Case Report

Anaesthesia concerns and perioperative management in a child with DiGeorge syndrome with corrected tetralogy of Fallot with pulmonary atresia posted for laparoscopic orchidopexy: Case report

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ABSTRACT

DiGeorge syndrome is afflicted with multiple congenital anomalies such as conotruncal and craniofacial anomaly, immune system dysfunction and hypoplasia/aplasia of parathyroid glands. Laparoscopy is a preferred surgical approach over open orchidopexy due to better visualisation of impalpable testis avoiding long incision, minimal tissue damage and a faster recovery. We report a case of DiGeorge syndrome with corrected tetralogy of Fallot with pulmonary atresia in a 1-year-old male child posted for laparoscopic orchidopexy. The anaesthesiologists face unique challenges due to the multisystem involvement and the effects of laparoscopic surgery on multiple organs. Thorough understanding of DiGeorge syndrome is essential for a good perioperative outcome.

Key words: Corrected tetralogy of Fallot with pulmonary atresia, DiGeorge syndrome, laparoscopic orchidopexy

INTRODUCTION

DiGeorge syndrome is the most common chromosomal microdeletion disorder affecting approximately one in 4000 live births.[1,2] Clinical manifestations include conotruncal anomalies including tetralogy of Fallot (ToF) in 10-15% patient,[3] craniofacial dysmorphism, thymus and parathyroid hypoplasia or aplasia. They pose several anaesthetic challenges such as pre-existing cardiac lesion or corrected cardiac lesion with residual disease, anticipated difficult airway, hypocalcaemia related haemodynamic instability, seizures during recovery, need for irradiated blood for blood transfusion and risk of perioperative infections.[4] We discuss the anaesthetic challenges in a 1-year-old male child suffering from DiGeorge syndrome with corrected ToF with pulmonary atresia (PA) posted for laparoscopic orchidopexy.

CASE HISTORY

A 1-year-old male child, known case of DiGeorge syndrome, was scheduled for laparoscopic orchidopexy for right undescended testis. The child was diagnosed with tetralogy of Fallot with long segment pulmonary atresia at birth. He underwent an emergency Blalock

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Taussig (BT) shunt on day 5. Consequently the child underwent early intracardiac repair with right ventricle to pulmonary artery 14 mm conduit with ventricular septal defect (VSD) patch and BT shunt take down and patent ductus arteriosus (PDA) ligation with small patent foramen ovale (PFO)/atrial septal defect (ASD) of 7 mm at 9 months of age. After the intracardiac repair, he was started on oral Sildenafil 1 mg/kg thrice a day and Tab. Bosentan 6.25 mg twice a day for residual pulmonary hypertension.

Operative record showed that the child had absent thymus and hypocalcaemia was treated in the peri-operative period. Irradiated blood was used in view of suspected DiGeorge syndrome. Diagnosis was later confirmed with Fluorescence *in situ* hybridization (FISH) assay. He had delayed milestones of development and growth retardation.

On physical examination, he weighed 8 kg and had atypical facial features like narrow up slanting of palpebral fissures, malar flatness, low set ears and broad nasal tip. His saturation on room air was 85-90%. On systemic examination, respiratory system was normal. Airway assessment revealed adequate mouth opening and normal neck movements but retrognathia was noted. Preoperative investigations included haemoglobin content 13.7 gm/dl, prothrombin time 13.4 seconds and international normalised ratio (INR) 1.15. Serum calcium levels were 10.10 mg/dl. Echocardiography suggestive of intracardiac repair with right ventricle to pulmonary artery conduit and ligation of PDA, PFO/ASD (7 mm) with bidirectional shunt, right ventricle systolic pressure of 55 mmHg, impaired right ventricle function, tricuspid annular plane systolic excursion (TAPSE) 10 mm, and estimated mean main pulmonary artery (MPA) pressure of 30 mmHg.

After informed consent, the child was shifted to operating room and monitors for electrocardiogram, non-invasive blood pressures, pulse oximetry, and end tidal carbon dioxide were attached. A 24G intravenous access was secured on right forearm in the operation theatre under inhalational anaesthesia with sevoflurane. The child was premedicated with injection glycopyrrolate 5 mcg/kg, ondansetron 0.1 mg/kg and midazolam 0.03 mg/kg intravenously (IV). Antibiotic prophylaxis given with injection cefuroxime 250 mg i.v before start of surgery.

