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Anthropometric Facial, Skeletal, and Airway Changes in Patients with Pierre Robin Sequence Treated with Mandibular Distraction Osteogenesis

Pérez N[1], Maldonado A[2] and Muñoz R[2,3]*

[1]Department of Oral and Maxillofacial, University of Carabobo Maxillofacial Residency Program, Dr. Ángel Larralde University Hospital, Venezuela

[2]Department of Oral and Maxillofacial Surgery, University of Carabobo Maxillofacial Residency Program, Dr. Ángel Larralde University Hospital, Venezuela

[3]Department of Craniofacial Surgery, Palmer Children Hospital, Maryland, USA

Abstract

Background: The following study is carried out, whose objective is to quantitatively determine the anthropometric changes of hard and soft tissues through skeletal, facial, and airway measurements in pediatric patients diagnosed with Sequence of Pierre Robin treated by Bilateral Osteogenic Distraction.

Methods: A descriptive and comparative study was carried out, performing a retrospective analysis of longitudinal design, defining a representative sample of 8 patients (5 male and 3 female), who attended the Oral and Maxillofacial Surgery service "Dr. Atilio Perdomo" from the "Dr. Ángel Larralde" located in the state of Carabobo, Venezuela, during the period 2015-2022 diagnosed with Pierre Robin Sequence that presented clinical characteristics micrognathia, glossoptosis and signs of upper airway obstruction.

Results: To collect the necessary information, several methods were used, such as reviewing the clinical history of the patient under study, performing a facial and intraoral clinical examination, and analyzing conventional radiographs such as orthopantomography, lateral cephalic radiography, and anterior-posterior skull radiography. Thilander, McNamara, and Bjork Jarabak tracings were also used, as well as Cone Beam CT scans and facial photographs. The Jarabak and Thilander tracings showed an improvement in mandibular body length and a more pronounced mentolabial groove. Additionally, an increase in facial height was observed. Despite this, growth was maintained with a brachiocephalic and horizontal pattern in relation to mandibular growth. On the other hand, McNamara's layout showed a favorable evolution in the airway, by measuring the upper and lower pharynx points, as well as the increase in the lower third. These findings were similar to those found in the Jarabak layout.

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***Correspondence:**
Nohelia Pérez, Department of Oral and Maxillofacial, University of Carabobo Maxillofacial Residency Program, Dr. Ángel Larralde University Hospital, Naguanagua, Carabobo, Venezuela

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Introduction

Craniofacial development is a process that involves the proliferation, differentiation, and migration of neural cells to multiple regions of the embryo and is distinguished into several cell types, such as peripheral neurons, enteric neurons, glial cells, melanocytes, smooth muscle and chondrocytes, these cells of the neural crest are pluripotent and are considered to be the fourth germinative leaf of the embryo, which, during neurulation, become mesenchymal and migrate following signals from the extracellular matrix throughout the body of the embryo [1,2].

Bone Morphogenetic Protein (BMP) and fibroblast growth factors play an important role in the formation and migration of neural crest cells, involved in the regulation of differentiation and proliferation [1,2].

Once the neural crest cells have migrated to the craniofacial region, contact-mediated signaling occurs between the tissues at the back of the neural tube, causing the neuroectodermal and non-neuroectodermal border cells to convert from epithelial cells to epithelial cells, highly invasive mesenchymal. For these cells to migrate from the dorsum of the neural tube to the craniofacial region, they must lose their apicobasal polarity and release chemotactic intercellular adhesion complexes, such as Fibroblast Growth Factor-2 (FGF-2) and Fibroblast Growth Factor-8 (FGF-8)

[1,2].

Once the neural crest cells have reached their destination in the craniofacial region, they proliferate and differentiate to form different craniofacial structures, such as bones and connective tissues. Craniofacial malformations are common congenital pathologies that can affect the respiratory tract, cause eating and speech problems, and even put the patient's life at risk, as well as leave irreversible consequences [1,2].

Mandibular Hypoplasia is a common craniofacial malformation that can be classified into three groups: Congenital, developmental, and acquired; it can be the result of trauma or injury, and it is characterized by incomplete or inferior development of the mandible compared to the maxilla [3,4]. This deformity can occur in isolation or be associated with other pathologies, such as the Pierre Robin sequence, hemifacial microsomia, Nager syndrome, Goldenhar syndrome, or some facial clefts [5].

