

Recovering Breathing and Feeding of a Newborn with Pierre Robin Sequence

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Abstract

Pierre Robin sequence (PRS) is characterized by the triad micrognathia, glossoptosis, and airway obstruction, commonly associated with cleft palate. This study reports the case of a patient with characteristics consistent with the diagnosis of PRS in the 1st week of life. To stabilize the airway, oro-tracheal intubation was performed without success, followed by tracheostomy and glossopexy and after a small improvement in symptoms, it was decided to perform the mandibular distraction osteogenesis (MDO). At the age of 6, her maxillofacial development was within expectations, with patent airway, the presence of deciduous teeth, and without neurological impairment. The MDO described in this case allowed patient's respiratory and feeding improvement, thus avoiding episodes of cyanosis, allowing timely removal of the tracheostomy and glossopexy, and execution of palatoplasty at the right time. While it is uncertain whether MDO will replace the possible need for future orthognathic surgery, treatment greatly improved the quality of life of the patient.

Keywords: Airway obstruction, mandibular distraction osteogenesis, Pierre Robin sequence

INTRODUCTION

Pierre Robin sequence (PRS) is named after the French stomatologist who described the problems associated with newborn micrognathia in 1923 and 1934.^[1] It is believed that the micrognathia causes upward and posterior displacement of the tongue resulting in obstruction of the airway^[2,3] and preventing the closure of the palatine shelves before the 10th week of gestation.^[4] Thus, the sequence is characterized by the triad of micrognathia (91.7% of the cases), glossoptosis (70%–85%), and airway obstruction.^[2,5] Although the cleft palate is not included in this triad, it is commonly associated with the sequence^[2,5] and it can worsen the airway obstruction, as the tongue may stay in an awkward position.^[5]

Infants with PRS should be evaluated by a multidisciplinary team to assess the anatomic findings, delineate the source of airway obstruction, and address airway and feeding issues.^[2] Positioning will resolve the airway obstruction in 70% of cases. In the correct position, most children will also be able to feed normally.^[2] However, if the infant continues to show evidence of desaturation, other interventions may be indicated as nasopharyngeal tube/stenting,

prolonged intubation, or surgical options as tongue-lip adhesion, mandibular distraction osteogenesis (MDO), and tracheostomy.^[6]

Difficulties in breathing and eating due to the mandibular hypoplasia may cause delayed growth and development, chronic aspiration, gastroesophageal reflux, defects in speech, obstructive sleep apnea, otitis media, and death.^[5] For such, it is imperative that the condition is early diagnosed and adequate measures are taken immediately.

The case of an infant with PRS and her management are discussed highlighting the current approaches for this condition.

Procedure

A request for the evaluation of an inpatient was addressed to the Cleft Lip and Palate Rehabilitation Center of the University General Hospital of the University of Cuiabá. The patient was a female 4-week-old infant presenting with congenital

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mandibular micrognathia, glossoptosis and cleft palate, leading to the diagnosis of isolated PRS.

Clinical signs of difficulty in breastfeeding and shortness of breath were present after birth, and hence the family was oriented about the child's prone or side positioning in case of upper airway obstruction crisis.

Thirty days later, the patient's condition was aggravated by the occurrence of two episodes of cyanosis. After trying to establish a patent airway through orotracheal intubation without success, the child was submitted to tracheostomy. During the procedure, the maxillofacial team also performed a glossopexy in an attempt to clear the hypopharynx for improved airway flow [Figure 1].

There was a little improvement of the symptoms and aiming to decannulate the patient, she was treated with MDO by applying distraction devices placed through a submandibular incision followed by bilateral osteotomy at the angles of mandible avoiding injury to the inferior alveolar nerves and placement of small external mandibular distractors. The pins were inserted into the proximal and distal parts of the osteotomy, the MDO device was installed and the surgical wound properly closed [Figure 2].

After a latency period of 2 days, the activation phase started at a rate of 1.5 mm/day, which lasted for 10 days to accomplish the objective of 15 mm of distraction. Consolidation period of 30 days was secured, then the distractor device was removed on clinical signs of stabilization of upper airway, as well as removal of the suture of the lip to the tongue.

At the age of 14-month-old, the patient was submitted to complete palatoplasty by Von Langenbeck technique [Figure 3] without any further signs of compromised airway at the postoperative period, a concern addressed to the proposal of the closure of the wide communication of the nasal to the oral cavity.

