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Anesthesia Management of a Child with Crouzon Syndrome and Turricephaly for MRI Brain And CT scan of Skull Bone Window- A Case Report

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ABSTRACT:
Crouzon syndrome is a rare genetic disorder involving craniofacial skeleton development. It is characterized by a triad of cranial deformities: premature craniosynostosis, facial anomalies (mid-facial hypoplasia), and exophthalmia[1]. The anesthetic challenges include the presence of a difficult airway, a history of obstructive sleep apnea, congenital cardiac disorders, hypothermia, blood loss and venous air embolism[1]. We present the successful anaesthetic management of a 3-year-old female child, weighing 10 kgs, posted for magnetic resonance imaging of the Brain and computed tomography Scan of the skull bone window.

1. Introduction

Crouzon syndrome is an autosomal dominant disorder with a prevalence of 1 in 60,000 live births with a higher male preponderance 3:1[2-4]. Stemming from fibroblast growth factor receptor 2 gene mutation, it is characterized by premature craniosynostosis of coronal and sagittal sutures, brachycephaly, midface hypoplasia, mandibular prognathism, hypertelorism, shallow orbit, proptosis, cleft palate, increased intracranial pressure, deafness, seizures and mental-impairment[2]. Skull growth perpendicular to fused suture gets restricted and compensatory growth occurs at the remaining open sutures, allowing continued brain growth causing abnormal shape of skull, resulting in difficult airway for Anaesthesiologists. Anticipation of difficulty in airway management helps anesthesiologists to avoid potential airway complications.

2. Case Report:

A 3-year-old female weighing 10 kg was planned for MRI Brain and Brain CT bone-window. Consent for publication was obtained from her parents. She is the second-born from a non-consanguineous marriage and her elder sibling also has similar features.

She was postnatally diagnosed with Crouzon's Syndrome with craniosynostosis of coronal and sagittal sutures and resultant turricephaly with features of facial dysmorphism – bilateral proptosis, hypertelorism, high arched palate and short-stature (Fig.1). Developmental history revealed ability to take few steps without support and vocalize monosyllabic words. There was no history suggestive of congenital heart disease, seizures, thyroid disorders, or hyperreactive airways though snoring history was present.

Airway examination revealed high-arched palate with adequate mouth opening and neck movements. Pulse rate was 132 bpm and blood pressure 90/58mm Hg with room air oxygen saturation of 99% in all four limbs. Systemic examination was unremarkable. A 22G intravenous cannula was secured in her right forearm. Blood investigations, chest Roentgenogram AP-view and 2D Echo were within normal limits for age.

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Figure 1: Child having short stature, Turricephaly, proptosis and high arched palate.

General anesthesia with minimal use of opioids and propofol was planned. The possible consequences of sedation and general anesthesia such as difficult intubation, the need for post-procedure intensive care unit stay were discussed with parents and written informed consent was taken. A difficult airway cart comprising oropharyngeal airway of size 0, laryngoscope with miller and macintosh blades 1 and 2, 2nd generation Supraglottic Device i-gel size 1.5 and 2, and endotracheal tubes of appropriate sizes were kept ready.

The co-operative child with her play toy was shifted to MRI room zone 3 and was positioned with rolled sheets under the neck and shoulders to facilitate neck extension during SGD placement (Fig.2). A gel head ring was placed underneath the head to optimize position during airway manipulation. Pre-induction monitors according to ASA standards were connected.

Inhalational induction was planned to preserve spontaneous respiration. After pre-oxygenation with 6 litres of 100 % Oxygen with a tight seal facemask, an incremental increase in Sevoflurane concentration to reach an end-tidal Sevoflurane concentration of 2%, and 1 mcg/kg intravenous fentanyl were administered to induce anesthesia. Chin lift, jaw thrust, and oral airway were needed to ensure adequate airway patency during spontaneous respiration. Bilateral chest movement and end-tidal carbon dioxide ($etCO_2$) were ensured. After confirming adequate bag and mask ventilation, an additional dose of iv Propofol 1 mg/kg was administered, and airway was secured with i-gel size 2. Eye protection was done with taping of eyes using a transparent dressing, and the everted right lower eyelid was properly covered with sterile gauze, taking care to avoid contact with bulbar conjunctiva and cornea (Fig.3).