The Pierre Robin Sequence (PRS) is a clinical triad consisting of glossoptosis, mandibular hypoplasia, and upper airway compromise, and in some cases, it may also be associated with cleft palate. SPR was named in honor of the French physician Pierre Robin who provided one of the first descriptions of patients with this condition, called glossoptosis cachexia, in 1923 [5].

To diagnose it, one begins with an evaluation of the airway, because patients with PRS may have breathing and feeding difficulties due to the posterior location of the tongue and mandibular hypoplasia. This can lead to the development of Obstructive Sleep Apnea (OSA), which is characterized by recurrent episodes of partial or total collapse of the upper airways during sleep, which can cause hypoxia, hypercapnia, acidosis, developmental problems, daytime sleepiness, malnutrition and, if left untreated, can lead to exhaustion, heart failure and ultimately death [6,7].

PRS may occur in isolation or be associated with other syndromes, such as Treacher-Collins syndrome, Nager syndrome, velocardiofacial syndrome, and Pfeiffer syndrome. The treatment of PRS can be conservative or surgical, with provisional measures being the use of a nasogastric tube, nasal cannulas, tracheostomy, management in ventral decubitus, and tongue traction, however, these measures can lead to complications such as tracheomalacia, chronic bronchitis, and disorders. of speech and feeding. Applicable surgical treatments include uvulopalatopharyngoplasty, glossectomy, orthognathic surgery, advancement genioplasty and mandibular distraction osteogenesis [8].

In the case of pediatric patients with RPS, Mandibular Osteogenic Distraction (DOM) is the best management method, as it corrects micrognathia by increasing the anteroposterior length of the mandible through gradual bone formation that can achieve significant advances in the jaw, improving the position of the tongue, elevating the hyoid bone and expanding the hypopharyngeal region to eliminate airway obstruction, being a safe surgical procedure during childhood, thus preventing secondary growth deformities and reducing the need for subsequent surgeries [9]. In patients with craniofacial deformities and atrophies of the maxillomandibular complex, this technique has proven to be effective in expanding the underlying bone and soft tissues, obtaining excellent clinical results in addition to reducing morbidity without the need for bone grafting and reducing relapse due to gradual tissue lengthening, restoring facial contour [10,11].

It is important that the treatment and management of PRS be individualized and adapted to each patient's specific needs, requiring appropriate medical evaluation and follow-up to ensure the best possible outcome.

Taking into account the biological and surgical benefits of this mandibular reconstruction technique, the following study is carried out, whose objective is to quantitatively determine the anthropometric changes of hard and soft tissues through skeletal, facial and airway measurements in pediatric patients diagnosed with Sequence of Pierre Robin treated by Bilateral Osteogenic Distraction who attended the Oral and Maxillofacial Surgery Service of the “Dr. Ángel Larralde” during the period 2015-2022.

Methodology

Type and Design

A descriptive and comparative study was carried out, performing a retrospective analysis of longitudinal design, defining a representative sample of 8 patients (5 male and 3 female), who attended the Oral and Maxillofacial Surgery Service “Dr. Atilio Perdomo” from the “Dr. Ángel Larralde” located in the state of Carabobo, Venezuela, during the period 2015-2022 diagnosed with Pierre Robin Sequence that presented clinical characteristics micrognathia and glossoptosis and signs of upper airway obstruction.

Inclusion criteria

Pediatric patients diagnosed with Pierre Robin Syndrome (PRS), with an average age of 5.12 years, underwent bilateral distraction osteogenic surgery (Table 1, 2). An immediate and long-term postoperative follow-up and control of 5 years was carried out. These patients had imaging studies, such as lateral head X-rays, to evaluate their evolution and Cone Beam tomography of the face for pre- and postoperative analysis.

Exclusion criteria

Pediatric patients who presented associated syndromic and systemic pathologies did not have corresponding imaging studies to evaluate their evolution.

Collection technique and instrument

To collect information, the clinical history of the patient under study, facial and intraoral clinical examination, conventional radiograph analysis (orthopantomography, lateral head X-ray, and AP Skull X-ray) with Thilander, McNamara and Bjork Jarabak tracings, as well as Cone Tomography were used Beam and facial photographs. The information was obtained through the measurement of various variables, where the anthropometric parameters included the evolution of Facial and Skeletal Changes, valued from:

Skeletal changes

1. Thilander
- Condyle height
- Ramus height
- Body length

2. Jarabak
- Joint Angle (S-Ar-Go)
- Anterior Cranial Length (SN)

Table 1: Age of the sample under study. Oral and maxillofacial surgery service. University Hospital "DR. "Angel Larralde". 2015-2022.

