Clinical Reports

Anaesthesia for Treacher Collins syndrome using a laryngeal mask airway

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Patients with Treacher Collins syndrome pose a serious problem to anaesthetists in maintaining their airway because of retrognathia. Two patients with Treacher Collins syndrome undergoing tympanoplasty are reported in whom a laryngeal mask was used in place of an endotracheal tube for airway maintenance.

Les patients atteints du syndrome de Treacher Collins posent un problème sérieux à l'anesthésiste pour le maintien des voies aériennes à cause de la rétrognatie. Deux patients atteints de ce syndrome devant subir une tympanoplastie sont rapportés ici où le masque laryngé fut utilisé au lieu du tube endotrachéal pour le maintien des voies aériennes.

Treacher Collins syndrome consists of congenital and familial deformities of the ears, eyes, maxilla and mandible.[1] The syndrome is often associated with cleft lip and palate, deafness due to meatal atresia and malformation of the middle and inner ear. The syndrome raises serious problems in maintaining the airway because of retrognathia.[2]

The laryngeal mask (LM) is a new type of oral airway which forms a seal around the larynx.[3,4] Its use in cases of difficult tracheal intubation in adults[5–7] and children[8,9] has been reported. We report here two patients with Treacher Collins syndrome undergoing tympanoplasty for malformation of the middle ear in whom a laryngeal mask was used in place of an endotracheal tube for airway maintenance.

Key words
AIRWAY: management;
EQUIPMENT: laryngeal mask;
SYNDROMES: Treacher Collins.

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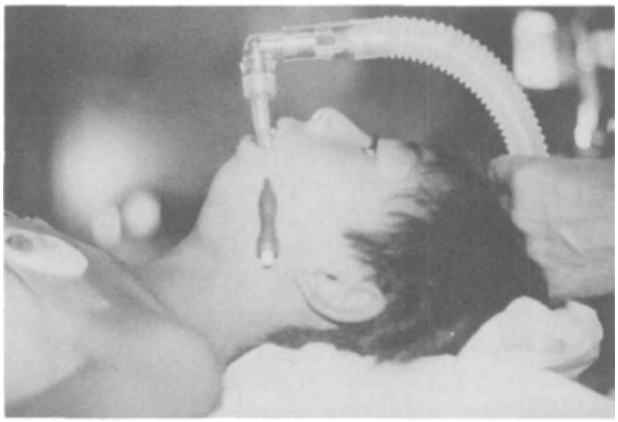

FIGURE Typical facial appearance of patient with Treacher Collins syndrome.

Case 1

A nine-year-old boy, weighing 25 kg, entered our hospital for tympanoplasty for hearing disturbance due to malformation of the middle ear associated with Treacher Collins syndrome. He had attended an orthodontist for hypoplasia of the mandible. There was no family history of Treacher Collins syndrome, and he had never suffered from any particular disease in the past. Routine preoperative findings showed no abnormality, and he was doing well at school without mental retardation. He showed the characteristic facial appearance of the syndrome, oblique palpebral fissures, protruding teeth and receding chin (Figure). Roentgenographic cephalometric analysis showed a considerable retrognathia, and difficult tracheal intubation was anticipated.

Premedication with hydroxyzine 25 mg and atropine 0.25 mg was given *im* 30 min before entering the operating room. After confirming that the lungs could be ventilated easily with a face mask, general anaesthesia was induced with thiamylal 75 mg *iv* and continued with nitrous oxide, oxygen and halothane until the depth of anaesthesia was sufficient for LM insertion. As expected, several attempts to view the larynx with a laryngoscope were unsuccessful. Then, an LM (Intavent, size #2) was placed over the larynx. The cuff was filled with 15 ml of air and manual ventilation was easily performed with minimal gas leak at an airway pressure of 15 cmH_2O. Anaesthesia was maintained with nitrous oxide, oxygen and isoflurane; no muscle relaxants were used. With the resumption of spontaneous respiration, assisted ventilation was applied during surgery. After the operation, the LM was removed with ease when the patient began to swallow.

Case 2

An eight-year-old girl, weighing 19.5 kg, underwent stapedectomy for hearing disturbance due to middle ear malformation associated with Treacher Collins syndrome. There was no family history of Treacher Collins syndrome. She had had palato-plasty surgery for cleft palate repair at the age of five years with general anaesthesia. The anaesthetic record showed that retrograde nasotracheal intubation had been performed, because orotracheal intubation was impossible. Apart from the characteristic appearance of the syndrome such as receding chin, the presence of severe snoring on sleeping, she was otherwise healthy. Cephalometric analysis also showed retrognathia.

Anaesthesia was conducted in a similar fashion to Case 1. Thiamylal and nitrous oxide, oxygen and halothane were administered for induction of anaesthesia. Direct laryngoscopy revealed that the epiglottis could not be visualized. An LM (size #2) was inserted and maintenance of the airway was satisfactory. The course of anaesthesia was uneventful and the patient was discharged nine days later without any trouble.

Discussion

Treacher Collins syndrome was first reported as a combination of congenital notching of the lower lid and deficient malar bones.[10] Since then Francescetti[11] and others have described the syndrome "mandibulo-facial dysostosis" with a broader series of deformities than those described by Treacher Collins. The condition has a strong familial incidence and the patients are usually mentally normal. It is sometimes associated with cleft lip or palate. The patients often need palato-plasty, tympanoplasty and other plastic procedures in childhood, and require general anaesthesia. The most serious problem during anaesthesia is maintenance of an adequate airway. Ross reported a three-year-old boy in whom difficulties were experienced in ventilating the lungs and intubating the trachea, and he advised that spontaneous respiration should be maintained for intubation of trachea.[2]

Clinical examination of the present two cases suggested that tracheal intubation would be difficult because of the receding chin and protruding teeth and this was confirmed by roentgenographic cephalometric analysis.[12]

When difficult intubation is anticipated, several alternatives to routine intubation are possible. These include awake intubation with topical anaesthesia, use of fibreoptic bronchoscopy, guided retrograde intubation, or tracheostomy. The LM was designed by Brain in 1983,[3] and has been in widespread use particularly in the UK as a new form of oral airway. The LM can be inserted to the level of the larynx without laryngoscopy, and it permits positive pressure ventilation with a gas-tight seal up to 15 mmHg of airway pressure. Its use in three adult cases of difficult intubation was reported by Brain.[5] In those patients, the intubation difficulties were associated with an anterior larynx, and the author succeeded in passing the LM to the level of the larynx easily. However, the author stated that insertion of an LM can be difficult in patients having a posteriorly placed larynx, which tends to block downward progress of the tip of the mask. There is also one report of a patient in whom LM insertion was difficult because of large pharyngeal tonsils.[13] In our patients with Treacher Collins syndrome associated with glossoptosis due to retrognathia, anaesthesia was induced carefully after confirming that the lungs could be ventilated easily with a face mask. No muscle relaxants were used. The LM was inserted easily and fitted well for adequate ventilation.

In conclusion, the use of a laryngeal mask was a safe and useful technique for airway maintenance in children in whom tracheal intubation was difficult because of retrognathia.

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