Case Report

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Anaesthesia for Complex Cyanotic Congenital Cardiac Disease in a Child for Emergency Laparotomy

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Abstract

Transposition of great arteries is a rare and serious congenital cyanotic disorder in which two main arteries leaving the heart are reversed or transposed. Transposition of the great arteries changes the way blood circulates through the body, leaving a shortage of oxygen in blood flowing through the heart to the rest of the body. These patients are at high risk of peri-operative mortality due to ventricular dysfunction, chronic hypoxia, hyper-cyanotic spells, polycythemia and infective endocarditis [1]. Very limited studies are available regarding anaesthetic consideration in patients with uncorrected D-TGA with other congenital cardiac defects like ventricular septal defect (VSD) and pulmonic stenosis. This case report highlights the anaesthetic challenges in an uncorrected transposition of great arteries along with other congenital cardiac defects for an emergency non-cardiac surgery in a critically ill pediatric patient.

Keywords: Transposition of Great Arteries (TGA); Ventricular Septal Defect (VSD); Pulmonic Stenosis; Cyanosis; Hypoxia; Laparotomy; Pediatric Anaesthesia.

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Introduction

D-Transposition is the commonest diagnosed cyanotic congenital heart disease. We hereby present a case of uncorrected transposition of great arteries (D-TGA) with severe infundibular pulmonary stenosis (PS) with bidirectional VSD, posted for emergency exploratory laparotomy for suspected sub-acute intestinal obstruction (acute abdomen).

Case Report

A 12-year old male patient, weighing 20 kg, presented to the pediatric emergency with complaint of fever, abdominal pain with distension and malena for three days. Thr child had history of cyanosis on crying and during any exertional activity. Preoperative examination revealed a body temperature of 101degrees Fahrenheit, heart rate 130 beats per minute (bpm), respiratory rate 24/min, SpO_2 57% (on the face mask with FiO_2 of 0.6), severe clubbing of fingers grade 3, and central cyanosis. On chest auscultation, breath sounds were decreased at the bases of lungs bilaterally, and a pan-systolic murmur of grade 3 was heard. Preoperative investigation showed haemoglobin 17.2 gm/dl, total leucocyte count 16,700/cu-mm, platelet count 1.8lakh/cu-mm, with normal kidney and liver function tests. X-ray of the abdomen

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showed multiple air-fluid levels. In echocardiography, TGA with severe infundibular pulmonary stenosis with peak pulmonary gradient of 80mm of Hg, severe tricuspid regurgitation and non-routable large mid-muscular bidirectional VSD was found. Electrocardiogram showed right axis deviation and right ventricular hypertrophy. In arterial blood gas analysis (pre-operative ABG), pH 7.498, pCO_2- 22.9, pO_2 – 38.3, HCO_3- 17.4 was found, indicating metabolic acidosis with respiratory alkalosis.

Emergency exploratory laparotomy was planned under general anaesthesia. A pediatric cardiologist opinion was sought and infective endocarditis prophylaxis was administered(the child was on vancomycin 360 mg QID and amikacin 360 mg OD).

A patient was wheeled into the operation theatre with oxygen by face mask. All standard ASA essential monitors were attached (ECG, Pulse, SpO_2, $EtCO_2$, BP, and Temperature). Blood pressure of 100/60 mm of Hg was recorded; SpO_2 of 57% on face mask oxygen was noted. Intravenous paracetamol 15mg/kg was given, as the patient was febrile. In view of the acute abdomen with sub-acute intestinal obstruction, a modified rapid sequence induction with cricoid pressure was planned after adequate pre-oxygenation. Premedication was done with injection midazolam 0.5mg IV, injection fentanyl 2microgram/kg i.v. was given in titrated doses slowly. Injection etomidate 0.25mg/kg i.v. and rocuronium bromide 0.9mg/kg i.v. were administered. Patients' trachea was intubated with a cuffed endotracheal tube of size 5mm internal diameter. Cricoid pressure was applied as soon as injection etomidate i.v. was given and the cricoid pressure was released when a patient's trachea was intubated with a cuffed ETT size 5mm and the cuff was inflated and bilateral air entry was confirmed.