Age (years)	Amount	Percentage
4	3	37.50%
5	2	25%
6	2	25%
7	1	12.50%
Total	8	100%

Table 2: Gender of the sample under study. Oral and maxillofacial surgery service. University Hospital "DR. "Angel Larralde". 2015-2022.

Gender	Amount	Percentage
Male	5	62.50%
Female	3	37.50%
Total	8	100%

- Gonial Angle (Ar-Go-Gn)
- Branch Height (Ar-Go)
- Body length (GoGn)
- SNB angle
- ANB angle
- Go-Gn-Sn angle
- Facial Depth (N-Go)
- Facial length on the Y axis (S-Gn)
- Posterior facial height (S-Go)
- Anterior Facial Height (N-Pl. Mand)
- Facial Plane (SN-Po)
- Facial convexity (NA-Pg)

3. McNamara

- Effective mandibular length Condylion- Gnathion (Co-Gn)
- Anteroinferior facial height (Ena-Me)
- Mandibular plane angle (Po-Or/Go-Me)
- Facial axis angle (Ba-N/Pt-GnI)
- Perpendicular nasion- Pogonion (Pg-Np)
- Airway: Upper and lower pharynx

Facial changes

- Inter-labial distance Stms-stmi
- Neck angle Sn, Gn-C
- Mento-labial groove

Airway

- Nasopharynx
- Velopharynx
- Oropharynx
- Hypopharynx

The analysis of all the results and elements of the patient's anthropometric changes was recorded in a statistical table as a summary instrument of the information collected, where they were detailed in 3 stages: Presurgical (T0), after the removal of the osteogenic distractors (T1) and in the late postoperative period of 5 years (T2), the skeletal, facial and airway changes, thus allowing comparison and demonstrating the evolution quantitatively.

Analysis technique

The techniques used to analyze the results of this research consisted of a comparative analysis of measurements in mm and degrees of skeletal changes, facial and airway changes, through conventional radiographs and facial photographs.

The average of the dimensions of the skeletal, facial, and airway analysis is obtained from the metric measurement, from the imaging of the patients, performing cephalometric tracings of radiographs such as orthopantomography, lateral cephalic, using the tracings of Thilander, McNamara and Bjork Jarabak, respectively.

Results

In the analysis of the Thilander tracing, the stability in the increase in length, both vertical and horizontal, of the ramus and mandibular body in the patients during the 5-year controls is observed. In patients undergoing bilateral distraction, a mean of 93.57 for the right side and 94.93 for the left side was found, maintaining body length compared to T1. In addition, vertical growth was observed in the height of the branch, with an average of 31.5 for the right side and 31.75 for the left side, which shows a change to T1 (Table 3).

During the 5-year follow-up after distraction osteogenesis, changes were observed in the McNamara tracings in the following aspects:

- An increase in the Co-Gn measurement was evident, presenting an average of T0 of 74.60 mm and an increase of T1 of 96.85 mm, evidencing an increase in the hypoplastic jaw, which is below normal standards.
- There was a vertical reduction in the Ena-Me measurement, which corresponds to the anterior part of the lower third of the face, presenting a decrease of 60.65 in T2 compared to T1, which presented 61.40.
- On the other hand, the Po-Or/Go-Me angles increased, with an average of 36.9°, which indicates an open gonial angle and reveals excessive vertical growth, resulting in a convex profile and a hyperdivergent pattern.
- Regarding the vertical position of the chin about the base of the skull (Ba-N/Pt-Gnl point), an increase in vertical growth was observed.
- In the case of the Pg-Np measurement, an average of -10.5° mm was found, which indicates a mandibular skeletal protrusion.
- Concerning the airway, represented by the upper and lower pharynx points, an increase was observed, since it presented T0 at 6.3 mm with an improvement in which it presented 12.80 mm related to growth (Table 4).

It should be considered that modifications were evident at different points of the Jarabak route. The changes are detailed below:

- At the S-Ar-Go point an increase was observed with respect

Table 3: Thilander analysis of patients with bilateral osteogenic distraction.

