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Case Report

Anaesthesia Section

Unravelling the Convoluted Story of Perioperative Care in Three-year-old Child with Tetralogy of Fallot Undergoing Repair Surgery

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ABSTRACT

Tetralogy of Fallot (TOF), one of the most prevalent cyanotic congenital heart diseases in children. Single step corrective surgery, early on in life, provides a fair expectation of favourable outcome in these individuals. However, complex, and skilled anaesthetic management is required by experienced team of healthcare providers. Preoperative surgical preparation, intraoperative key anaesthesia principles and postoperative care Intensive Care Unit (ICU) are all perioperative considerations in these patients. The present article reports challenges faced in the perioperative anaesthesic management of a 3-year-old male child having uncorrected TOF, who underwent Waterston shunt and later Intracardiac Repair (ICR). He presented with postoperative complication like cyanotic spell, gastrointenstinal bleeding, sepsis; but later, recovered successfully.

Keywords: Anaesthesia, Congenital cyanotic heart disease, Cyanotic spell, Intracardiac repair, Waterston shunt

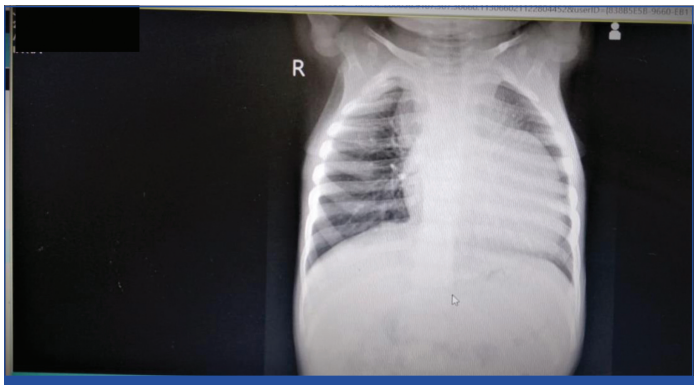
CASE REPORT

A 3-year-old male child having congenital heart disease of tetralogy of fallot (diagnosed at birth) was posted for one step corrective surgery and preanaesthetic check-up was requested. He had chief complaints of bluish discolouration of lips and nail beds, breathlessness, and intermittent loss of consciousness after crying or excessive playing. The child became limp off and on after any rigorous activity. These episodes lasted for 3-5 minutes and occurred 5-6 times in a month. Increased frequency of these episodes was observed since last 3-4 months, which subsided after maintaining a knee-chest position or on reassurance by the mother. The child was born vaginally, at term gestation and was admitted in Neonatal Intensive Care Unit (NICU) for difficulty in breathing and had a birth weight of 1.5 kg. There was delay in achieving developmental milestones with a reported ability to sit at age of 1.5 years. The child is not able to stand and/or walk at the age of three years and can speak only in monosyllables. He was receiving tablet propranolol 5 mg twice daily since birth.

The weight and height of the child was 6.1 kg and 77 cm, respectively, with a body surface area of 0.36 m^2. He was afebrile, with a pulse of 102 bpm (beats per minute), respiratory rate of 26/min. There was no pallor, icterus, lymphadenopathy, or oedema on the feet. Jugular venous pressure was normal. Cyanosis was present on lips, tongue, fingers and toes, there was grade 3 clubbing and oxygen saturation was 40% on room air. On inspection of the precordium, there was visible apical impulse, and thrill was palpable at second intercostal space along the sternal border. Parasternal heave was present. S1 and S2 heart sound were heard normally. Pansystolic murmur was present on lateral border of sternum in 2nd intercostal space. There was no chest wall deformity and accessory muscle for respiration were not used.

The ejection fraction was 65% on 2D echocardiography examination. There was a large, bidirectional, Ventricular Septal Defect (VSD) of 12 mm size. The pulmonary arteries were grossly hypoplastic with a severe infundibular pulmonary stenosis with pressure gradient of 68 mmHg. The pulmonary valve annulus was 6 mm. The superior vena cava was grossly dilated and Tetralogy of Fallot (TOF) was seen. Boot-shaped heart was seen on chest x-ray [Table/Fig-1].