At 3 years of age, a discreet extra-oral scar in the submandibular region and satisfactory mandibular projection were observed clinically [Figure 4]. Maxillofacial development was within expectation. All primary teeth were present and undamaged. Upper airway was patent and no neurological impairment was perceived. The panoramic radiograph showed good bone continuity at the mandibular angle, where osteotomies were previously performed [Figure 5].

At the age of 6 years, facial esthetic and functional harmony were observed [Figure 6]. In spite of Class II jaw pattern, significant improvement in the airway space was confirmed by the lateral view radiography [Figure 7] and symmetrical mandibular growth could be observed through panoramic radiograph [Figure 8].

The patient will continue to follow-up until the end of mandibular growth, until the end of adolescence, when the need for a new surgical procedure for correction discrepancy between the jaws will be evaluated.

DISCUSSION

PRS infants face two main problems: feeding difficulties and upper airway obstruction.^[6] All patients should be closely evaluated by qualified professionals in regard to their ability to be fed. If necessary, early nasogastric feeding shall be initiated to supplement oral feeds and improve weight gain. Most infants will be successfully managed with nonoperative measures alone. If these measures fail to relieve obstruction as evidenced by inadequate results on sleep studies and poor weight gain, surgical options are to be considered.^[2]

In the absence of professional assistance, children with RS and significant airway obstruction may succumb to asphyxia, hypoxia, respiratory failure, cor pulmonale, malnutrition, and death.^[4] Failure to maintain the free airway can lead to chronic obstruction and consequently, carbon dioxide retention, development of pulmonary vasoconstriction, hypertension with right ventricular failure and associated delayed development.^[6] In cases of mild upper airway obstruction, children can be managed by nonsurgical treatment, including careful instruction regarding appropriate feeding techniques, an alertness to symptoms of increased obstruction, and lateral or prone positioning, where the infant is placed in ventral decubitus to displace the tongue anteriorly.^[2,4,6] Patients with severe or refractory upper airway obstruction require more aggressive management. Surgical options most commonly include glossopexy, tracheostomy, and MDO.^[4,6]

In the described case, the infant could not be managed by more conservative feeding and positioning techniques, evidenced by the two episodes of cyanosis. Even the orotracheal intubation was unsuccessful, so both tracheostomy and glossopexy were performed. However, as those procedures are expected to be temporary and still led to poor improvement in the symptoms, MDO was indicated.

Although numerous algorithms are described in regards to surgical decision-making, no uniform consensus currently exists. The clear benefits of one surgical technique versus another have not clearly been determined and neither procedure can be expected to provide normal occlusion for individuals at the time of skeletal maturity.^[2]

Tracheostomy remains the most effective option for airway management for children with upper airway obstruction refractory to other measures, although it is still used as the main surgical option for children with PRS and airway obstruction at some institutions.^[2,4] However, tracheostomy is associated with complications, such as accidental decannulation, obstruction of the tube or long-term problems such as tracheomalacia, growth retardation, delayed speech, articulation difficulties, and behavioral problems.^[6]

Glossopexy, also known as tongue-lip adhesion, is also effective in relieving tongue-base obstruction as the anterior ventral tongue is anchored to the lower lip (mucosa with or without the muscle) and the posterior tongue remains anchored to the mandible.^[4] Promising results are reported to



Figure 1: Glossopexy was performed in an attempt to clear the hypopharynx for improved airway flow

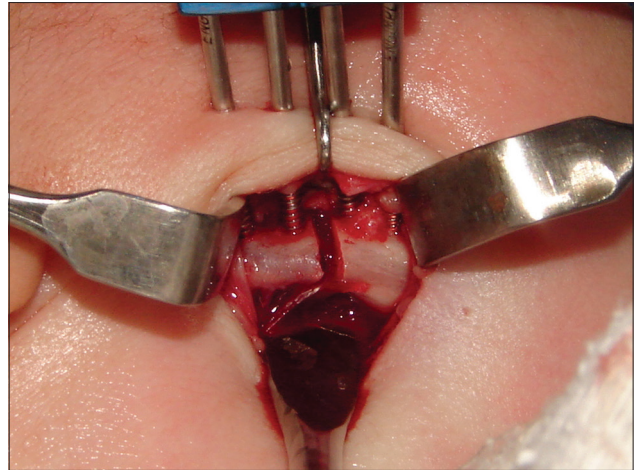


Figure 2: Small external mandibular distractors were placed through submandibular incisions followed by bilateral osteotomy at the angles of mandible

