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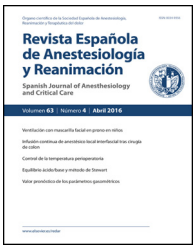

CASE REPORT

A case of severe perioperative hypoxia in uncorrected tetralogy of fallot: Anesthetic management

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KEYWORDS
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Abstract Tetralogy of fallot (TOF) is one of the most common congenital heart disease (CHD) in children. With the development of pediatric surgery and intensive care units, increasing number of grown-up CHD patients are presenting for non-cardiac surgeries. Non-operated TOF patients suffer from chronic hypoxia and decreased pulmonary blood flow resulting in considerable alteration in the physiology. The optimal management of these patients, therefore, require a thorough understanding of the pathophysiology of the uncorrected TOF. We hereby report a case of successful management of a 10-year-old child with an uncorrected TOF posted for tibial external fixation device.

PALABRAS CLAVE
Anestesia;
Cardiopatía congénita;
Tetralogía de Fallot;
Cirugía no cardíaca;
Hipoxia perioperatoria

Caso de hipoxia perioperatoria grave en tetralogía de Fallot no corregida: Tratamiento anestésico

Resumen La tetralogía de Fallot (TF) es una de las cardiopatías congénitas más habituales en niños. Con el desarrollo de la cirugía pediátrica y las unidades de cuidados intensivos cada vez se presentan más casos de pacientes adultos con cardiopatías congénitas para cirugías no cardíacas. Los pacientes con TF no operada padecen hipoxia crónica y un flujo sanguíneo pulmonar reducido, lo que supone una alteración considerable de la fisiología. El manejo óptimo de estos pacientes requiere, por tanto, un profundo conocimiento de la fisiopatología de la TF no corregida. El presente artículo expone el caso de tratamiento exitoso de un paciente de 10 años con TF no corregida intervenido con dispositivo de fijación externa tibial.

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Introduction

Congenital heart diseases (CHD) are one of the most common inborn errors affecting around 0.8% of newborns. With the development of pediatric cardiac surgery and intensive care units, increasing number of grown up CHD patients are presenting for non-cardiac surgeries.[1] Among these, TOF is the most prevalent cyanotic heart disease accounting for 10–14% of all congenital heart defects.[1,2] Non-operated TOF patients suffer from chronic effects of hypoxia and decreased pulmonary blood flow resulting in a considerable alteration in the physiology.[2] Hence, the perioperative management of grown-up CHD patients with uncorrected tetralogy of fallot (TOF) is a challenge for the anesthesiologists and substantial understanding of its pathophysiology is inevitable. We hereby describe the case of a 10-year-old child with uncorrected TOF posted for an external tibial fixation device.

Case summary

A 10-year-old male child, weighing 20 kg, American Society of Anesthesiologist grade II, victim of a road traffic accident presented to emergency department with dyspnea, tachypnea and decreased consciousness. There was no history of loss of consciousness, vomiting or ear/nasal/oral bleed. Initial physical examination revealed a compound fracture tibia with respiratory distress. Vital signs: pulse rate (PR) – 140 beats/min, blood pressure (BP) – 72/54 mmHg, respiratory rate (RR) – 28/min, oxygen saturation (spO_2) – 60%, despite cyanosis and clubbing there was bilateral equal air entry on auscultation of the chest. Neurological assessment showed Glasgow coma score (GCS) – E3V4M5, pupils – normal size and reaction. Immediately 100% oxygen was applied via face mask, and fluid resuscitation with ringer lactate (20 ml/kg) and one unit of packed red blood cells (PRBC) was done. Following which, vital signs increased to BP – 90/60 mmHg and spO_2 – 80%. Laboratory tests showed hemoglobin – 7.6 g/dl, haematocrit – 23%, TLC – 9400/mm^3, DLC – P80L14M6, platelets – 230×10^{-3} mil/mm^3, RBS – 90 mg/dl, blood urea – 18 mg/dl, serum creatinine – 0.8 mg/dl, arterial blood gas analysis: pH – 7.26, paO_2 – 55 mmHg, $paCO_2$ – 42 mmHg, HCO_3 – 18.7 mequiv./L, BE – 5.3, SaO_2 – 65%, lactate – 5.3 mmol/L. Chest radiograph showed right ventricular enlargement, electrocardiogram (ECG) showed right axis deviation while e-FAST (extended focused assessment sonography in trauma) was normal. Cardiology evaluation revealed ejection systolic murmur best heard over the pulmonary area and left sternal border. Echocardiography demonstrated ventricular septal defect (VSD) with right-left shunt, overriding aorta and ejection fraction (EF) of 0.7.