As a part of preoperative preparation, the baby was kept nil by mouth for 4 hours, intravenous (i.v.) cannula was secured, and i.v. fluids



[Table/Fig-1]: Chest x-ray showing boot-shaped heart.

Dextrose Normal Saline (DNS) (2 mL/kg) was started to maintain adequate hydration. Adequate blood and blood products were cross matched and kept ready. Informed consent was obtained from patient's father after counselling, about risk of anaesthesia, postoperative complications of surgery and chance of prolonged Intensive Care Unit (ICU) stay. Precaution was taken to avoid hypothermia. Initially, Blalock-Taussig (BT) shunt was planned, but because of unfavourable anatomy, the surgical team had opted to perform Waterston's shunt. During preinduction of anaesthesia, the child went into a cyanotic spell, blood pressure dropped to 60 mmHg and SpO_2 (oxygen saturation) fell to 10%. To increase the systemic vascular resistance, the anaesthetic team tried to calm the child and make him comfortable in knee-chest position and 100% O_2 was given. A fluid bolus was given, and acidosis was corrected after injecting sodium bicarbonate in bolus dose as per body weight. Injection phenylephrine was given as per requirement to treat hypotension. Patient responded to the given treatment and became vitally stable.

Anaesthesia was induced with inj. midazolam (0.05 mg/kg), inj. fentanyl (1 mcg/kg), inj. ketamine (2 mg/kg), inj. vecuronium (0.1 mg/kg) as per calculated dose per kg body weight in i.v. route and the child was intubated with No.5 uncuffed endotracheal tube and maintained on oxygen and air. Injection vecuronium was used for topping up of anaesthesia. Central Venous Pressure (CVP), Invasive Blood Pressure, temperature was monitored as per standard American Heart Association (AHA) monitoring. Normothermia and normocapnia was maintained. Intravascular fluid volume with attention to blood sugar monitoring was done.

Meticulous care was taken to avoid air bubbles entering the tube used to deliver intravenous fluid. Waterston Shunt was done using pericardial tube. Waterson shunt is side-side anastomosis of right pulmonary artery to ascending aorta. Anastomosis of Right Pulmonary Artery (RPA) to aorta was done. Postoperatively, the patient was shifted to intensive care unit on ventilatory and ionotropic (noradrenaline and adrenaline) support. In postoperative period, good analgesia was achieved with Inj. dexmedetomidine infusion and Inj. paracetamol as per body weight. The child was extubated on Postoperative Day (POD) 4, was vitally stable and maintained saturation of 80% on two litres of oxygen and was shifted to regular ward on POD 8.

On POD 10, the child suddenly desaturated and SpO_2 fell to 20%. He was shifted to ICU and was diagnosed to have a cyanotic spell and severe metabolic acidosis as per Arterial Blood Gas (ABG) report {pH-6.8, (partial pressure of carbon dioxide) pCO_2-48, pO_2-25, (partial pressure of oxygen) lactate-15) [Table/Fig-2]. He was managed conservatively (knee-chest position). On Transthoracic Echocardiography (TTE), good flow across the shunt was noted. He was shifted to ICU and intubated. Intravenous heparin 500 IU was given followed by 100 IU per hour, intravenous infusion. In view, of the deteriorating condition and haemodynamics, the patient was taken up for Intracardiac Repair (ICR) under high risk.

Parameters	Results
1st ABG report during cyanotic spell	
pH	6.80
pCO_2	48 mmHg
pO_2	25 mmHg
Lactate	15 mmol/L
Bicarbonate	Not recordable
Base deficit	Notable record
SpO_2	Not recordable
Haemoglobin	10.2 g/dL
2nd ABG on 10 lit O_2	
pH	7.28
pCO_2	38 mmHg
pO_2	10 mmHg
Lactate	15 mmol/L
Bicarbonate	17.9 mmol/L
Base deficit	8.8 mmol/L
SpO_2	7%
Haemoglobin	10.2 g/dL

[Table/Fig-2]: The Arterial Blood Gas (ABG) reports.