Anaesthesia was induced with intravenous fentanyl 2 mcg/kg and sevoflurane and air-oxygen mixture. Muscle relaxation was achieved with intravenous atracurium 0.5 mg/kg. Intubation was done with 4.0 mm cuffed endotracheal tube (ETT) and was fixed after ensuring adequate bilateral air entry. Anaesthesia was maintained with sevoflurane 2% in oxygen: air ratio (70%:30%) along with intermittent doses of atracurium. Ventilation was with pressure control ventilation- volume guaranteed (PCV-VG) with positive end-expiratory pressure (PEEP) 5 cm H2O during pneumoperitoneum. Minute ventilation was adjusted by modifying tidal volume (10-12 ml/kg) and respiratory rate (25-30/min) to maintain EtCO2 between 25-30 mm Hg and higher Fi02 of 70%. The intraoperative monitoring – ECG (within normal range with no ST/T changes or ectopics), peak airway pressure (within the range of 18-24 mmHg), non-invasive arterial pressure (90/50-80/40 mmHg), pulse oximetry (98-100% on Fio2-70%), $EtCO_2$ (25-30 mmHg), and fluid balance (using lactated Ringer and 1.25% dextrose) were essentially within normal limits. Laparoscopy revealed low intra-abdominal testis adjacent to deep inguinal ring.

Intravenous calcium gluconate 0.5 ml/kg was given slowly under cardiac monitoring. Child was haemodynamically stable throughout the surgery. Paracetamol suppository 175 mg was inserted rectally for postoperative analgesia. Procedure lasted for 1.5 hours and the patient was extubated fully awake in the operating room. His postoperative recovery was uneventful and was discharged on the third post-operative day.

DISCUSSION

Pre-anaesthetic assessment in DiGeorge syndrome is vital to assess cardiac anomaly and previous corrective surgeries with residual disease, facial dysmorphism leading to anticipated difficult airway, defects in thymus with low lymphocyte count increasing risk of perioperative infections, use of irradiated blood to prevent Graft versus Host Disease (GvHD) and low ionic calcium levels to prevent seizures related hypocalcaemia and perioperative risk of haemodynamic instability.

Laparoscopic orchidopexy is the surest way to locate the site of impalpable testis with lesser tissue damage and minimal need for postoperative analgesia, reduction of postoperative respiratory and wound complications and hastens recovery.[5,6] But laparoscopic surgeries itself pose several anaesthetic challenges such as

increase in Intra-Abdominal Pressure (IAP) causing basal atelectasis and ventilation perfusion mismatch. CO2 insufflation leads to increase in EtCO2 which further causes increase in systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR).[7] Factors that may further increase pulmonary hypertension or decrease ventricular function such as relative hypovolaemia, depressant anaesthetic agents, hypoxia, acidosis, hypothermia, arrhythmias, stress, and pain should be avoided.[8]

Our main goal was to prevent further increase in pulmonary vascular resistance and maintain systemic vascular resistance to avoid decrease in coronary perfusion and right ventricular perfusion pressures and right to left shunting through PFO. We did an inhalational induction with sevoflurane avoiding hypotension and tachycardia and maintained the child on higher fiO2. Adverse effects of pneumoperitoneum were counteracted with lower intraabdominal pressures (6-10 mmHg), hyperventilation (respiratory rate of 25-30/minute) to maintain normocapnia and PEEP (5 cm H20) to prevent hypoxaemia.[9] Peri-operatively pulmonary vasodilator therapy with oral sildenafil and bosentan was continued to maintain the PVR. We maintained adequate analgesia with paracetamol suppository.

Though we did not encounter difficult intubation, difficult airway cart was kept ready and patient was intubated in the head up position after adequate preoxygenation. Hypocalcaemia secondary to hyperventilation and alkalosis was prevented with intraoperative intravenous calcium supplementation 0.5 ml/kg. Antibiotic prophylaxis with careful aseptic technique was used to prevent the risk of perioperative infections and infective endocarditis in a corrected cardiac lesion. Irradiated blood was kept as a standby.[10]

To conclude, sound knowledge and thorough understanding of various anatomic anomalies and pathophysiologic considerations are required to prevent any perioperative complications.

Declaration of parental consent

The authors certify that they have obtained all appropriate parent consent forms. In the form, the parent(s) has/have given his/her/their consent for his/her/their child's images and other clinical information to be reported in the journal. The parents understand that their child's names and initials will not be published and due efforts will be made to conceal their child's identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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