Perioperative management of a child with Crouzon syndrome with bilateral partial nasal obstruction

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Crouzon syndrome is an autosomal dominant, genetic disorder characterized by craniofacial dysostosis.[1,2] Perioperative management of these children for craniosynostosis surgery imposes lot of challenges for an anaesthesiologist including management of difficult airway, massive blood loss, hypothermia and difficult extubation.[3,4] Immediate postoperative management is equally important for better outcome. Adequate pain relief and proper positioning after awakening is very important for successful extubation. Here, we want to highlight the immediate postoperative management of a child with crouzon syndrome with bilateral nasal obstruction.

A one-and-half-year-old male child (10 kg), known case of Crouzon syndrome with bilateral choanal artesia was posted for fronto orbital advancement. Child had episodes of cyanosis while crying for which he was evaluated by cardiologist and congenital cyanotic heart diseases were ruled out. On examination, he had typical facial features of Crouzon syndrome (craniosynostosis, exophthalmos, hypertelorism and mid-face hypoplasia) with large tongue. He was breathing by mouth with loud snoring. Parents were asked about the child's usual sleeping position and found that the child used to sleep in prone position on the mother's chest. Since he was obstructing even during sleep and had history of cyanosis in the recent past, he was not given premedication in the ward.

After preparing the operating room (OR) with adequate equipments and drugs, the child was re-examined by the anaesthesiologist in the preoperative holding area. The child was crying and restless and intranasal midazolam (3 mg) was given while the child was monitored by the anaesthesiologist. After 10 minutes, child was calm and the stranger anxiety was completely relieved, child was taken in to the OR and induced with 100% oxygen and sevoflurane. While inducing the child in lateral position (partial obstruction of airway was noted), a 20 G intravenous (i.v) cannula was placed in the hand and anaesthesia was deepened with propofol (2 mg/kg) and fentanyl (2 µg/kg). Adequate mask ventilation required insertion of oral airway. After placing the standard monitors [peripheral capillary oxygen saturation (SpO2), non-invasive blood pressure (NiBP), electrocardiography (ECG), end-tidal carbon dioxide ($ETCO_2$)] patient was paralysed with atracurium and intubated with 4.5 mm endotracheal tube (ETT). A 22 G invasive arterial line and 20 G additional peripheral line was established. Anaesthesia was maintained with air, oxygen (FiO_2 50%) and Isoflurane (1–1.1 Mac) and 5 ug/kg/min of atracurium. Surgery lasted for 6 hours with 250 ml of blood loss which was replaced with crystalloids (500 ml of RL and 350 ml of 0.9% NS) and 200 ml of blood. The child had received total of 7 µg/kg of fentanyl and 0.2 mg/kg of morphine and 2 doses of i.v paracetamol (15 mg/kg) during the start and at the end of procedure. Tranexamic acid was administered to minimise the blood loss (10 mg/kg bolus soon after induction followed by intraoperative infusion of 1 mg/kg/hr). Temperature was maintained between 36.3–36.9^0 C throughout the procedure. At the end surgery, child was reversed with neostigmine (0.5 mg) and glycopyrrolate (0.1 mg). Child was extubated awake, and the vitals were stable. Since he was awake and was trying to sit up and move vigorously, we could not keep him in lateral position or prone position in the OR trolley. So the child was given 0.25 mg of i.v midazolam. While the child was sleeping,

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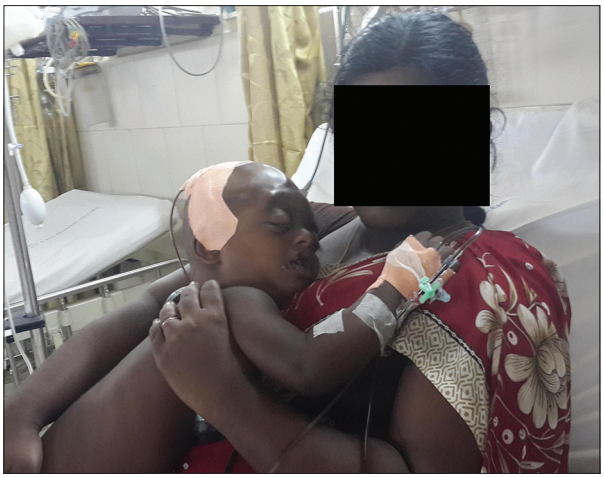

Figure 1: Mother lying in the ICU bed with the baby sleeping on her chest in prone position

the drains and i.v lines were secured on his body. As the child was getting awake, he was placed on the anaesthetist shoulder with oxygen mask and transferred to neonatal intensive-care unit (NICU). In the NICU, the mother was asked to lie down on the ICU bed and the child was placed on the mother's chest on his usual sleeping position [Figure 1]. He was responding to his mother's voice and slept comfortably while maintaining 100% saturation on room air over night without any airway related issues. Postoperatively the child received i.v paracetamol (15 mg/kg) every 6th hourly for analgesia. The child was shifted out to ward the next day and got discharged on the 5th day from the hospital.

Keeping the child awake and calm is very important in the immediate postoperative period to avoid airway-related complication; catheter induced urethral injury, i.v line, drain disconnections and head trauma. Since children are obligate nasal breathers, maintaining a patent airway in a child with bilateral nasal obstruction in the immediate perioperative period is very difficult. Sedating the child with i.v medication to keep the child calm can lead to airway obstruction, especially in difficult airway cases. Smooth induction, administration of adequate analgesia and positioning the child in which the child used to sleep preoperatively will avoid administration of unwanted sedation and airway related complications especially in a difficult airway senario.

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