In view of the urgent nature of surgery, patient received a dose of antibiotic prophylaxis and posted for external tibial fixation device under general anesthesia. In the operating room, standard monitors were attached and was premedicated with midazolam 0.03 mg/kg, fentanyl 1 μg/kg, and ondansetron 0.08 mg/kg via intravenous route. After preoxygenation for 3 min, anesthesia was induced with ketamine 2 mg/kg/iv followed by rocuronium 0.9 mg/kg to facilitate endotracheal intubation. The patient was mechanically ventilated (IPPV) with minimal PEEP plus 50% nitrous oxide in oxygen. Anesthesia was maintained with isoflurane – 0.6% and 0.1 mg/kg/h vecuronium infusion. During intraoperative period, the patient has an episode of severe desaturation to 35–40% which was unresponsive despite of 100% oxygen. Therefore, the patient was given a bolus of 20 ml/kg ringer lactate solution and estimated blood loss was replaced by 1 unit of PRBC. Besides, a bolus of phenylephrine 1 μg/kg followed by an infusion of noradrenaline at 0.1 μg/kg/min after right-sided internal jugular cannulation, and esmolol 0.5 mg/kg/iv bolus was given. After that, his spO_2 improved to 78–84%. The remainder of the operative course was uneventful, the patient was extubated in the operative room and was transferred to the intensive care unit for observation overnight. After 24 h, the inotropic support was withdrawn and the child was transferred to the orthopedic ward.

Discussion

TOF, described by Etienne-Louis Arthur Fallot in 1888 is the commonest cause of cyanotic CHD in children. Though its cause is unknown, but many patients are associated with Di George syndrome, cleft lip and palate, hypospadias and skeletal anomalies. A thorough understanding of underlying CHD including anatomy, physiology and identification of risk factors is vital before anesthetizing these children.

The TOF consists of four anatomical components: 1. a large VSD, 2. right ventricular (RV) outflow tract obstruction – RVOTO (commonly infundibular than valvular or supravalvular), 3. RV hypertrophy and 4. overriding aorta. Despite of the presence of all four defects, TOF presents with a spectrum of minimal pulmonary stenosis to virtual pulmonary atresia. The physiology depends on how severe the RVOTO is; favoring the right-to-left shunt and deoxygenated blood to flow into systemic circulation, causing cyanosis and reduced pulmonary blood flows (PBF). When there is a minimal pulmonary obstruction, child may appear pink and becomes blue only during cry, while others may appear blue all the time. The RVOTO can be dynamic, secondary to increased sympathetic tone, these are known as hypercyanotic attacks or ''tet spells''. Examination of the child's cardiorespiratory system will reveal features of poor physiological compensation. The presence of cardiac failure or cyanosis must be searched for to know the high risk for perioperative complications.[3]

The majority of children will have intracardiac repair, consisting of a ventricular septal defect patch and enlargement of RV infundibulum. If left untreated, more than 50% of children with TOF die before their tenth birthday.[4] Those who survive generally present with the problems of chronic hypoxia and cyanosis, polycythemia with thrombotic complications, coagulopathies, congestive heart failure (CHF), perioperative tet spell and kidney abnormalities associated with decreased glomerular filtration rate.[5,6]

Perioperative cyanosis is generally because of imbalance in pulmonary and systemic vascular resistance, favoring decreased pulmonary flow and right-to-left shunting, so precipitously arterial oxygen saturation drops, leading to minimal effect of oxygen therapy[7] (Table 1). Though pulmonary stenosis is the primary factor governing the

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