Variables	T0		T1		T2	
	Right	Left	Right	Left	Right	Left
	Average		Average		Average	
Condylar Height	9.87 mm	9.75 mm	11.62 mm	11 mm	13.85 mm	13.5 mm
Branch Height	25.75 mm	26.12 mm	29.50 mm	30.37 mm	31.5 mm	31.75 mm
Mandibular body length	84.40 mm	86.93 mm	93.50 mm	94.43 mm	93.57 mm	94.93 mm

Table 4: Descriptive analysis plot of MC Namara of the sample under study. Oral and maxillofacial surgery service. University Hospital “DR. “Angel Larralde”. Period 2015-2022.

Variable		T0	T1	T2
	Normal	Average	Average	Average
Co-Gn	120.2 mm	74.6	96.85	94.25
Ena- Me	66 mm	60.8	61.4	60.65
Po-Or/Go-Me	22.7°	35.75	36.05	36.95
Ba-N/Pt-Gnl	0.2°	2.8	3.7	9.7
Pg-Np	-1.8 mm	-22.85	-10.85	-10.55
Upper Pharynx	17.4 mm	6.3	11.5	12.8
Lower Pharynx	11.3 mm	6	9.95	10.45

Table 5: Descriptive analysis of Jarabak in the study sample. Oral and maxillofacial surgery service. University Hospital “DR. “Angel Larralde”. Period 2015-2022.

Variable		T0	T1	T2
	Rule	Half	Half	Half
Sargo	143°	153	162.2	156.6
Ar-Go-Me	130°	137.15	132.1	133.1
S.N.	71 mm	67.6	67.6	70.2
Argon	55°	60.75	51.4	51.55
N-Go-Gn	75°	79.45	78.7	79.85
Ar-Go	44 mm	29.8	34.3	34.6
Go-Gn	71 mm	55.45	68.2	68.55
SNB	78°	64	72.9	71.7
ANB	2nd	15.2	13.3	13.2
Go-Gn-Sn	32°	44.2	46.3	47.85
N-Go	-	96.45	92.9	92.8
S-Gn	-	96.35	108.3	107.4
S-Go	-	55.5	60.5	60
N-Pl. mand	-	99.4	107.9	112.8
SN-Po	-	64.95	77.5	76.3
N -A-Pg	-	29.25	18.9	18.65

to T1, however, an open joint angle related to greater vertical growth of the mandibular ramus was maintained.

- ArGoMe increased compared to T1 demonstrating positive growth of the ramus and mandibular body

- Although the Ar-Go-N value was maintained, a decrease in the superior gonial angle was represented, indicating forward growth of the branch.

- The N-Go-Gn point increased, which represents a clockwise rotation of the mandibular body.

- The SNB and ANB angles presented an average of 70.7° and 13.2°, respectively, which indicates a small decrease in the T1 value, evidencing a retruded mandible about the base of the skull.

- The Go-Gn-Sn points showed an increase, reflecting vertical growth, while the N-Go point remained stable, with a mean of 92.8°.

- Regarding the S-Gn points, a decrease was observed, referring to horizontal growth of the jaw, and the S-Go point remained stable compared to T1, interpreting it as the posterior vertical growth of the face.

- The N-Pl-Mand anterior facial height presented an increase, reflecting excessive vertical growth.

- About the NA-Pg facial convexity, an average of 18.65° was found, while the SN-Po point decreased (Table 5).

Significant changes can be observed in the airway (Table 6). An increase was recorded in the different analysis points at the level of the pharynx compared to T0, which is within normal measurements. Regarding the interlabial distance, it increased during the postoperative period of distraction osteogenesis and continued to increase in the late postoperative period of 5 years. This increase may be related to the presence of an open bite in the patient, which will be corrected through children's orthopedics and orthodontics.

Regarding the angle of the neck, a lower angulation is observed, while the mento labial groove has increased, which indicates a greater projection of the mental region.

In terms of aesthetic improvement, an increase can be seen in the lower third of the face. Unlike the initial photographs where this was practically non-existent, you can see a distance between the chin

Table 6: Analysis of changes in the airway of the sample under study. Oral and maxillofacial surgery service. University Hospital “DR. “Angel Larralde”. PERIOD 2015-2022.

