. Anaesthesia consisted of a balanced general anaesthesia combined with regional anaesthesia by an infraorbital nerve block. The child developed hypercyanotic spells postoperatively which were successfully managed with noradrenaline, morphine, fluid, and oxygen therapy.

Conclusion

Children with unrepaired Tetralogy of Fallot coming for non-cardiac surgery have increased risk of complications during anaesthesia. The anaesthesia provider should be aware and ready to manage them promptly.

Key words: Anaesthesia, cleft lip, Tetralogy of Fallot, hypercyanotic spell, Malawi

Introduction

Children scheduled for cleft lip or palate repair may present with congenital abnormalities in 20 to 30% of the cases. Cardiac abnormalities are the most frequent associated comorbidity. Tetralogy of Fallot (TOF), atrial septal defect and ventricular septal defect represent the most common congenital heart diseases seen in this patient population^{1,2}. Proper understanding of the cleft condition, the associated abnormalities and the surgery is the key element to administration of safe anaesthesia for cleft lip and palate and effective management of complications¹⁻⁴.

TOF is one of the commonest cyanotic congenital heart malformations and is characterised by four cardinal features: ventricular septal defect (VSD); right ventricular outflow tract obstruction (RVOTO), which has a fixed and a dynamic component in most patients; an overriding aorta; and RV hypertrophy (RVH). Pathophysiology might range from a "pink" Fallot with almost normal saturations and only marginally impaired physiology to real "blue" babies with very low saturations, severely impaired circulation and frequent hypoxic spells⁵⁻⁸. Patients with TOF can present for extracardiac surgery either before or after correction of the malformation^{5,9}. Anaesthesia for patients with TOF coming for a non-cardiac surgery represents a substantial added risk of increased perioperative morbidity and mortality¹⁰. We report here a case of a child with unrepaired TOF coming

for cleft lip repair.

Case presentation

A 8 years old boy known to have unrepaired TOF was referred for cleft lip repair at Mercy James Centre for Paediatric Surgery and Intensive Care at Queen Elizabeth Central Hospital from an Operation Smile campaign which was held at the Malamulo Hospital in Blantyre, Malawi. He had no subjective complaint and was not on any medication. The history obtained from the mother revealed that he was born at term from a spontaneous vaginal delivery. He had been hospitalized several times for exacerbation of his cardiac condition. He had not had any exacerbation for the last 6 months. He had no history of allergy and had completed his vaccines calendar.

On examination, vital signs were heart rate (HR) 116 beats per minutes, blood pressure (BP) 105/76 mmHg, respiratory rate (RR) 18 cycles/minutes, SpO₂ 80-85% on room air, temperature 36.2°C. The weight was 14 Kg. Conjunctives were pink with a central cyanosis. Chest was clear. The cardiovascular system examination revealed a pansystolic murmur 3/6 on the Levine scale. He had clubbed fingers. The rest of the examination was normal. He had had an echocardiography which confirmed the diagnosis of TOF. On the chest X-ray, the lung fields were normal with a boot shaped heart. Electrocardiogram (ECG) was not done preoperatively. Full blood count revealed white blood cells

6.5 103/ μ L, haemoglobin 12.9g/dL, haematocrit 40.5%, platelets 180 103/ μ L. Covid test was negative. The paediatric cardiologist review favoured the operation. The patient was asked to fast for 6 hours and allowed to drink clear fluid until 1 hour before surgery. Consent was obtained from the mother for both surgery and anaesthesia.

On the day of surgery, standard monitoring included pulse oximeter, ECG, non-invasive blood pressure, capnography, and temperature. After a bolus of Ringer lactate 140ml, anaesthesia was intravenously induced with ketamine 28mg, atracurium 7mg, Fentanyl 20 μ g. The patient was intubated with a 4.5 RAE tube cuffed at 13 cm. Three packs were inserted orally. Patient received Cefazolin 700mg and dexamethasone 2.8 mg. Analgesia consisted of 210 mg paracetamol intravenously (iv) and an infraorbital nerve block with 1ml of bupivacaine 0.5% by intra-oral approach by the anaesthetist. Lidocaine 1% with adrenaline 1:200000 was infiltrated by the surgeon for haemostasis. Anaesthesia maintenance was achieved with isoflurane in oxygen and air at a FiO₂ 45%. Patient was mechanically ventilated with pressure control mode. He received additional 10mg of Ketamine to deepen the anaesthesia in the middle of the procedure. HR varied between 95-100 bpm, BP was in normal range throughout the procedure, SpO₂ improved to 94-96%, and end tidal Carbon dioxide (EtCO₂) was kept around 35 mmHg. Temperature was kept normal using a forced air warming system. Fluid maintenance with plain Ringer's lactate was run at 48ml/hr. The procedure lasted 1h15 from the beginning of the anaesthesia including 45 minutes of surgery. No intraoperative complications were noted. Patient was extubated at the end of the procedure. After 5 minutes in recovery, patient was noted to be agitated and crying. He was tachycardic with HR 175, tachypnoeic with respiratory rate 40, hypoxemic with SpO₂ 50% on Oxygen via a face mask at 5l/min. He was laying in the bed, the legs flexed in a knee-to-chest position. We concluded to a postoperative hypercyanotic episode or "tet spell". The management consisted of administration of a total of 50 μ g of noradrenaline iv slowly and morphine iv 0.5 mg and continued 100% oxygen and fluid at the same maintenance rate. Patient improved and stabilised in the recovery. He was sent to the HDU for observation overnight. Postoperative analgesia consisted of paracetamol iv 210mg every 6 hours. Patient was discharged home on day 2 postoperative with review in 6 months.

