

TONGUE-LIP ADHESION AND OSTEOGENIC DISTRACTION IN ROBIN SEQUENCE

Adhesión-labio lingual y distracción osteogénica en Secuencia de Pierre Robin

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ABSTRACT

Pierre Robin sequence is characterized by micrognathia, glossoptosis, and airway obstruction, with or without cleft palate, which can compromise neonatal respiratory function. The management of this condition has evolved towards surgical strategies to avoid invasive procedures such as tracheostomy. Among the alternatives, tongue lip adhesion (TLA) has been used temporarily to stabilize the airway. At the same time, mandibular distraction osteogenesis (MDO) has proven to be an effective therapeutic option for gradual lengthening of the mandible and widening of the oropharyngeal space. This article presents the case of a neonate with Pierre Robin sequence treated by a combination of TLA and MDO, achieving resolution of the respiratory obstruction without the need for tracheostomy. The importance of these therapeutic options in the integral management of Pierre Robin sequence is emphasized, demonstrating their efficacy in improving neonatal ventilatory function.

Keywords: Tongue-lip adhesion, osteogenic distraction, Pierre Robin sequence.

RESUMEN

La secuencia de Pierre Robin se caracteriza por micrognatia, glosoptosis y obstrucción de la vía aérea, con o sin paladar hendido, lo que puede comprometer la función respiratoria neonatal. El manejo de esta condición ha evolucionado hacia estrategias quirúrgicas para evitar procedimientos invasivos como la traqueostomía. Entre las alternativas, la adhesión labio-lingual se ha utilizado como medida temporal para estabilizar la vía aérea, mientras que la distracción osteogénica mandibular ha demostrado ser una opción terapéutica eficaz para el alargamiento gradual de la mandíbula y la ampliación del espacio orofaríngeo. Este artículo presenta el caso de un neonato con secuencia de Pierre Robin tratado mediante una combinación de adhesión labio-lingual y distracción osteogénica mandibular, logrando la resolución de la obstrucción respiratoria sin necesidad de traqueostomía. Se destaca la importancia de estas opciones terapéuticas en el manejo integral de la secuencia de Pierre Robin, evidenciando su eficacia en la mejora de la función ventilatoria neonatal.

Palabras clave: Adherencia lengua-labio, distracción osteogénica, secuencia de Pierre Robin.

INTRODUCTION

Pierre Robin sequence (PRS) is characterized by the triad of micrognathia, glossoptosis, and upper airway obstruction, conditions in which the retrograde position of the tongue compromises airway patency, resulting in significant airway obstruction.¹

Historically, this sequence may manifest in isolation or association with other syndromes, such as Stickler syndrome, fetal alcohol syndrome, Nager syndrome, or Treacher-Collins syndrome. The incidence of PRS varies from country to country, from 1 in 8,500 to 1 in 30,000 live births.²

Early diagnosis and treatment of PRS are essential to prevent severe complications from respiratory obstruction, such as chronic hypoxemia, increased pulmonary vascular resistance, and even death.³

The failure of prone postural treatment has led to the development of several surgical strategies. Among them, tongue lip adhesion (TLA), introduced by Shukowsky in 1911 and formally described by Douglas in 1946, has been widely used. Over the last decades, this technique has undergone several modifications, incorporating intermuscular sutures, anterior mandibular anchors, retention buttons at the lingual base, and division of the genioglossus muscle for anterior mobilization.⁴

Mandibular corticotomies assisted by intraoral or extraoral devices have proven to be an effective alternative allowing gradual tissue elongation and pharyngeal space enlargement. This technique not only corrects hypoxemia, the principal manifestation of PRS but also reduces the need for tracheostomies and prolonged intubations, thus minimizing the risk of severe pulmonary complications.⁵

CLINICAL CASE

A six-day-old neonate was admitted due to frequent episodes of obstructive apneas and hypopneas, with oxygen saturation dropping to critical levels. Immediate intervention was required to stabilize respiratory function. The procedure, performed under general anesthesia with inhalation and local anesthesia, involved anterior tongue traction. Normal oxygen saturation was maintained both intraoperatively and postoperatively (Fig. 1).

