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INHALATIONAL INDUCTION: A SAFE ANAESTHETIC MANAGEMENT IN A PATIENT WITH CROUZON SYNDROME

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Abstract

Crouzon syndrome is a rare genetic disorder involving craniofacial skeleton development. It's characterized by a triad of cranial deformities: premature craniosynostosis, facial anomalies (mid-facial hypoplasia), and exophthalmia. The anaesthetic management challenges include the presence of a difficult airway, history of obstructive sleep apnea, congenital cardiac disorders, hypothermia, blood loss, and venous air embolism. We present the case of an infant with Crouzon syndrome who was scheduled for a ventriculoperitoneal shunt placement managed with inhalational induction.

Keywords

Crouzon syndrome • craniofacial skeleton • genetic disorder • anaesthetic management

Introduction

Crouzon syndrome (CS) is a rare genetic disorder involving craniofacial skeleton development. It's characterized by a triad of cranial deformities: premature craniosynostosis, facial anomalies (mid-facial hypoplasia), and exophthalmia [1,2]. It has a prevalence of 1 in 25,000 live births worldwide and constitutes approximately 4.8% of all craniosynostosis. The anaesthetic management challenges include the presence of a difficult airway, history of obstructive sleep apnea, congenital cardiac disorders, hypothermia, blood loss, and venous air embolism [3,4] . Herein, we describe the anaesthetic management of an infant with Crouzon syndrome scheduled for ventriculoperitoneal (VP) shunt placement.

Case Report

A 7-month-old female baby with a large head weighing 6.5 kg was scheduled for a ventriculoperitoneal (VP) shunt. She was admitted with chief complaints of cold, cough, and nasal discharge since birth. Her mother reported the infant had a history of snoring episodes while sleeping along with mouth breathing. There was no significant family history. An ear, nose, and throat consultation was obtained, and a diagnosis of adenotonsillitis had been established. The abnormal presentation of the head warranted a neurosurgery opinion, which revealed premature craniosynostosis. The craniosynostosis had led to hydrocephalus which required placement of a VP shunt on an emergency basis. Airway examination revealed a narrow oral aperture. The neck was short with restricted movement. Craniofacial examination revealed a malformed skull with a prominent occiput, hydrocephalus, a prominent forehead with frontal bossing, mild exophthalmos, mild convergent strabismus, a compressed nasal bridge, and mild mandibular prognathism (Figure 1).

Upon clinical examination, the pulse rate was observed to be 122 bpm and blood pressure to be 80/52 mmHg. Conducted sounds could be heard throughout the lung surface area. No heart abnormalities were detected by auscultation. Results from other investigations were within normal limits, and hemoglobin was estimated at 11.2 gm/dl. The patient was planned to undergo placement of a VP shunt by the neurosurgeon. The various implications and complications of general anaesthesia were discussed with the parents. Stress was laid on the possibility of a difficult intubation and the need for mechanical ventilation in the paediatric intensive care unit postoperatively. After the counselling session, the parents agreed to general anaesthesia and the written informed consent was taken.

In the pre-anaesthetic room, a 22 G IV cannula was secured, and intravenous fluid was started immediately with Ringer's lactate (60 mL) plus 10% dextrose (40 mL) at the rate of 20 mL/hour. One unit of 100 mL paediatric purified red blood cells was arranged. In the operating room, a difficult airway cart was prepared, and standard monitors were attached. Positioning of the patient was particularly challenging in this case due to the unusual prominent occipital protuberance. A rolled cotton pad was used to support the occiput.

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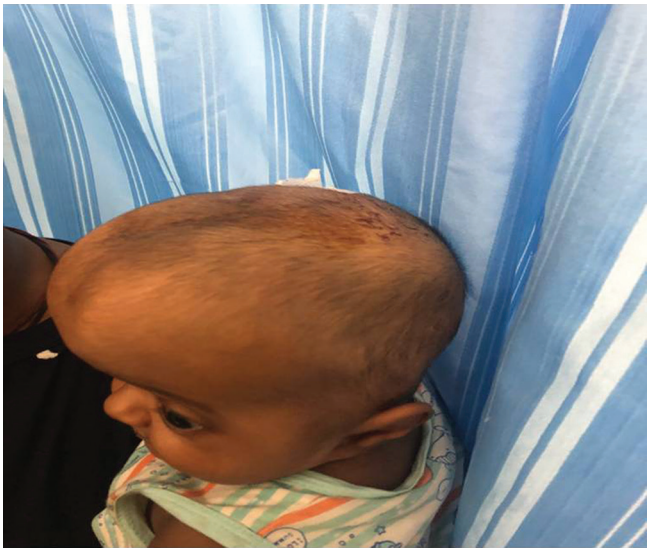

Figure 1.

It was planned for inhalational induction to preserve spontaneous respiration in view of anticipated difficult intubation. Anaesthesia was induced with 100% oxygen and incremental increase in sevoflurane concentration with 1ug/kg of intravenous fentanyl. An oral airway was needed to ensure adequate bag mask ventilation. A lubricated 4.0 mm uncuffed endotracheal tube (ETT) was inserted into the trachea. Bilateral chest movement and end tidal carbon dioxide graph, ETT was secured. Injection propofol 5 mg IV and injection atracurium 0.5 mg/kg IV was administered. Anaesthesia was maintained using O2, air, and sevoflurane. Analgesia was provided with 15 mg/kg of intravenous paracetamol. Surgery lasted for 30 minutes with minimal blood loss. After the administration of injected neostigmine 0.25 mg IV and glycopyrrolate 0.05 mg IV, the extubation of the trachea was carried out. When the patient was fully awake, she was shifted to a recovery room and observed for about one hour.

Discussion

Crouzon syndrome is a rare, autosomal dominant disease arising from a fibroblast growth factor receptor 2 gene mutation, and it's characterized by premature craniosynostosis of coronal and sagittal sutures, brachycephaly, midface hypoplasia, mandibular prognathism, hypoplastic maxilla, hypertelorism, proptosis, short upper lip, crowding of teeth, cleft palate, and other abnormalities [5]. Once a suture gets fused, growth perpendicular to that suture gets restricted, and the fused bones act as a single bony structure. Compensatory growth occurs at the remaining open sutures to allow continued brain growth but causing abnormal bone growth. Multiple premature sutural synostosis in the skull base sutures eventually lead to facial deformities such as those visible in our patient. Characteristic dysmorphic features are detected at birth or in infancy.

The most challenging aspect in a case of Crouzon syndrome is airway management under general anaesthesia. Upper airway obstruction in Crouzon syndrome can be due to septal deviation, mid-nasal abnormalities, and nasopharyngeal narrowing. Tracheotomy may be required to relieve airway obstruction [6]. Most of the time, these patients present with obstructive sleep apnea (OSA) due to craniofacial anomalies. Tracheal cartilaginous sleeves and complete cartilaginous trachea leading to congenital tracheal stenosis have also been reported [7]. Laryngeal mask airway and endotracheal intubation by conventional methods have proved to be challenging [8]. Problems in the present case included increased ICP and difficult airway due to a large occiput, large tongue, small submandibular space, and a narrow oral aperture. Preserving respiration with induction by inhalational anaesthesia was safe.

Though anaesthetic management is very challenging in patients with Crouzon syndrome, counselling and proper preoperative planning in anticipation of a difficult airway can help manage things safely.

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