Discussion

We have presented the case of a child with an unrepaired TOF undergoing non cardiac surgery. Children with congenital heart disease (CHD) such as TOF undergoing non-cardiac surgery have an increased risk of morbidity and mortality^{11,12}. Anaesthesia for patients with unrepaired TOF is challenging for the anaesthetist¹³. It is advised for them to be operated in a paediatric cardiac centre^{5,11}. In the settings of Malawi with no paediatric cardiac centre, the existing paediatric surgery centre represents adequate settings for anaesthesia for non-cardiac surgery. It has a dedicated paediatric intensive care. The referral from the campaign was thus justified.

Understanding the pathophysiology of the TOF is the key to deliver safe anaesthesia to these patients and manage complications^{4,11}. The anatomy of TOF allows mixing of blood between the pulmonary and systemic circulations which usually occurs at the VSD, with a right-to-left shunt adding deoxygenated blood to the systemic circulation,

causing cyanosis. The compensation includes polycythaemia, hyperventilation, and chronic respiratory alkalosis. The right-to-left shunt is determined by the relative pressure gradient between the right ventricle (RV) and the left ventricle (LV). The amount of pulmonary blood flow (the RV stroke volume) is determined by the degree of the RVOTO. Anaesthetic management of patient with unrepaired TOF must respect an imperative: prevention of worsening right-left shunt⁵⁻⁷. This can be achieved by adequate hydration preoperatively avoiding long fasting times, adequate intraoperative fluid management targeting euvolemia thus preventing exacerbation of dynamic RVOTO from hypovolemia and reflex increasing in HR and contractility, and ventilation strategies which include minimising mean airway pressure to reduce pulmonary vascular resistance (PVR) and avoid mechanical obstruction of pulmonary blood flow and its effects on preload^{5,14}.

Our patient was induced with ketamine and maintained with isoflurane. Analgesia was multimodal. Ketamine is a good agent of induction of patients with TOF to maintain balance between systemic vascular resistance (SVR) and PVR^{5-7,11,14}. Considering an infraorbital nerve block in a multimodal analgesia strategy in these patients present several advantages including reducing inhalational anaesthetic requirement which may lower the SVR, fastening of emergence, reducing the need of opioids in addition to provide adequate perioperative pain control^{1,2}.

The child presented a hypercyanotic episode (tet spells) in recovery which responded well on administration of morphine, noradrenaline, fluid and oxygen. Tet spells may arise under anaesthesia or postoperatively. Tet spells triggers include sympathetic stimulation (pain and anxiety), exercise, breath holding or Valsalva manoeuvre, crying, feeding, and defaecation, vasodilatation and decrease in SVR (e.g. hot baths), hypoxia, hypercarbia, acidosis, induction of and awakening from anaesthesia and sympathomimetic drugs.

Spelling describes a unique acute desaturation and clinical deterioration in a patient with unrepaired TOF due to an acute reduction in pulmonary blood flow caused by a sudden increase in right-to-left shunt. It constitutes an emergency and requires adequate intervention. Treatment goals are to support the airway, breathing and circulation (ABC approach); and to reduce the right-to-left shunt by reducing the dynamic RVOTO, decreasing the PVR, and increasing the SVR to raise the left ventricle end diastolic pressure (LVEDP). Strategies for this management comprise administration of 100% oxygen, intubation, placing the patient in a knee-to-chest position, manual compression of femoral arteries or the abdominal aorta, morphine sulphate (0.05 to 0.1 mg/kg), fluid 15 to 30 mL/kg, phenylephrine at 0.5-2mcg/kg to high doses 5 -10 mcg/kg or noradrenaline. Though phenylephrine is preferred to noradrenaline as a first choice, in the case of limited resources, noradrenaline is a good choice^{6,7,9,10,15}. We successfully reversed tet spells with a relatively high dose of iv slow bolus noradrenaline in 1:1 ratio compared to phenylephrine. There is a need for more investigations to determine efficient and safe dosages of bolus noradrenaline in the management of tet spells in TOF considering the scarcity of its usage in the literature. Administration of sodium bicarbonate (1 to 2 mEq/kg) may be considered to treat the severe metabolic acidosis. Propranolol (0.025-0.1mg/kg) or esmolol (0.1-0.5mg/kg followed by an infusion of 50 to 300mcg/kg per minute)

may reduce infundibular spasm. Extracorporeal membrane oxygenation (ECMO) may be useful in refractory episodes⁵⁻⁸.

In conclusion, children with unrepaired TOF coming for non-cardiac surgery represent a challenge for the anaesthesia provider. Balanced anaesthesia that follows the pathophysiology of the TOF condition avoiding worsening of the right to left shunt is the key management. The anaesthesia provider should be aware that complications mostly hypercyanotic episodes may arise at any time perioperatively and should be ready to manage them promptly.

Ethical Statement

Written informed consent from the mother of the patient and written assent from the child were obtained for publication of this case report.

Conflict of Interest

No conflict of interest was declared by the authors.

Financial Disclosure

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Authorship Contributions

FNBP and BA managed the case. FNBP conceptualised and wrote the first draft of the manuscript. BA, SC, GP reviewed and edited the manuscript. All authors read and approved the final version of the manuscript.

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