

Anesthetic management of a preterm infant with Crouzon's syndrome

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Abstract : Crouzon syndrome is a rare genetic disorder, characterized by craniofacial dysostosis including skull deformities, facial anomalies and ocular problems. Management of the difficult airway of these patients is the most challenging aspect for an anesthesiologist. We describe successful anesthetic management of a preterm infant with Crouzon's syndrome with difficult airway, inspiratory stridor and laryngomalacia for permanent tarsorrhaphy to prevent exposure keratopathy.

Key Words : Crouzon Syndrome, preterm infant, laryngomalacia, difficult airway, ophthalmic surgery, fiberoptic bronchoscopy, inspiratory stridor

INTRODUCTION

Crouzon syndrome is also known as branchial arch syndrome. These patients have multiple system involvement including craniosynostosis, maxillary hypoplasia and ocular problems such as proptosis due to delayed and abnormal growth of the cranium (1-3). They may also have respiratory symptoms like upper airway obstruction, obstructive sleep apnea (OSA) and difficult airway. They usually need proptosis, craniosynostosis and airway surgery. We describe the anesthetic management of a preterm infant with Crouzon's syndrome scheduled for ophthalmic surgery. Written consent for the publication of the patient's details and photographs has been obtained from parents.

Case Description

A 20 weeks old female baby weighing 3.5 kg, known case of Crouzon syndrome was scheduled for permanent tarsorrhaphy. The baby was born preterm at 32 weeks (postmenstrual age 52 weeks) and stayed in the neonatal intensive care unit (NICU) for 10 days. Oxygen was supplemented using oxygen hood for 3 days without need for mechanical ventilation.

On examination, she had grossly deformed head, proptosis, exposure keratopathy, and retrognathia (Fig. 1). Her computed tomography of



Fig. 1. — Preterm infant with grossly deformed head, proptosis, exposure keratopathy and retrognathia.

the head revealed craniosynostosis with abnormal configuration of skull, including bilateral temporal bossing, flat occiput, and features of raised intra cranial pressure (ICP). She was receiving syrup phenytoin 10 mg twice daily for seizure prophylaxis. Echocardiography, hematological, and biochemical investigations were within normal limit.

She also had inspiratory stridor with supraglottic and intercostal retraction since birth. Airway examination revealed small nasal cavities, small mandible, high arch palate, large tongue, and narrow submandibular space. Preoperative nasal fiberoptic laryngoscopic (FOL) evaluation (2.2 mm size) showed floppy epiglottis, elongated aryepiglottic fold, and prolapsing arytenoids with signs of laryngomalacia.

The patency of the nasopharyngeal area of the patient was checked during pre-anesthetic examination by fogging glasses with the exhaled

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air of the baby. An informed parental consent was taken for possibility of postoperative ventilation and emergency tracheostomy. Indeed, being premature, she was more prone to perioperative apnoea and respiratory distress. Arrangements for NICU with postoperative ventilation facility were made. In the preoperative room, one drop of 0.025% oxymetazoline was instilled in both nostrils.

In the operating room (OR), a difficult airway cart was prepared and standard monitors were attached. A 24 G intravenous cannula was inserted on the left foot at first attempt. Anesthesia induction was started using 100% oxygen (O₂) and incremental increase in sevoflurane concentration through an Ayers' T piece. Assisted ventilation was not possible despite all efforts. Adequate chest rise was not seen with different sizes and types of face mask and bag, despite appropriate positioning of the head and neck. Even after changing head position and using oral airway, and, later, using a Classic size 1 laryngeal mask airway (LMA), the child could not be ventilated. As the patient was breathing spontaneously, arterial oxygen saturation (SpO₂) was maintained between 96 and 98%. A lubricated 3.0 mm uncuffed endotracheal tube (ETT) was cut at 9 cm and inserted in the right nasal cavity to the nasopharynx, after softening it with warm water. Assisted ventilation was possible using a nasopharyngeal tube, and the end tidal carbon dioxide (EtCO₂) concentration was kept between 40 and 45 mmHg. Oral fiberoptic bronchoscopy (FOB) (2.8 mm size) was done under inhalational anesthesia, and, once the glottis was visible, the trachea was intubated using the size 3.5 mm uncuffed ETT. The patient's stomach was decompressed using an orogastric tube. After confirming bilateral equal air entry, ETT was fixed and volume controlled ventilation was started with closed circuit. There was no episode of desaturation during this period. Both eyes were protected with gauge pad and taped. Anesthesia was maintained using O₂, nitrous oxide, and sevoflurane. A balanced salt solution was administered as maintenance fluid. Analgesia was provided with 7.5 mg/Kg of intravenous paracetamol and local infiltration using 2 ml 1% lignocaine. Surgery lasted for 30 minutes, with minimal blood loss. At the end of surgery, sevoflurane was switched off and a 3.0 mm ETT was inserted through the nasal cavity to the nasopharynx. Extubation of the trachea was performed when she was awake and after returning to spontaneous ventilation. Further oxygen supplementation was performed using the nasopharyngeal tube in the NICU during the initial two hours, and using a

face mask during the next 4 hours. There was no increase in the severity of stridor or desaturation postoperatively.

