CASE REPORT

Airway management in a patient with Crouzon syndrome proposed to orthognathic surgery

Melissa Fernandes,¹ Ana Eufrásio,¹ João Bonifácio,¹ João Marcelino²

¹Anesthesiology, Centro hospitalar e Universitário Coimbra, Coimbra, Portugal
²Oral and maxillofacial surgery, Centro hospitalar e Universitário Coimbra Coimbra Portugal

Correspondence to
 Dr Ana Eufrásio,
 eufra@gmail.com

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SUMMARY

Airway management is one of the most important anaesthetist's skills as major complications of airway management, although rare, can be among the most life threatening in medicine. Crouzon syndrome is a rare condition with physical characteristics that can result in difficult airway manipulation. A correct preanaesthetic evaluation and a planned preinduction strategy should be designed to facilitate intubation.

BACKGROUND

Crouzon syndrome is an autosomal dominant condition caused by mutations in the fibroblast growth factor receptor-2 (*FGFR2*) gene which is mapped to chromosome locus 10q25–10q26.¹ It is characterised by cranial synostosis, hypertelorism, exophthalmos, parrot-beaked nose, short upper lip, hypoplastic maxilla and a relative mandibular prognathism. The craniofacial dysostosis can cause severe airway problems and often obstructive apnoea.²

In the presence of a predictive difficult airway, it is essential that a preplanned preinduction strategy is designed to facilitate intubation. Awake intubation is considered a non-invasive intervention intended to manage a difficult airway. Studies with observational findings indicate that awake fiberoptic intubation is successful in 88%–100% of patients with difficult airway.³

Facial appearance improvement, a possible malocclusion correction and severe obstructive sleep apnoea are often the reasons why patients with Crouzon syndrome are submitted to orthognathic surgery.^{4 5} Le Fort III midface advancement projects components of the midface, cranium, zygoma and mandible with the purpose to reduce exorbitism and malocclusion along with airway improvement.⁴ Usually this procedure is performed between the ages 8 and 12 years or from the age of 18 years.⁵

This paper presents a case of difficult management of fiberoptic intubation in a patient with Crouzon syndrome proposed for Le Fort III procedure.

The patient reviewed the case and gave written permission for the authors to publish the report.

CASE PRESENTATION

A 27-year-old female patient with Crouzon syndrome and class III malocclusion, proposed for Le Fort III osteotomy to improve airway obstruction.

The preoperative medical evaluation showed a Crouzon syndrome diagnosed when she was an infant based only on the anatomic changes. No genetic study was performed. She presented complete blindness of left eye and deafness of left ear, in a sequence of craniosynostosis surgery that was performed at 2 years of age. She had right eye glaucoma with a visual acuity of 1/10, a severe decrease in auditory acuity in the right ear, which was being partially corrected with a hearing prosthesis and a description of snoring and sleep apnoea. She was only medicated with dorzolamide ophthalmic solution and oral contraception. The physical examination showed exophthalmia, hypertelorism, hypoplastic maxilla, small oral cavity filled with the tongue and small nares. Airway was evaluated as Mallampati class IV. Preoperative tests did not present any significant abnormalities.

Despite her clinical condition, she was a single woman fairly autonomous in her daily life and very comprehensive about her situation. She postponed this surgery since 13 years of age, but because of airway obstruction during sleep she was now scheduled. She was in the presence of her father, both very anxious concerning surgery but mainly with the postoperative period.

In the presence of a predictive difficult airway, the patient and her father were previously informed and consent the risks, the possibility of awake intubation and the need for mechanical ventilation in the postoperative period.

On the day of surgery, an additional anaesthetist with high experience in fiberoptic intubation was available to serve as an assistant. A portable storage unit containing special equipment for difficult airway management and an emergency trolley were available in the operating room. The patient was carrying her hearing prosthesis to guarantee the possibility of communication and cooperation.

Heart rate, ECG, invasive left radial artery blood pressure and oxygen saturation were continuously displayed.

Face mask preoxygenation was administered for 3 min and the patient was sedated with propofol maintaining spontaneous ventilation. A laryngoscopy using Airtraq device (regular size blade) was attempted but it resulted in a visualisation Cormack and Lahane class IV and a consequent failure in tracheal intubation. Fiberoptic intubation, with the patient sedated, was attempted three times unsuccessfully. Both strategies had no complications and the patient was then awakened. It was attempted an awake fiberoptic intubation that was explained

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once more to the patient. The oropharynx and larynx were nebulized with lidocaine spray 2%. Preoxygenation was administered for 3 more minutes, intravenous fentanyl and midazolam were administered slowly (100 µg fentanyl and 1.5 mg midazolam). Oxygen was provided by face mask positioned near open mouth. After rechecking fiberoptic bronchoscope (Karl-Storz) a cuffed armoured endotracheal tube 7.0 mm was lubricated and applied through the mouth. Three millilitres of lidocaine 2% were administered topically by a spray-as-you-go technique. During the procedure the only abnormal registry was a decrease of oxygen saturation to 60% during a few seconds with no cardiac changes. Nevertheless, during the all procedures a stress-free environment was kept.