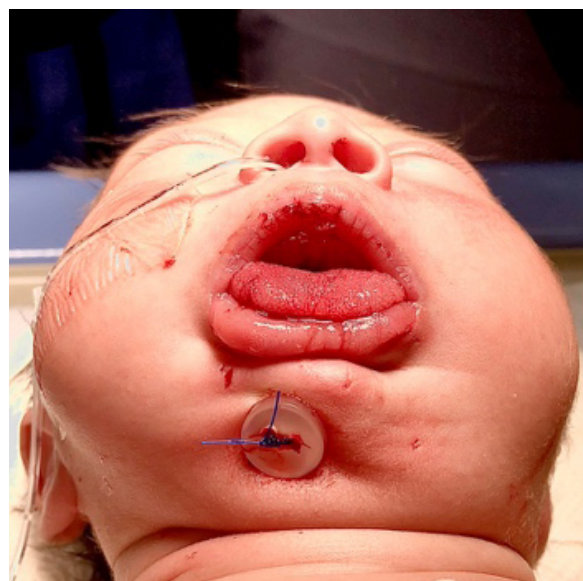


Fig 1. Transoperative image (tongue-lip adhesion)

To further stabilize the tongue and prevent airway obstruction, a polypropylene (1-0) suture was placed through the chin skin, labial and lingual flaps, and anchored at the base of the tongue, maintaining the tongue in an advanced position. A button was placed at the posterior part of the tongue to prevent retroversion, securing it to the chin skin. This approach successfully ensured proper tongue positioning and contributed to the stabilization of respiratory function.

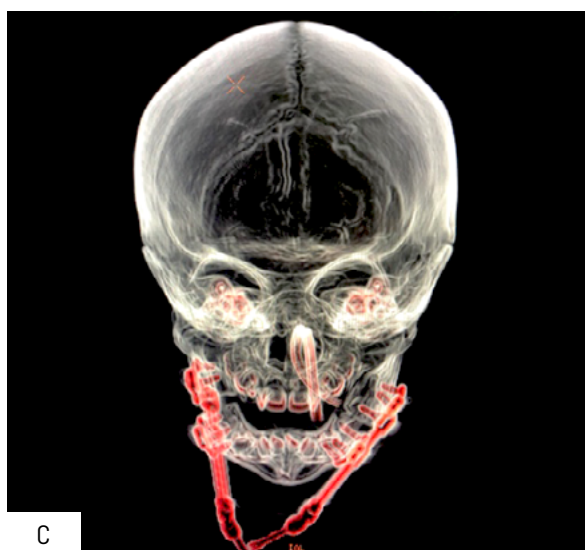
Surgical planning included tomographic studies to assess the mandibular anatomy (Fig. 2A) and stereolithography models to plan the placement of the mandibular distractors (Fig. 2B). At 19 days of age, under general anesthesia and nasotracheal intubation, mandibular distraction osteogenesis (MDO) surgery was scheduled. An osteotomy was performed on the mandibular body, perpendicular to the occlusal plane, preserving the inferior alveolar nerve. Subsequently, unidirectional titanium intraoral distractors with horizontal vectors were placed, and secured with miniplates and screws. Activation began on the third postoperative day (Fig. 2C).



