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Changes in airway dimensions after mandibular distraction in patients with Pierre-Robin sequence associated with malformation syndromes☆

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ABSTRACT

Introduction: The Pierre Robin syndrome, or sequence, is a triad characterised by micrognathia, glossoptosis and upper respiratory obstruction, with or without cleft palate. Most patients respond to postural treatment, although tracheotomy is necessary on extreme occasions. Mandibular distraction is currently an effective therapeutic alternative that elongates the jaw and resolves the respiratory obstruction. The choice of vector for distraction is essential for modifying the dimensions of the airways.

Patients and methods: The objective of this study is to evaluate the changes produced in the dimensions of the upper airways in eight children with Pierre Robin sequence, treated with mandibular distraction, depending on the vector of distraction planned. To this end, a lateral cranial X-ray was performed pre- and post-distraction, tracing a line from the mandibular plane to the base of the tongue and as far as the posterior pharyngeal wall, measuring the millimetres of separation between the two structures.

Results and conclusions: The results showed that the horizontal distraction vector, in the first place, and the oblique vector in the second place, would be the procedures of choice in view of their positive effects on the airways.

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Cambios en la dimensión de la vía aérea en pacientes con secuencia de Pierre-Robin asociada a síndromes malformativos tras distracción mandibular. Planificación del vector de distracción

Palabras clave:
Síndrome Pierre Robin
Micrognatia
Apnea obstructiva
Distracción mandibular

RESUMEN

Introducción: La secuencia de Pierre Robin es una tríada caracterizada por micrognatia, glosoptosis y obstrucción respiratoria alta con o sin paladar hendido. La mayoría de los pacientes responden al tratamiento postural, aunque en ocasiones extremas hay que realizar traqueotomía. En la actualidad la distracción mandibular es la alternativa eficaz de tratamiento que elonga la mandíbula y resuelve la obstrucción respiratoria. La elección del vector de distracción es importante en los cambios de dimensión de la vía aérea.

Pacientes y métodos: El objetivo del estudio es evaluar los cambios producidos en las dimensiones de la vía aérea superior en 8 niños, con secuencia de Pierre Robin, tratados con distracción mandibular dependiendo del vector de distracción planificado. Para ello realizamos una radiografía lateral de cráneo pre y posdistracción, trazamos una línea que une el plano mandibular con la base de la lengua hasta la pared posterior de la faringe y medimos los milímetros de separación entre ambas estructuras.

Resultados y conclusiones: Analizando los resultados obtenidos, el vector de distracción horizontal en primer lugar y en segundo lugar el oblicuo son de elección por su repercusión positiva en la vía aérea.

Introduction

Pierre Robin sequence is a triad characterised by micrognathia, glossoptosis and airway obstruction with or without cleft palate.[1] Its incidence is 1:8500 to 1:30,000 live births.[2] It is frequent in members of the same family. 25% of patients with Pierre Robin sequence may present alterations in deglutition and breathing.[3]

In the Pierre Robin sequence, altered mandibular development appears to be the initial condition, which is altered during the development of the embryo at about the 7th and 11th week of gestation. The position of the jaw towards the back keeps the tongue in a higher position, which prevents the joining of the palate plates, causing a cleft palate. Mandibular micrognathia may cause a downward displacement of the tongue (glossoptosis) towards the pharynx, causing airway obstruction,[4,5] with clinical manifestations ranging from a minor breathing difficulty to severe respiratory distress with cyanosis.

The Pierre Robin sequence develops in an isolated manner, but it may sometimes be associated with different syndromes, the most frequent being Stickler (44%), velocardiofacial (7%), craniofacial microsomia (3%) and Treacher-Collins (5%).[6,7]

The upper airway of a new-born is smaller and anatomically different to the upper airway of an adult. The tongue is relatively bigger, completely occupying the oral and oropharyngeal cavity. Newborns have narrow external nares and breathe only through the nose, since the epiglottis is in a very high position, very close to the soft palate, causing oral breathing difficulty; also in the new-born, the tongue is at the level of C3-4, while in adults it is at the level of C4-5.[8] In patients with Pierre Robin sequence, since the jaw is smaller, the tongue is retracted, making the pharynx size even smaller.

In the Pierre Robin sequence, there are several functional mechanisms causing airway obstruction[9]: (1) backward displacement of the tongue, (2) in this position, it compresses the soft palate and takes it closer to the posterior pharyngeal wall, thus obstructing the airflow, (3) displacement of the lateral pharyngeal walls, and (4) the pharynx contracts as a sphincter.

Airway obstruction can cause hypoxia, hypercapnia, acidosis, development problems, daily drowsiness, inability to gain weight, malnutrition and, if not treated, it may lead to exhaustion, cardiac failure and, finally, death.

The treatment of Pierre Robin sequence may be from a conservative therapy to surgical treatment. Most of these children can improve by being placed in a prone decubitus position until proper growth of the mandible to bring the tongue forward and clear the airway.[10] In patients with severe mandibular hypoplasia and obstructive crisis, surgical treatment is necessary, such as glossopexy, tracheostomy or mandibular distraction osteogenesis.[11,12] A tracheotomy treatment is associated in the long term with a high rate of morbidity and even mortality; also, logically, there are alterations in the learning of the language and social integration.[13]

McCarthy was the first to use mandibular distraction osteogenesis to elongate the jaw, move the base of the tongue forward, enlarge the retropharyngeal space and improve the airway obstruction.[14]

The distraction osteogenesis has many advantages over traditional mandibular advancement osteotomies. We can achieve advances of 20 mm or more with no bone grafts.[15,16] There is consensus that distraction osteogenesis is superior to other techniques because it is a gradual biological process, with positive effects on muscles, nerves and soft tissues. The newly generated bone will be of a better quality bone and can be modelled, and it is a rehabilitation technique for severe