Anesthetic dilemma in planning bilateral cataract surgery for an infant associated with congenital cardiac anomaly

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In a patient with tetralogy of Fallot (TOF) and pulmonary atresia, treating the cardiac problem or the associated congenital illness is always a challenge. We describe the challenges and successful initial management of bilateral cataract to prevent visual loss in an infant with TOF with pulmonary atresia.

Key words: Anesthesia, tetralogy of Fallot, neonatal cataract

Tetralogy of Fallot (TOF) with pulmonary atresia present unique challenges to the anesthetist in terms of maintenance of oxygenation and systemic blood flow. Patients can also have other associated anomalies, which require surgical intervention before correction of the cardiac defect. The successful management of intra-operative period and postoperative complication in a child with TOF with pulmonary atresia and associated bilateral congenital cataract posted for cataract surgery is discussed.

Case Report

A 5-month-old male child weighing 4 kg, having bilateral microcornea and total cataract, presenting with nystagmus, was planned for lens aspiration and intraocular lens implantation. Parents mentioned that the child had a history of recurrent cough; however, there was no history of hurried breathing, feeding difficulty, cyanosis, or seizures. On examination, the child was found to have cyanosis, clubbing, grade four systolic murmurs in left second intercostal space with room air saturation of 60–70% only. In addition to complete hemogram and serum electrolytes, which were within normal limits an echo-cardiogram was done which revealed TOF with pulmonary atresia, decreased pulmonary blood flow, large ventricular septal defect (VSD), large patent ductus

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arteriosus (PDA), and normal biventricular function. As the patient had presented late, after consultation with the Cardiologist the correction of the cataract was deemed more urgent than immediate correction of TOF defect.

On the day of surgery, the child was prepared as per American Society of Anesthesiologists (ASA) fasting guidelines. As per the advice of the cardiologist, the patient received antibiotic prophylaxis with intravenous (IV) cefazolin 200 mg 60 min before the procedure. Prostaglandin (PG) E1 infusion was kept ready to maintain the patency of PDA, which could be guided by a fall in hemoglobin saturation below 45%. Electrocardiography (ECG), noninvasive blood pressure, O₂ saturation, and temperature monitoring were done as per standard ASA guidelines. EMLA cream was applied on the dorsum of left hand for securing IV access to prevent pain leading to crying spell, which could have led to de-saturation. The patient was induced with IV ketamine (8 mg) and mask ventilation with 100% O₂ was done. Proseal laryngeal mask airway (LMA) was used to secure the airway as oxygenation and ventilation are as effective as endotracheal intubation with far less hemodynamic changes. Maintenance was done with O_{γ} , air, sevoflurane, and IV fentanyl (8 ug) and vecuronium (0.4 mg) was used for its cardiostability. Fluid deficit was replaced with Ringer Lactate solution. Patient's hemodynamics were stable and saturation was maintained around 60-65% throughout the intraoperative period. Surgery lasted for 30 min and LMA was removed after giving reversal. The child received supplemental O₂ and monitored (SpO₂ and ECG) in the postanesthesia care unit for 4 h in an incubator. Thereafter the child was shifted to the ward. The child received paracetamol rectal suppository for pain relief. On the first postoperative day, child developed breathing difficulty, fever and increasing cyanosis for which he was shifted to the cardiac Intensive Care Unit (ICU) and after a week Blalock-Taussig shunt procedure was done. The child recovered well and was sent home with due instructions. On the first postoperative day and 2 weeks after lens aspiration, the child's visual axis was clear and was following light.

Discussion

Tetralogy of Fallot is the most common cyanotic congenital heart disease.^[1] It is characterized by the presence of a VSD aortic overriding, pulmonary artery outflow obstruction and right ventricular hypertrophy. Pulmonary atresia is considered as the extreme form of TOF.^[2] Patients with TOF present at various ages most common being infancy. Problems associated with TOF such as chronic hypoxia, polycythemia, coagulopathy in congestive heart failure, embolism, and altered acid-base status cause significant mortality and morbidity in patients undergoing anesthesia for various surgical procedures.

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Our patient was brought to the hospital for ocular complaints rather than cardiac manifestations primarily because of unawareness of the uneducated parents. The decision to correct the ocular problem first was based on an attempt to prevent any further visual loss in the child. Reversibility of amblyopia depends on the stage of maturity of the visual system at which the abnormal visual experience began, the duration of deprivation, and the age at which therapy was instituted. The most critical period is probably when the patient is younger than 2 months.^[3] If amblyopia is not managed properly, visual deprivation during this period usually causes severe and permanent visual loss and permanent nystagmus.^[4] If visual deprivation occurs after the age of 2-3 months, amblyopia is usually reversible to some extent. Visual loss and the development of amblyopia are also dependent on the size and location of the cataract. As in this patient there was total dense cataract occluding pupil, early surgery was required. The visual evoked response in this child showed delayed latency of 132 ms in right eye and 136 ms in the left eye. Hence, despite having cyanotic heart disease, early lens aspiration was preferred than cardiac shunt surgery to prevent irreversible amblyopia.

We preferred IV induction with ketamine in order to maintain systemic vascular resistance and flow from the aorta to the pulmonary artery. Proseal LMA was used to secure the airway. Maintenance of anesthesia with sevoflurane in oxygen and air with a minimum alveolar concentration of 0.9-1 provided stable hemodynamics. Saturation remained more than 60 throughout, and so PG was not required intra-operatively.

The child developing fever, breathing difficulty, and increasing cyanosis 24 h after surgery could be because of dehydration as the child was only on oral fluids. Infective endocarditis could also be a cause due to a minor injury during manipulation of the airway and LMA placement could have also resulted in migration of bacteria into the bloodstream. The fever could also be due to the physiological response to surgery. The child was shifted to cardiac ICU and managed. Modified Blalock–Taussig procedure was done when the child was stable after 1-week. Postoperative recovery after cardiac surgery was unremarkable.

Conclusion

The decision to correct the cardiac defect or cataract first was a dilemma. The urgency of preventing irreversible amblyopia in the child lead us to carefully plan and manage anesthesia in this child prior to correction of cardiac defect. Postoperative problems which occurred in this child indicate the need of intensive monitoring and keeping a standby cardiac set up for if the need arises in the postoperative period.

Learning Points

In tetralogy of Fallot patients with severe compromised cardiac functions, noncardiac surgeries can be undertaken with necessary precautions. Pediatric ophthalmologic surgeries which have a definite time period for correction should not be delayed whenever possible. Prevention of irreversible amblyopia is of paramount importance.

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Conflicts of interest

There are no conflicts of interest.

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