A



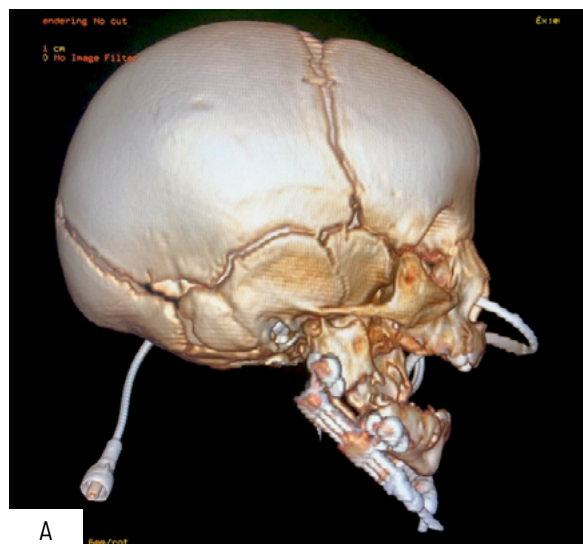
B



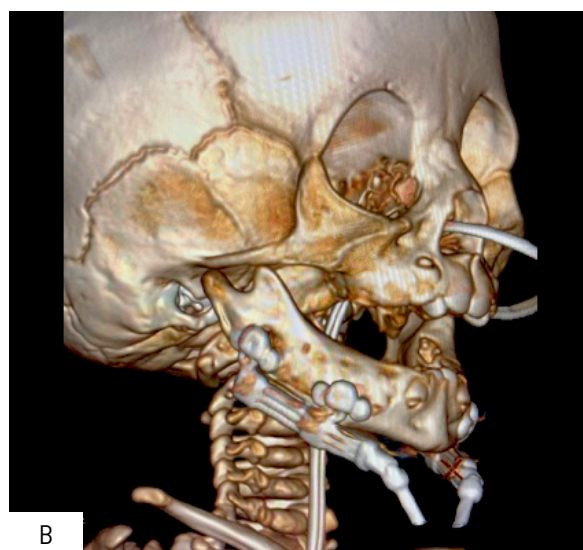
C

Fig 2. Studies to assess the mandibular anatomy. A: Initial tomographic image; B: Stereolithography model image – mandibular distractors; C: Post-surgical tomographic image.

Distraction progressed at a rate of 1 mm per day until a final length of 19 mm, increasing pharyngeal space and resolving respiratory distress (Fig. 3A). The consolidation period was extended up to 6 weeks, with radiological follow-up before the removal of the distractor. Clinical and radiological assessments were conducted to monitor the patient's mandibular development, airway patency, and nutritional status (Fig. 3B). No immediate complications were observed, although a prophylactic antibiotic regimen was administered for 7 days to prevent infections. The final evaluation showed significant improvement in the resolution of respiratory obstruction and adequate functional adaptation, without the need for additional interventions.



A



B

Fig 3. Progress of Distraction. A: Tomographic image 19 days of distraction; B: Final image of the consolidation period

DISCUSSION

In 1891, Lannelongue and Ménard described for the first time two cases of newborns with micrognathia, cleft palate, and glossoptosis. However, it was not until 1923 that Pierre Robin characterized this condition more precisely, establishing it as a pathological sequence resulting from a series of anomalies triggered by an initial malformation. Its clinical expression is variable and may be associated with chromosomal abnormalities, genetic mutations, or the influence of environmental and teratogenic factors. PRRS is considered an isolated condition without a defined inheritance pattern, which excludes its transmission to future generations.⁶

The pathogenesis of Pierre Robin sequence has been extensively studied, and to date, the mechanical theory is the most widely accepted. According to Logjes et al, this theory postulates that oligohydramnios could play a key role in the development of the anomaly, as amniotic fluid restriction would contribute to mandibular deformation and tongue impaction against the palate, preventing its proper closure.⁷ Other hypotheses include the neurological maturation theory and rhombencephaly, which suggest that alterations in brainstem development may be involved in the dysfunction of neuromuscular control of the tongue. In addition, neurogenic theory suggests that defects in fetal muscle tone and innervation may contribute to the characteristic Pierre Robin sequence phenotype.⁸

Respiratory obstruction is one of the main complications in patients with PRS, as it can generate hypoxemia, hypercapnia, and pulmonary edema, also affecting suction and swallowing capacity. According to Khouri et al, upper airway obstruction and alterations in feeding function are crucial aspects of the treatment plan, since a lack of adequate management can lead to chronic hypoxia. These metabolic disturbances can lead to severe consequences, such as right heart failure and cerebral hypoxia.^{3,9}