After tracheal intubation, correct position was confirmed watching respiratory movements by capnography and thorax auscultation.

Anaesthesia was induced using 50 µg fentanyl, 200 mg propofol (2.5 mg/kg) and 40 mg rocuronium (0.5 mg/kg) and was maintained using sevoflurane 2% and a perfusion of remifentanyl. The endotracheal tube was then transferred by the surgeon to the submental position and intubation was again checked using the same procedures stated previously.

The patient underwent an attempt of Le Fort III procedure with no success, so a Le Fort I procedure was completed. After 9 hours of surgery, the patient was transferred to the recovery room sedated and ventilated to prevent obstruction due to surgery oedema. In the recovery room, the correct position of endotracheal tube kept in the submental position and was again confirmed using thorax radiography.

Patient remained haemodynamic stable, sedated and intubated in the recovery room for 3 days on corticotherapy, analgesia and antibiotic therapy. On the fourth day postsurgery, with surgeon's approval, she was kept in spontaneous ventilation and after emerged from sedation, she was successfully extubated and transferred to the oral and maxillofacial surgery ward.

OUTCOME AND FOLLOW-UP

Due to the unsuccessful surgical procedure attempt, the patient maintains most of the previous complaints related to obstructive sleep apnoea. She has been proposed to a rhinoplasty surgery to be carried out briefly, seeking to partially reduce her symptoms. She attended an anaesthesia consultation and because of her predictable difficult airway she will be subjected to a new awake fiberoptic intubation.

DISCUSSION

Airway management is one of the most important anaesthetist's skills as major complications of airway management, although very rare, can be among the most life threatening in medicine and accounts for a significant percentage of anaesthesia death related.⁶ Difficult airway claims arose throughout the perioperative period mostly on induction.⁷ For these reasons establishing airway predictors to foresee a difficult airway becomes vital. Although common preoperative evaluation scores lack sensitivity and specificity for stratification of airway management risks, they represent practical means, especially for those unskilled in airway management, to detect and prevent a possible difficult airway.⁸

In a patient with Crouzon syndrome, it is predictable that the airway abnormalities, allied to the small oral cavity filled with tongue and a high resistance to airflow via nasal route, make ventilation by mask difficult.² As airway management in Crouzon syndrome can be challenging, it is relevant to consider

a regional approach if it is indicated. Neuroaxial anaesthesia may circumvent the problems that Crouzon syndrome presents, but may be difficult to carry out, because of vertebral fusion and the presence of scoliosis.⁹ Regional anaesthesia may be an alternative choice.

In cases of general anaesthesia, it is necessary a preplanned preinduction strategy that considers various interventions designed to facilitate intubation if a difficult airway occurs.^{3 10}

Airtraq laryngoscope is designed for the difficult airway as it shows a view similar to direct laryngoscope with the additional advantage of less airway manipulation and a skilled technique with shorter learning curve.¹¹

An awake fiberoptic intubation is a non-invasive intervention intended to manage a difficult airway.³ Although awake intubation can be achieved using local anaesthesia alone, sedation is often required to allow patient tolerance.¹² In this clinical case, the anxiety level shown in preanaesthetic evaluation and the absence of pulmonary disease, despite the risk of difficult airway, were the key factors taken in consideration to perform sedation.

Despite there are no references on this subject, we believe that the success of tracheal intubation using awake fiberoptic intubation, rather than sedated, may be explained by the preservation of upper airway muscle tone and the help of voluntary glottic movements which improve the larynx structures visualisation.

Intraoperative airway management in a facial reconstruction, when intubation via oro and nasal route is contraindicated, is another challenge to both surgeon and anaesthetist. Tracheotomy is often the choice in this subset of patient nevertheless it has some documented complications and should be carefully considered when none or short period postoperative ventilation is needed.¹³ Submental intubation gained a wide acceptance as an alternative approach in these situations because it is very simple to perform and has lower morbidity than tracheotomy.^{13 14}

Our report shows the challenge of airway management in the Crouzon syndrome, despite being rare, is very serious and can be life threatening.

Learning points

- ▶ Airway management complications, although rare, are among the most life threatening in medicine and it is of utmost importance for anesthesiologist to predict and handle situations like these.
- ▶ Crouzon syndrome is a rare condition with physical characteristics that can result in difficult airway manipulation.
- ▶ A correct preanesthetic evaluation and a planned preinduction strategy should be designed to facilitate intubation.
- ▶ Awake fiberoptic intubation is a viable choice for tracheal intubation in surgeries that require general anaesthesia and should be performed by skilled professionals.
- ▶ Submental intubation is a feasible alternative to tracheotomy in patients with predictable short-term postoperative ventilation.

Contributors MF was responsible for data collection and analyses and prepared the manuscript draft. AE, JB and JM conducted the clinical case during the all procedure including patient recruitment and management of clinical situation. All authors approved the final manuscript.

Competing interests None declared.

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