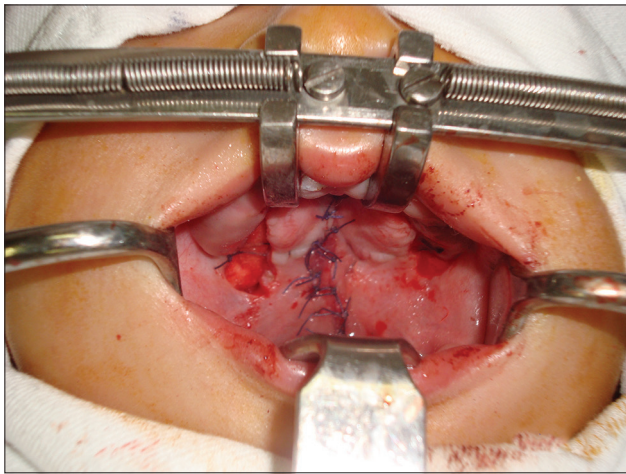


Figure 3: Patient was submitted to complete palatoplasty at age of 14 months



Figure 4: Discreet extraoral scar in the submandibular region and satisfactory mandibular projection



Figure 5: Panoramic radiograph showing good bone continuity at the mandibular angles, where osteotomies were previously performed

be successful enough to clear the hypopharynx of the patient.^[7,8] However, the present case resulted in little improvement of the symptoms and was replaced by a more stable solution, the MDO.

MDO is a relatively new treatment option for neonates with PRS,^[3,6] which has been shown to be very effective in relieving the upper airway obstruction^[3,6,9] by gradually increasing pharyngeal airway size and lengthening the mandible.^[3,4] As the mandible is lengthened, the tongue base moves forward by

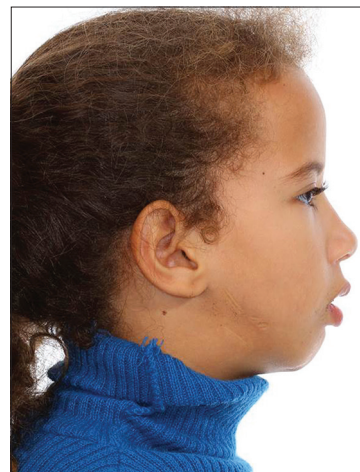


Figure 6: Facial esthetic and functional harmony were observed at age 6 its anterior muscular attachments to the mandible, increasing the airway space and relieving airway obstruction.^[6]



Figure 7: Significant improvement in the airway space was confirmed by the lateral view radiography

The procedure includes bilateral osteotomies and placement of distraction devices, which can be internal or external with percutaneous pins.^[4] External devices are easy to adjust and remove but can be dislodged and are associated with scarring.^[4,6] Internal devices are usually better tolerated than external ones^[4,6] but require repeat dissection for removal under general anesthesia.^[4] In this case, the external device was selected because it was a national brand with affordable costs (considering limitations of the Brazilian Public Health System) and prompt delivery whereas internal devices (all foreign) could only be bought by request and were much more expensive.

High rates of dental complications associated with the MDO have been reported in the past.^[3] The complications included device failure to support resistance; breakdown^[5] damage to the inferior alveolar nerves; infections; failure of distraction; dislodgement of pins or distractors, and damage to the tooth buds.^[6] The use of the newer distractor devices with the improved planning of mandibular osteotomies may result in reduced dental complications.^[3]

When newborns undergo MDO, frequently there are no recognizable dental structures on intraoral examination. Thus, there is a real chance for the tooth buds to be injured by either the osteotomy and/or screw or pin placement. There is also a risk of tooth migration that may occur with the application of the distraction forces.^[3] Therefore, it is important to examine the long-term consequences of the primary dental development in these children.^[3] The patient presented was followed-up for 6 years and no complication was yet observed other than discrete submandibular scars at the distraction sites.

CONCLUSION

The neonatal MDO described in this case allowed the patient to breathe and be fed properly, preventing new episodes of cyanosis. It also enabled timely removal of the tracheostomy and glossopexy



Figure 8: Symmetrical mandibular growth is observed through panoramic radiograph

as well as the execution of the palatoplasty at the proper time. Although it is unsure whether MDO will replace the eventual need for orthognathic surgery in the future, the treatment was found to be safe as it improved patient's quality of life and made possible her development with cosmetically acceptable surgical scars.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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