Figure 2: Positioning during airway management.

Figure 3: Eye care measures.

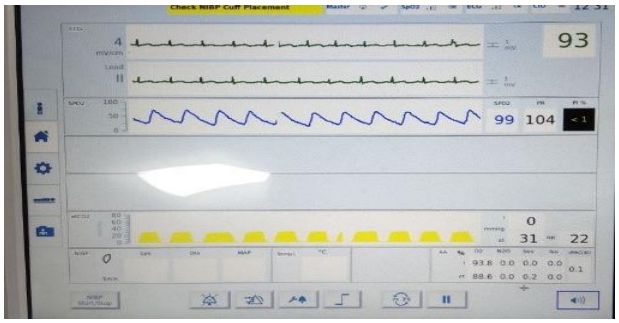

Figure 4: Monitoring including etCO2.

Anesthesia was maintained using oxygen, air, and sevoflurane in Zone III and after shifting into Zone IV, anesthesia was maintained with

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Isoflurane present in MRI-compatible vaporizer in the anesthesia machine in our institute. Nitrous oxide was avoided to prevent further increase in intracranial pressure. Following MRI which lasted for 45 minutes, child was shifted to CT room with Modified Jackson Rees circuit connected to Oxygen cylinder and intravenous Propofol 0.5 mg/kg was used for maintenance during the transit. The intra-hospital transfer was uneventful. After CT scan, the Supraglottic Airway device was removed when the child was fully awake and shifted to recovery room for observation. Post-procedure monitoring was done for an hour, and the child was shifted back to ward. Interestingly MRI Brain showed a narrow nasopharyngeal space, large tongue and mandibular prognathism that explained the difficult mask ventilation during anesthesia (Fig.5).

Figure 5: MRI – narrow nasopharyngeal space, large tongue and mandibular prognathism.

3. Discussion

The most challenging aspect in this case is airway management. Upper airway obstruction can be attributed to septal deviation, mid-nasal abnormalities, and nasopharyngeal narrowing. Hence, using nasopharyngeal airway to maintain upper airway patency can be traumatic. Hence, we opted for the oropharyngeal airway along with airway manoeuvres- chin lift, jaw thrust and neck extension, which can be complex in a child with turricephaly.

The combination of reduced consciousness and/or excessive opiates, sleep disorder, and any co-existent soft tissue edema or secretions can lead to recurrent airway obstruction upon emergency and recovery which may be relieved by airway manoeuvres and suction.

Another important aspect to consider is Tracheal Cartilaginous Sleeve which is a congenital malformation caused by fusion of tracheal arches that may be isolated to few tracheal arches/ include the entire trachea/ extend beyond the carina into the bronchi. The fusion of cartilaginous rings of trachea makes the trachea rigid. Postnatally, neonates with TCS present with biphasic stridor requiring airway intervention. The dynamic airflow of the rigid trachea interrupts mucociliary clearance causing mucous stasis and plugging, infection and bronchospasm.[5,6] However, there was no history of neonatal respiratory distress or recurrent respiratory tract infections in this child.

Mandibular prognathism makes bag-mask ventilation difficult, while laryngoscopy and intubation are easier than in patients with retrognathism[7]. Turricephaly can impair neck extension in supine position, hence optimal positioning is achieved with rolled blanket underneath the neck and shoulders and soft-gel head ring for head support. Eye protection is crucial in proptosis.

Considering MRI's remote location from theatre premises, anticipation of difficult airway helped in optimal pre-procedure planning. A structured airway-management plan to tackle failed mask ventilation/laryngoscopy/intubation essentially led to the successful management of this case.

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4. Conclusion:

Anaesthetic management of a child with Crouzon syndrome presents with its unique challenges in airway management, intra-hospital transfer and other concerns with non-operating room anesthesia. Individualized anesthesia plan with attention to detail for airway management strategy and vigilant monitoring during the procedure is essential for successful anaesthetic outcome.

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