Thereafter, an arterial line was secured in the left radial artery and the central line was placed right internal jugular vein, using Seldinger technique. Invasive, beat-to-beat blood pressure monitoring and central venous pressure monitoring was done. Urinary bladder catheterization was done for hourly urine output monitoring. Nitrous oxide was not used, instead, the air was used as carrier gas. During surgery, adequate hydration was maintained with (room temperature) Plasmalyte crystalloid solution to maintain a CVP of 8 to 10 cm H_2O. Anaesthesia was maintained with isoflurane (MAC of 1 -1.2) with oxygen along with intravenous, titratedPropofol infusion was started and on volume controlled ventilation to maintain an end-tidal CO_2 between 34-38 was achieved. Cold sponging of fore-head and axilla was done to reduce body temperature. Injection phenylephrine was prepared and loaded in a syringe so that it can be given promptly whenever there is a need to increase the systemic vascular resistance in the event of a hyper-cyanotic spell. The patient was hemodynamically stable for the entire duration of surgery, lasting 2 hrs. Urine output at the end of surgery was 50 ml. Intraoperatively, a stricture was found in the ileum for which ileostomy was done by the surgical team. Injection tramadol was given for postoperative pain relief and the abdominal incision was infiltrated with 10 ml of 0.25% bupivacaine. The patient was shifted to pediatric ICU in intubated condition and after 4 hours, he was extubated, after evaluating the ABG and ventilatory parameters. After extubation, the patient maintained SpO_2 of 70-75%. Fever subsided after 24 hours. The child was later referred to the cardiac surgery unit for definitive correction of the congenital cardiac anomaly.

Discussion

Transposition of great arteries is a cyanotic congenital heart disease where both the arteries arising from the ventricles are reversed, meaning the aorta arises from the right ventricle and the pulmonary trunk arises from the left ventricle. There is a parallel circulation in which blood from the right atrium goes to the right ventricle and then to the aorta, blood from the left atrium goes to the left ventricle and then to the pulmonary artery. Both the pulmonary and systemic circulations are separate and parallel. Survival1 depends on the presence of atrial, ventricular or aortopulmonary communications. These communications allow mixing of oxygenated and deoxygenated blood. Oxygen saturation depends on the inter-circulatory mixing of blood through the shunt. The pulmonary vascular resistance should be maintained so that pulmonary blood flow is maintained and oxygenation of blood takes place so that the SpO_2 is maintained. If the pulmonary vascular resistance is increased, then blood flow to the lungs decreases and deoxygenated blood increases in circulation, causing a fall in SpO_2.

Our patient had D-TGA with VSD with infundibular pulmonary stenosis, this warrants avoidance of peri-operative dehydration, maintenance of systemic vascular resistance, pulmonary vascular resistance and minimizing an increase in oxygen consumption, all of which are central to a successful outcome [2]. These patients have polycythemia as a compensatory response to

improve oxygen saturation at the expense of hyperviscosity. Hence, they are at increased risk of thrombosis and stroke. There is an increased risk of perioperative bleeding due to multiple coagulation factor deficiencies, necessitating coagulation studies. Chronic hypoxia has adverse effects on the heart leading to the reduction in ventricular diastolic compliance and myocardial reserve. Chronic renal hypoxia leads to an increase in serum urea and creatinine levels [3]. A preoperative echocardiography and chest radiography should be done. Factors increasing sympathetic drive should be avoided such as light anaesthesia, pain, acidaemia, hypoxia, hypercarbia, and hypothermia [4,5]. Fentanyl, Sevoflurane and Etomidate were chosen because they have minimal effect on systemic vascular resistance and pulmonary vascular resistance [6]. N_2O was avoided because it increases pulmonary vascular resistance and can also expand the air bubbles which are accidentally entrained from the surgical site or intravenous line leading to paradoxical air embolism. Utmost care should be taken to avoid air bubbles going in the circulation, either through the intravenous tubing or through the surgical site [7]. Care must be taken to adequately hydrate the patient in the peri-operative period to avoid hyper-viscosity. There is a risk of peri-operative "tet spells" or hyper-cyanotic episodes in these patients, due to sudden decreases in pulmonary blood flow, which may be difficult to manage. Injection phenylephrine hydrochloride was kept ready, as this drug increases the systemic vascular resistance and increases the pulmonary blood flow and in turn, increases the oxygen saturation.

Conclusions

Complex and mixed congenital cardiac lesions can be quite challenging to manage, especially in an emergency setting, where there may be limited time available for evaluation and optimization. This case was unique as the child had several, serious uncorrected cardiac lesions in the presence of fever, clubbing and central cyanosis. Prevention, prompt recognition and treatment of 'tet' spells are of paramount priority.

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Corporate (collective) author

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