Intracardiac repair was done under anaesthesia and the child was managed postoperatively in ICU, ventilated mechanically, and kept on ionotropic support. He was extubated on POD 2 of ICR. On POD 3 of ICR, there was sudden fall in saturation, and we suspected aspiration due to Gastrointestinal (GI) bleed, secondary to thrombocytopaenia as platelet was 32000 cells/μL. Ryle's tube bleed and melena was present. Cold saline irrigation was done. The child was again intubated and shifted to mechanical ventilation in view of drop in saturation and suspected aspiration. Dexamethasone and heparin were stopped to avoid steroid or heparin induced thrombocytopenia. Multiple Packed Red Cells (PRC), Fresh Frozen Plasma (FFP) and platelets were transfused as per body weight to maintain haemoglobin and platelet within normal range.

On post operative day 4 of ICR, the child developed with sepsis i.e high grade fever, tachycardia, hypotension, high total leucocyte counts (22600 cumm) and low platelet count (32000 cells/μL); so blood culture, Endotracheal Tube (ET) culture and urine culture were immediately sent. Blood investigations- Complete Blood Count (CBC), C-Reactive Protein (CRP), serum lactate and chest X-ray was done. It was found that Total Leucocyte Count (TLC) (18000 cumm) and CRP (47.28 mg/L) values were higher than normal values. Platelet count was low (28,000 cells/μL). In x-ray, there was bilateral haziness present in lower lobes. The antibiotics were escalated and Inj. colistin 90,000 IU TDS and Inj. meropenem 200 mg TDS were started, within the 1st hour of recognising sepsis. Fluid challenge was given and multiple inotropes started in view of persistent hypotension. Chest physiotherapy and nebulisation were given to manage the secretion overload.

In postoperative period, anaesthesia team maintained the child, with adequate filling and started phosphodiesterase inhibitors (sildenafil) and ionodilators (milrinone, dobutamine). The child responded well to the treatment and was extubated on POD 6 of ICR and managed conservatively (Inj. digoxin 0.06 mg i.v. once a day, Inj. furosemide 3 mg i.v., Inj. pantoprazole). Then he was shifted to step-down ward and discharged from hospital on POD 10 in stable condition. The 2D echocardiography on discharge showed signs of TOF repair. The Ventricular Septal Defect (VSD) patch was in-situ and there was no residual VSD. There was mild residual Pulmonary Stenosis (PS) with pressure gradient of 34 mmHg. Free pulmonary regurgitation was present with normal biventricular function.

DISCUSSION

Tetralogy of Fallot (TOF) is one of the most prevalent cyanotic congenital heart disorders [1]. Four main characteristics of TOF include [2]:

1. Ventricular Septal Defect (VSD)
2. Right Ventricular Outflow Tract Blockage (RVOTO), which is often dynamic
3. Over-riding aorta and
4. Right Ventricular Hypertrophy (RVH).

The presentation and prognosis of this disease are determined by the degree of RVOTO, relative pressures in the right and left ventricles and the proportion of the aorta over-riding the VSD [2]. Several perioperative risk factors need to be considered for patients with TOF, whose presentations might vary in complexity. The first factor is the severity of disease, which is assessed by the heart's preoperative oxygenation and function.

In most circumstances, corrective surgery is performed in a single phase, and if done, quickly enough, positive results can be anticipated. Anaesthetists may encounter this condition before to or during repair, thus, they should be familiar with its relevant anatomy, physiology, and emergency care. Its presence increases the perioperative risk and mortality. Preoperative planning for surgery, managing the anaesthesia during surgery, and managing common postoperative problems in the critical care unit are all perioperative issues for these patients.

Anaesthesiologists face various challenges while managing a child undergoing repair surgery for uncorrected TOF. They must have a full understanding of the pathophysiology, events, and effects of drugs that can change the degree of right to left shunting. Maintaining normovolemia, minimising hypoxia, and avoiding changes in Systemic Vascular Resistance (SVR) and Peak Vascular Resistance should all be goals of. In the present case, during preinduction of anaesthesia, the child went into a cyanotic spell, blood pressure dropped to 60 mmHg and SpO_2 fell to 10%. Cyanotic episodes are typically brought on by either a drop in SVR or a spasm of the heart muscle in the vicinity of the RVOT as a result of sympathetic activation (infundibular spasm) [3]. Adrenergic agonists like phenylephrine or norepinephrine and intravenous (i.v.) fluids are used to treat any drop in SVR, whereas beta blockers like propranolol or esmolol are used to treat infundibular spasm.