Several surgical techniques have been described for the management of PRS, each with specific indications and limitations. Tracheostomy continues to be used in critical cases, especially in syndromic patients, although the application is associated with high morbidity, including tracheomalacia, chronic bronchitis, laryngeal stenosis, and risk of death due to complications such as mucous plugs or tube extrusion.¹⁰

Alternatively, lingual adhesion has proven to be ineffective, as it does not resolve the obstruction and may affect phonation and dentition. Subperiosteal release of the lingual musculature of the buccal floor has shown limited results in terms of efficacy. Other methods such as tongue traction with Kirschner wire, subperiosteal release of the buccal floor musculature, and mandibulopexy have been used with varying degrees of success.¹¹

In contrast, MDO has established itself as the first line of treatment today due to its ability to correct micrognathia and improve airway patency. Safri et al, emphasize that this procedure allows controlled elongation of soft tissues, muscles, and neurovascular structures, favoring the resolution of respiratory obstruction without the adverse effects of other techniques.¹² According to Pendem et al, although labiolingual adhesion and ODM have gained greater acceptance, the most frequent complication in labiolingual adhesion is dehiscence, which reinforces the role of ODM as a stable and effective surgical strategy.¹³

Despite its benefits, ODM is not without complications, including infection, neuromuscular disturbances, bone resorption, and bone healing defects, which may require additional interventions. Clinical evaluation by pulse oximetry and objective studies with polysomnography have shown that ODM and tracheostomy are significantly more effective than labiolingual adhesion in reducing the apnea-hypopnea index.^{14,15}

CONCLUSION

The management of Pierre Robin sequence (PRS) requires an individualized approach based on the severity of airway obstruction. In this case, tongue-lip adhesion (TLA) was used as an initial measure to stabilize the airway, allowing immediate improvement in oxygenation. However, given the persistence of respiratory distress, mandibular distraction osteogenesis (MDO) was performed, achieving a progressive increase in pharyngeal space and the resolution of obstructive episodes. The combination of these two surgical strategies proved to be an effective and safe approach, avoiding the need for tracheostomy and its associated complications. Postoperative follow-up demonstrated successful mandibular growth, improved respiratory function, and adequate nutritional adaptation without additional interventions. This case underscores the importance of early diagnosis and timely intervention in PRS management. While TLA can serve as a temporary solution, MDO remains the gold standard for long-term correction of mandibular hypoplasia and airway obstruction. Further studies are

needed to refine patient selection criteria and optimize surgical outcomes for PRS treatment.

CONFLICTS OF INTEREST

The authors declare that they have no conflict of interest.

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Ethical approval

No authorization was required, but the article is in law with the declaration of Helsinki and the patient signed an informed consent regarding his participation and publication of the data obtained.

Informed consent

Consent was obtained from the patient, under the signed consent of the parents of the newborn.

Publication ethics

All images collected and displayed in this article have been obtained with the signed consent of the parents of the newborn.