Variable		T0	T1	T2
	Rule	Half	Half	Half
Nasopharynx	25 mm ± mm	6.15	10.08	11.98
Velopharynx	9 mm ± 3 mm	7.78	10.01	12.22
Oropharynx	16 mm ± 4 mm	7.45	10.09	13.02
Hypopharynx	12 mm ± 3 mm	9.61	12.82	14.88

Table 7: Analysis of soft tissue changes of the sample under study. oral and maxillofacial surgery service. University Hospital “dr. “Angel Larralde”. Period. 2015-2022.

Variable		T0	T1	T2
	Rule	Half	Half	Half
StmS-Stml	02 mm	15.5	16	10
Sn-Gn-C	100°	132.25	122.5	99.25
Labial groove	04 mm	8	5.75	5

Image 1: Imaging evolution Lateral Cephalic X-ray: T0, T1, T2.

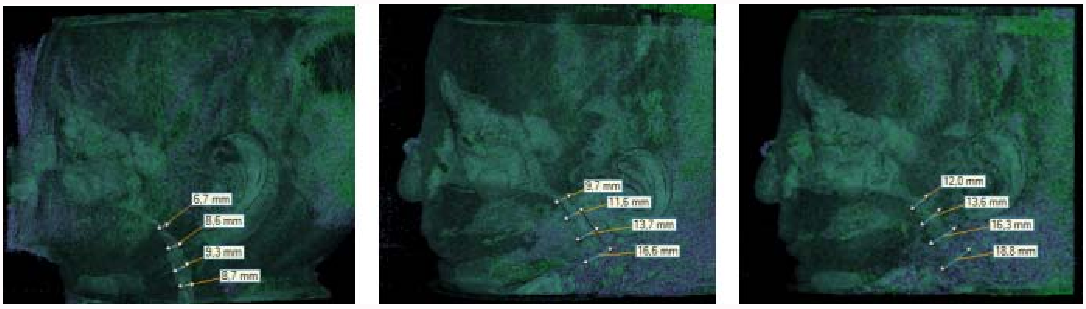

Image 2: CT-type imaging for airway assessment.

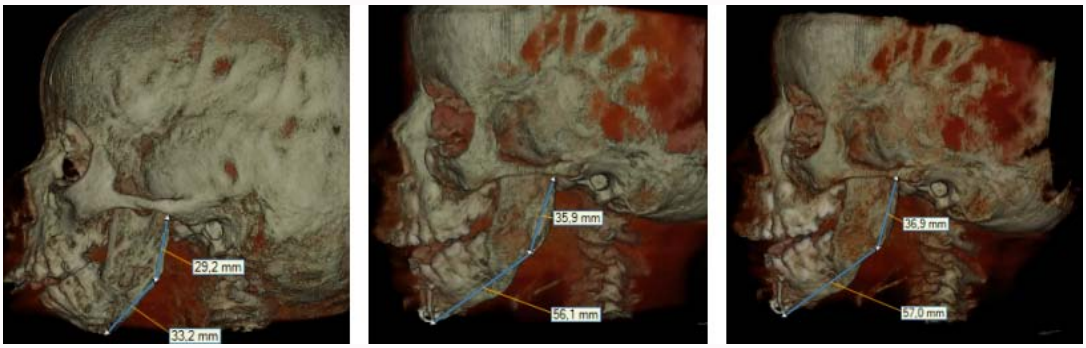

Image 3: CT-type imaging for assessment of mandibular length growth.

and the neck. The distraction osteogenic procedure performed on the mandibular body has contributed to this increase in the mento labial sulcus. Importantly, after treatment, the patient has improved not only her facial appearance but also her functional activity, improved airway layout, and returned to her normal daily activities.

Discussion

In a study conducted in Spain in 2015, Dr. Plaza and her team investigated changes in upper airway dimensions in 8 children with the Pierre Robin sequence treated with mandibular distraction. Two types of distraction vectors were taken into account, oblique in 8 cases and horizontal in 3. Pre- and post-distraction radiographs were performed, measuring the separation between the mandibular plane and the base of the tongue to the posterior wall of the pharynx. The latency period was 2 to 3 days, with a distraction rate of 1 mm/day or 0.5 mm/12 h. Sagittal advancement of the mandible and base of the tongue was observed immediately after distraction. The containment period was 4 weeks for children under 6 months and 6 weeks for those over 6 months. Regarding the results, 3 cases did not present occlusal changes, while in 3 cases a mild anterior open bite occurred that was corrected after the restraint period. Four of the 8 patients needed physical therapy to rehabilitate the masticatory muscles and sucking and swallowing reflexes. Complications related to the pins used in the procedure were recorded, including extrusion in 3 patients and loss on one side requiring correction. No inferior alveolar nerve injuries were observed and infectious complications were minimal [7].