In present case, on postoperative day 10, the child suddenly desaturated due to cyanotic spell and severe metabolic acidosis. He was managed medically and heparinised, however due to

deteriorating condition and haemodynamics, he was taken up for ICR. As there was unfavourable anatomy for BT shunt i.e evidence of large 12 mm subaortic VSD, grossly hypoplastic Pulmonary Arteries (PA) (MPA-5 mm, LPA/RPA -4.2 MM), and severe infundibular pulmonary stenosis with pressure gradient 68 mmHg, surgeons decided to perform Waterston shunt. However, the child could not maintain oxygen saturation and had to be taken for intracardiac surgery. The Waterston shunt is preferred by for palliative treatment of cyanotic congenital heart disease with decreased pulmonary blood flow [4], though intracardiac repair surgery is definitive treatment.

However, on postoperative day 4 of ICR, the child developed sepsis i.e high grade fever, tachycardia, hypotension, high total leucocyte counts (22600 cumm) and low platelet counts (32000 cells/μL). Yaroustovsky M et al., suggested that one of the main issues with paediatric intensive care to this day is sepsis [5]. Infants with chronic illnesses and congenital problems, as well as, low birth weight newborns, are particularly vulnerable. It is estimated that 15-30% of paediatric heart surgery patients would experience infection problems [5]. As a result, early sepsis treatment reduces the length of ICU stays and increases the survival rate of children after cardiac surgery. According to Oliveira DC et al., sepsis after cardiac surgery is a rare occurrence, with a reported prevalence of 0.39% to 2.5%. Patients who develop severe sepsis following heart surgery, have a significant death rate, ranging from 65 to 79% [6]. They also require prolong mechanical ventilation, as well as, intensive care and hospital stay. Early detection, aggressive fluid resuscitation, appropriate antibiotic therapy, source control and organ support should be done for sepsis management [7].

After intracardiac surgery, Gastrointestinal (GI) bleeding, is a serious and often fatal complication [8]. Elgharably H et al., reported Gastrointestinal Complications (GICs) in 1037 patients (3.5%) out of 29,909 cardiac surgical procedures, with overall in-hospital mortality of 14% compared to 1.6% in those without GICs. Mesenteric ischaemia, Hepatopancreatobiliary (HPB) dysfunction, and gastrointestinal haemorrhage were the most fatal GICs [9]. Welsh GF et al., reported that within 30 days after open-heart surgery, 16 (0.22%) of the 7,333 patients developed significant gastrointestinal bleeding [8]. Early recognition and aggressive treatment of GI bleed are necessary to improve the postoperative outcomes [9].

The present case report, mainly focusses on expert anaesthesia management and excellent nursing care, which helped in managing the complicated case successfully. The patient response was positive to conservative management. The patient was recovered satisfactorily and discharged on 10th postoperative day of ICR.

CONCLUSION(S)

Children with tetralogy of fallot are at high risk for anaesthesia management. Anaesthesiologists have to deal with TOF patient in cardiac catheterisation laboratory, TOF with cerebral abscess posted for Magnetic Resonance Imaging (MRI), neurosurgery, TOF with non cardiac surgery, TOF posted for BT shunt/ICR. Management starts with proper preanaesthesia check-up, preoperative advice and preparation for surgery. Postoperative monitoring and care is equally important as intraoperative care management. Cyanotic spells may prove life-threatening and deserve prompt treatment. Sepsis is another life-threatening condition, which may adversely affect the outcome of children affected by RVOTO and needs prompt recognition and aggressive management to save life. Multidisciplinary approaches involving anaesthesiologists, surgeons, paediatric cardiologists and neonatologist is essential in managing these patients.

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