DISCUSSION

Crouzon syndrome is a rare, autosomal dominant disease arising from a fibroblast growth factor receptor 2 gene mutation, characterized by premature craniosynostosis of coronal and sagittal sutures, brachiocephaly, midface hypoplasia, mandibular prognathism, hypoplastic maxilla, hypertelorism, proptosis, short upper lip, crowding of teeth, cleft palate, and other abnormalities (3,4).

Proptosis may lead to exposure keratopathy, corneal opacity, and poor vision, therefore increasing the odds of eye trauma. Severe exposure keratopathy requires permanent tarsorrhaphy like in the present case. Problems in the present case included prematurity, increased ICP, and difficult airway due to maxillary and mandible hypoplasia, large tongue, small submandibular space, and laryngomalacia (4).

Upper airway obstruction in Crouzon syndrome can be due to septal deviation, midnasal abnormalities, and nasopharyngeal narrowing. Tracheotomy may be required to relieve airway obstruction (5). These patients may also have obstructive sleep apnea (OSA) due to craniofacial anomalies. Sudden infant death has been reported due to severe OSA at home (6). Tracheal cartilaginous sleeves (7) and complete cartilaginous trachea leading to congenital tracheal stenosis (8) have also been reported.

Our case had inspiratory stridor with supraglottic and intercostal retraction, floppy epiglottis, and prolapsing arytenoids with laryngomalacia. General anesthesia may increase the severity of respiratory distress, with an inability to assist ventilation and requiring emergency tracheostomy. In these patients, tracheostomy is also technically difficult, and may lead to complications (9). Therefore, we were prepared for emergency tracheostomy.

Difficult airway patients can be best managed with awake intubation. Although there are few case reports in the literature regarding techniques of awake intubation in children, it was not a suitable technique in our case due to raised ICP (10,11). As a consequence, we planned general anesthesia with sevoflurane, allowing precise titration of the depth of anesthesia in comparison to intravenous induction. Eyes protection was insured to prevent trauma by the face mask.

We had different types and sizes of pediatric face mask and oral airway ready, insofar as we suspected difficult mask ventilation. In the present case, assisted face mask ventilation was not possible. With a shortened nasopharyngeal tube and manual closure of the other nostril and mouth, spontaneous as well as assisted ventilation with O₂ and sevoflurane became possible. This technique has been used in other difficult airway patients (12). KAMLIN *et al.*, in a randomized controlled trial, suggested that the naso-pharyngeal tube may be considered a suitable alternative to the face mask for stabilization of preterm infants at birth, or in the neonatal ICU (13). Specific pediatric nasopharyngeal tubes that are softer than endotracheal tube (i.g. the Rusch nasal airway, Teleflex medical inc., NC, USA) are now available.

Tracheal intubation can be assisted with a videolaryngoscope, fiberoptic bronchoscopy (FOB), Bonfil's rigid bronchoscope, and using supraglottic devices, provided that they are available and have proper size, and that the practitioner is familiar with their use. Adequate depth of anesthesia is also essential for the use of those devices. Initially, we tried to avoid the insertion of a nasopharyngeal tube, in order to avoid trauma of the airway, which may lead to difficult visualization through FOB. Although nasal intubation has the advantage of an easier manipulation for FOB, and of greater postoperative comfort without the need of sedation in the NICU, we did choose nasal intubation at first. Indeed, our patient had a small nasal cavity, which could not accommodate 3.5 mm ETT. We were able to intubate the trachea orally at first attempt with a 3.5 mm ETT. In case of non-availability of proper size FOB, nasal fiberoptic guided oral intubation has also been done successfully in neonates and infants with difficult airway (12). Two nasal ETT were also used, one serving as a nasopharyngeal airway to assist the ventilation, and other serving as the tracheal tube for FOB (14).

Extubation of infants with severe stridor is difficult, and they may have increased the risk of airway obstruction. In the present case, we inserted a nasopharyngeal tube before extubation. The trachea was extubated when the infant was fully awake and further O₂ supplementation was provided through the ETT. The patient did not have desaturation postoperatively.

CONCLUSION

Infants with Crouzon syndrome have difficult airway, stridor and, craniosynostosis, which should be managed with proper planning. Small ETT can be used as a nasopharyngeal airway to maintain anesthesia during FOB. Premature infants with stridor can be managed with intravenous paracetamol and local anesthetic agent to achieve good postoperative recovery following short duration procedures.

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