Bibliography

- Wright M, Knowles RL, Cortina-Borja M, Javadpour S, Mehendale F, Urquhart DS. Airway management in infants with Robin sequence in the United Kingdom and Ireland: A prospective population-based study. *Pediatr Pulmonol*. 2024 Nov;59(11):2839-2849; DOI: <https://doi.org/10.1002/ppul.27140>
- Oubejja H, Menbahi I, Elhasnaoui C, Qurouach S, Benomar I, Ettayebi F. Cleft palate in Pierre Robin sequence. In: *E3S Web of Conferences*. EDP Sciences; 2021 Nov;319: 1-5. DOI: <https://doi.org/10.1051/e3s-conf/202131902013>
- Khoury E, Bisher O, Hamdy J. Tongue lip adhesion (TLA) in the management of airway obstruction and feeding in Pierre Robin sequence, a case report. *Int J Surg Case Rep*. 2024 Aug;121:109932. DOI: <https://doi.org/10.1016/j.ijscr.2024.109932>
- Miloro M. Mandibular Distraction Osteogenesis for Pediatric Airway Management. *J Oral Maxillofac Surg*. 2010 Jul;68(7):1512-23. DOI: <https://doi.org/10.1016/j.joms.2009.09.099>
- Flores R, Tholpady S, Sati S, Fairbanks G, Socas J, Choi M, et al. The surgical correction of pierre robin sequence: Mandibular distraction osteogenesis versus tongue-lip adhesion. *Plastic and Reconstructive Surgery*. Lippincott Williams and Wilkins. 2014 Jun;133(6):1433-9. DOI: <https://doi.org/10.1097/PRS.0000000000000225>
- Leger M, Baudouin R, Thierry B. Debunking the Myth: Should Pierre Robin be Credited for Defining the Pierre Robin Sequence? *Otolaryngol - Head Neck Surg (United States)*. 2024 Nov;172(3):1114-1117. DOI: <https://doi.org/10.1002/ohn.1064>
- Logjes RJH, Mermans JF, Paes EC, Muradin MSM, Don Griot JP, Breugem CC. Assessment of Health-Related Quality of Life in Robin Sequence: A Comparison of Mandibular Distraction Osteogenesis and Tongue-Lip Adhesion. *Plast Reconstr Surg [Internet]*. 2019[cited: 06/11/2024];143(5):1456-65. Available from: <https://www.researchgate.net/publication/331269836>
- Thiers Oliveira Carneiro D, Ferreira da Cunha Filho J, Nascimento da Silva R. Multidisciplinary treatment of Pierre Robin sequence: a review. *Brazilian J Dent Oral Radiol*. 2022 Jun-Dec;1(1):1-6 DOI: <https://doi.org/10.52600/2965-8837.bjdor.2022.1.bjd10>
- Wang C, Shi B, Li J. Management of cleft palate among patients with Pierre Robin sequence. *British Journal of Oral and Maxillofacial Surgery*. Churchill Livingstone. 2023 Sep;61(7):475-481. DOI: <https://doi.org/10.1016/j.bjoms.2023.06.003>
- Morrison K, Collares M V, Flores R. Robin Sequence: Neonatal Mandibular Distraction. *Clin Plast Surg*. 2021 Jul;48(3):363-73. DOI: <https://doi.org/10.1016/j.cps.2021.03.005>
- Horikiri M, Park S, Kinoshita M, Matsumoto D. Respiratory management of Pierre Robin sequence using nasopharyngeal airway with Kirschner wire. *J Plast Reconstr Aesthetic Surg*. 2010 Apr;63(4):390-394. DOI: <https://doi.org/10.1016/j.bjps.2009.09.023>
- Safri S, Wickemeyer J, Valika T. Paradigm Shift in Airway Management for Pierre Robin Sequence: Tongue Base Suspension. *J Craniofac Surg*. 2021

May 1;32(3):E288-90. DOI: <https://doi.org/10.1097/SCS.00000000000007198>

13. Pendem S, Jayakumar N, Gopalakrishnan S, Arakeri G. Systematic review and meta-analysis of surgical approaches for improving airway stability in infants with Robin sequence: evaluating complications and outcomes. *British Journal of Oral and Maxillofacial Surgery*. Churchill Livingstone. 2024 Jul;65(6):511-22. DOI: <https://doi.org/10.1016/j.bjoms.2024.04.003>
14. Scott A, Tibesar R, Sidman J. Pierre Robin Sequence. Evaluation, Management, Indications for Surgery, and Pitfalls. *Otolaryngologic Clinics of North America*. 2012 Jun;45(3):695-710. DOI: <https://doi.org/10.1016/j.otc.2012.03.007>
15. Ribeiro C, Padovani L, Ferreira-Donati G, Almendros M, Castro C, Maximino L. Babies With Pierre Robin Sequence: Neuropsychomotor Development. *Pediatr Neurol*. 2023 Apr 1;141:72-6. DOI: <https://doi.org/10.1016/j.pediatrneurol.2023.01.010>

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