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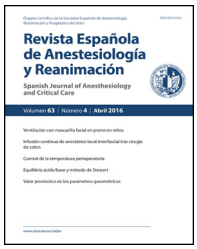

CASE REPORT

The airway approach to a neonate with Treacher Collins syndrome – Case report

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KEYWORDS
Mandibulofacial dysostosis;
Newborn;
Airway management;
Bronchoscopes;
Laryngeal masks

Abstract Neonates and small infants with syndromes characterized by the presence of craniofacial abnormalities may represent great challenges regarding the management of the airway. We describe the case of a 9-day-old neonate with Treacher Collins syndrome, in which a laryngeal mask was essential to improve the airway obstruction, ventilate the patient and serve as an airway conduit for a fiberoptic intubation. By presenting this case, we intend to show that in neonates with Treacher Collins syndrome, in whom difficulties ventilation and intubation are expected, a thoughtful airway management planning is mandatory.

PALABRAS CLAVE
Disostosis mandibulofacial;
Recién nacido;
Manejo de la vía aérea;
Broncoscopios;
Máscaras laríngeas

Manejo de la vía aérea en un recién nacido con síndrome de Treacher Collins. Caso clínico

Resumen Los recién nacidos y los lactantes pequeños con síndromes caracterizados por la presencia de anomalías craneofaciales pueden representar grandes desafíos en el manejo de la vía aérea. Describimos el caso de un recién nacido de 9 días de edad con síndrome de Treacher Collins, en el que una mascarilla laríngea fue esencial para mejorar la obstrucción de la vía aérea, ventilar al paciente y servir como guía para una intubación con fibrobroncoscopio.

Con la presentación de este caso se muestra cómo en los recién nacidos con síndrome de Treacher Collins, donde se prevé una ventilación y una intubación difíciles, es obligatoria una planificación cuidadosa del manejo de las vías aéreas.

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Introduction

Treacher Collins syndrome (TCS), Franceschetti-Zwahlen-Klein, or mandibulofacial dysostosis, is a rare disorder of craniofacial development (incidence of approximately 1:50,000 live births) secondary to mutations of the *TCOF1* gene, which encodes a nucleolar phosphoprotein known as treacle.[1] TCS is mainly characterized by maxillary, zygomatic, and mandibular hypoplasia combined with a small oral aperture, a high arched palate and temporomandibular joint abnormalities. Patients with this syndrome are particularly difficult or even impossible to mask, ventilate or intubate.[2]

Management of difficult airway in children remains one of the most relevant and challenging task for anesthesiology. Children have limited pulmonary reserve and these are prone to upper airway obstruction.

Usually, neonates with TCS present a difficult airway, and for this reason, a thorough knowledge of pediatric airway management techniques is critical on order to minimize complications and to achieve a successful tracheal intubation. Several options to traditional direct laryngoscopy have been described: since blind intubation through the laryngeal mask airway (LMA), video laryngoscopy intubation until oral or nasal fiberoptic intubation; however, usually, these techniques are a challenge in the pediatric population.[2–5]

We report the case of a TCS neonate, in which the LMA improved the airway obstruction and was useful as a canal for a flexible fiberoptic bronchoscope (FFB). The FFB was passed through the LMA and in situ a tracheal tube into the trachea.[6–8]

Clinical case

A 9-day-old male neonate, weighing 2650 g was admitted to the neonatal intensive care unit with respiratory distress due to upper airway obstruction, that was partially relieved by the insertion of a LMA I-Gel® 1 after of some unsuccessful attempts of orotracheal intubation with direct laryngoscopy. He had the craniofacial abnormalities normally associated with TCS, and the difficulty to ventilate and intubate was predictable.

Although adequate oxigenation was achieved with LMA, ventilation was impaired due to air leak when in mechanical ventilatory support. Therefore, it was decided to attempt intubation in the operating room, where difficult airway equipment was available.

One senior pediatric anesthesiologist and pediatric intensivist were present in the operating room, and after a case discussion, planned the different airway approaches trying out the different airway devices available, starting with the most easy and common used ones during inhaled anesthetic with spontaneous ventilation.

In the operating room, before any procedure, the airway devices were checked: Frova® intubating introducer (catheter 8.0 Fr, 35 cm), Airtraq® video laryngoscopy (size 0), FFB (PortalView®: Working Length, 600 mm) and the FFB would fit easily through a 3.0 and 3.5 mm uncuffed tracheal tube, and it could pass both together through a size 1 LMA I-Gel®. During the procedure, standard monitoring was used, administering an air/oxygen mixture and increasing inhaled anesthetic through the LMA in order to maintain spontaneous ventilation. LMA was useful to assure patient airway and to support ventilation when it was needed.

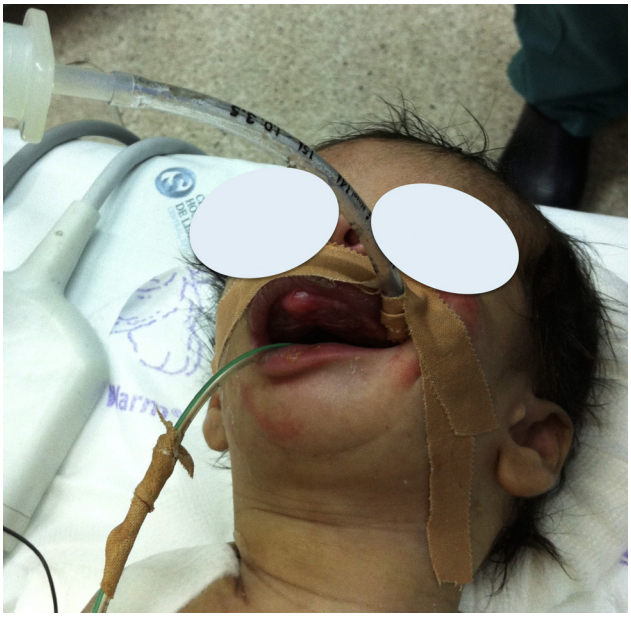

Figure 1 Successful intubation with a 3.5 uncuffed tracheal tube.

First-attempt, blind insertion of the Frova® introducer through the LMA, and the second-attempt, after through out the LMA, a combination of Airtraq® video laryngoscopy with Frova® intubating introducer, were unsuccessful, so finally, we used FFB through the LMA.

However, we had a problem with the last technique, because the tracheal tube length was too short, and the proximal end of tracheal tube disappeared into LMA when the tube was advanced through the trachea. This makes it difficult to safely remove the LMA without dislodging the uncuffed tracheal tube. For this reason, we loaded a second tube on the FFB in order to extend the length of the first tracheal tube, screwing the distal end of 3.0 mm uncuffed tracheal tube to a proximal end of a 3.5 mm uncuffed tracheal tube. This method can be used as a stabilizer during removal of the LMA, is easy to assemble, and avoids the need to cut or modify the LMA.

The FFB was threaded through the tracheal tube and both together were introduced easily through the size 1 LMA I-Gel® and advanced under video-screen visualization into the trachea until the carina was visualized. Then, the two contiguous uncuffed tracheal tubes were passed completely through the LMA and advanced down over the FFB into the trachea. The LMA was removed from the mouth, and then we removed the FFB. We disconnected the two tracheal tubes, and the proper tracheal tube position was confirmed by end tidal CO_2 and the auscultation of bilateral breath sounds.

The patient was intubated with a 3.5 uncuffed tracheal tube railroaded by the FFB, through the LMA, at the second attempt, and vital signs were always within normal range (Fig. 1).

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