CASE REPORT

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

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Received 27 June 2016; accepted 10 October 2016
Available online 13 January 2017

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

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

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.
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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 craniofaciales 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 laringea 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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http://dx.doi.org/10.1016/j.redar.2016.10.003
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TCS, Treacher Collins syndrome, 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. 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.

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.

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.

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 8.0Fr, 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.

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 CO₂ 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).
Discussion

In neonates and other Pediatric patients with predicted difficult ventilation and intubation, a thoughtful airway management planning is mandatory. Usually, difficult intubation in these patients is anticipated, which gives us some time to be prepared. But sometimes the difficult airway (not intubation and/or not ventilation) is unexpected, and the management could be a challenge.

The "gold standard" managing of the difficult airway in all age groups is to maintain spontaneous ventilation until the airway is secure. It is challenging to perform an awake fiberoptic intubation on a child with a difficult airway, not only due to the degree of difficulty itself but also the lack of cooperation from the child. Therefore, experience in difficult pediatric airways and knowledge of the different available airway devices is the key. Many airway management techniques had been described in pediatric patients with craniofacial malformations. Another alternatives to direct laryngoscopy are blind intubation through the LMA, video laryngoscopy intubation and oral or nasal fiberoptic intubation. As above described, in our case we used different devices, starting with the simplest and most used one for us. Our first attempt was blind introducing of a Frova® through the LMA into the trachea, followed by a combination of video laryngoscopy, Airtraq® with intubating introducer, Frova®. Both techniques were unsuccessful, therefore, we passed the FFB through the LMA. Failed to tracheal intubation in the first two approaches is justified by the craniofacial abnormalities of TCS. It is well described in the literature that reduces and deforms the oral cavity and the posterior protrusion of the tongue due to mandibular hypoplasia. The decision on using these two devices before step in FFB was based on our technical skills when comparing the different equipment at our disposal. Besides that, there are successful cases describing the use of Frova® and/or Airtraq®. It is important to note that, in order to prevent the risk of airway trauma inherent to the possibility of multiple attempts, each device was only used once.

The LMA has a relevant role in management of difficult airway in children, not only as a rescue airway device used during the failed intubation scenario ensuring a patent airway and assisted ventilation, but also as a canal to blind intubation. LMA can be used as a conduit to blind tracheal intubation or fiberoptic intubation. In our case, LMA was essential to relieve upper airway obstruction and achieve good oxygenation, as well as a canal to FFB tracheal intubation. But sometimes, we are in trouble when we need to remove the LMA and FFB without dislodging the tracheal tube. So many ways have been suggested in the literature to achieve it. Since leave the LMA placed to passing a wire through working channel FFB and after remove the LMA in order to use the wire as a guide to advance the tube into the trachea or use two connected traqueal tubes joined by either wedging the two together, taping them together or with adapted female-to-female connector. In our case, we were successful adding a second tube stabilizer, screwing the distal end of 3.0 mm uncuffed tracheal tube (second tube) to the proximal end of 3.5 mm uncuffed tracheal tube (first tube), to length and maintain the first tracheal tube position while removing the LMA and FFB. Beside, the LMA left in place allowed us to oxygenate and to keep the neonate adequately sedated. Although this airway approach has been described in the literature for some children and adults, we did not find previous reports of the successful use of this intubation method in a 9-day-old neonate with TCS, using LMA I-Gel®.

In summary, we have realized that neonates with predicted difficult airway management, as TCS, a thoughtful and carefully planned airway management is the key. Although there are other techniques described, our case confirms that the FFB remains an excellent choice to manage the difficult airway in all ages. Despite of the LMA insertion might be useful administering oxygen and relieving upper airway obstruction as well as device to pass the FFB to intubate the trachea. Learning to intubate the trachea through a supraglottic airway is a basic skill; it require several discrete steps and can be complicated by the inability to remove the LMA without dislodging tracheal tube.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Conflict of interests

The authors declare that they have no conflict of interest.

References