Image 4: Facial analysis evolution of 05 years.

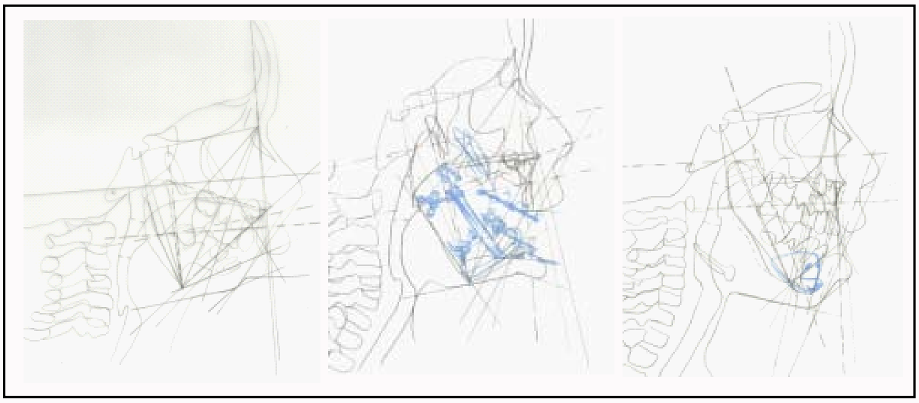

Image 5: Diagnostic tracings in imaging Lateral cephalic X-ray.

Complications related to the pins used in the procedure were recorded, including extrusion in 3 patients and loss on one side requiring correction. No inferior alveolar nerve injuries were observed and infectious complications were minimal [7]. Complications related to the pins used in the procedure were recorded, including extrusion in 3 patients and loss on one side requiring correction. No inferior alveolar nerve injuries were observed and infectious complications were minimal [7].

In the study carried out by Gürsoy and Coll, it was shown that improvements in facial appearance resulting from mandibular distraction osteogenesis gradually decrease with growth. During the 5-year follow-up period, the gradual return of the jaw and facial profile toward its original pre-distraction shape was initially assumed to be a relapse. However, the results showed that the mandibular lengthening achieved was stable and there was no additional growth. Instead, considerable posterior rotation was observed in the mandible. On the other hand, the maxilla continued to grow normally during long-term follow-up. Regarding the correction of mandibular retrognathia and facial convexity, the results indicated that it was not stable in the long term. During the first year, there was a statistically significant relapse in mandibular interrelationship, mandibular retrognathia, and facial convexity. Over the 5 years of follow-up, the mandibular skeletal and soft tissue profiles continued to regress, and the shape and position of the mandible returned almost to their original state before distraction. On the other hand, maxillary, nasal, and labial contour measurements lengthened and moved forward and downward [12]. Compared to the present study, where all gained mandibular lengths were preserved but did not increase as expected through normal mandibular growth, it is evident that there are multiple variables and interactions, many of which are unrecognized, that influence long-term stability after treatment. Therefore,

Conclusion

Due to the previously mentioned results, the facial and skeletal analysis, and the discussion of the present investigation, it is concluded that the control cephalometric analysis after 5 years of having applied distraction osteogenesis, aesthetic, and functional improvements were observed with stability in facial appearance in children with PRS. All mandibular lengths gained were preserved, but no considerable increase was observed as expected through normal mandibular growth.

The Jarabak and Thilander tracings showed an improvement in the length of the mandibular body and a more pronounced mentolabial groove. On the other hand, facial height increased. Despite this, growth with a brachiocephalic and horizontal pattern was maintained in relation to mandibular growth. Likewise, the McNamara layout showed a favorable evolution in the airway, by measuring the upper and lower pharynx points, as well as the increase in the lower third, in a similar way to the Jarabak layout, a convex profile was observed.

Osteogenic distraction is completely safe in pediatric patients with RPS, specifically, the best results were observed in the preschool and school stages because it manages to induce mandibular growth in patients, which results in an improvement in facial appearance, and functionality of the patient since it provides airway patency as a result of the anteroposterior mandibular advancement and the development in the psychological aspect of the patients.

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