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RESEARCH ARTICLE

CONGENITAL AGLOSSIA: AN ALTERNATIVE TREATMENT WITH RAPID MAXILLARY EXPANSION AND MANDIBULAR DISTRACTION OSTEOGENESIS

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ABSTRACT

Purpose: Aglossia is a rare condition referring to the congenital absence of the entire tongue that is associated with activities such as sucking, swallowing, chewing, and phonation playing an important role in facial growth. The objective of this clinical case report is to demonstrate that adequate orthopedic and orthodontic procedures can re-adjust the buccal bone structures by redirecting the growth and the muscular adaptation, contributing to the improvement of the functions of the stomatognathic system.

Methods: A girl 9 years and 4 mouths complained of difficulty to chew and diagnosed with congenital aglossia. The treatment included rapid maxillary expansion and mandibular midline distraction osteogenesis to correct transverse deficiencies and to provide conditions of myofunctional adaptation. It may occur in the course of growth and development of craniofacial complex.

Results: Maxillary and Mandibular bones were successfully expanded allowing a better development of adaptive mechanisms to compensate for absence of the tongue.

Conclusions: The use of a tooth-borne distraction device with a Hyrax-type screw to expand the mandibular by symphysis osteogenesis was efficient and improved the tongue tonus and occlusion. However, multidisciplinary continuous cares are required due to lack of proper stomatognathic function.

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INTRODUCTION

Tongue malformations are congenital structural changes that may vary in size and, rarely, appear to be virtually absent (Cappellette *et al.*, 2013). Often hypoglossia represents part of the oromadibularand limb hypogenesis syndrome (OLHS) that represents a spectrum of disorders characterized by congenital malformations involving multiple sites such as the tongue, mandible, and maxilla with or without reductive limb anomalies (Cappellette *et al.*, 2013; Kalaskar *et al.*, 2016). In the literature, few cases of aglossia and hypoglossia are described and were associated with other manifestations as deafness, situs inversus, and genetic syndromes (Gupta, 2012), such as Moëbius and Pierre Robin (Kantaputra and Tanpaiboon, 2003; Kumar and Chaubey, 2007). All these syndromes are believed to belong to a family of oromandibular limb hypogenesis syndrome (OLHS) (Gupta, 2012).

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The incidence of OLHS is very low (1/175,000 live births) and most cases are actually hypoglossia rather than true aglossia (Kalaskar *et al.*, 2016; Nevin *et al.*, 1975). Most of the cases are sporadic with no sex or racial predilection but a few intra-familial cases led to the hypothesis of mutation in an autosomal recessive gene though no genetic mutations or chromosomal abnormalities have been identified (Gupta, 2012). Moreover, maternal febrile illness, hypothyroidism, and cytomegalovirus infection have also been implicated as etiological factors (Gupta, 2012). Apart from this, teratogenic etiology has also been implicated (Hall, 1971), and drugs like Tigan, Imipramine, Diazepan, and Chlorpromazine are involved in the etiology, but their effect in the causation of syndrome has not been proved (Kalaskar *et al.*, 2016) heat-induced vascular disruption has also been considered as one of the etiological factors for these syndromes (Meundi *et al.*, 2013). Aglossia is an extremely rare condition and refers to the congenital absence of the entire tongue (Gupta, 2012; Bommarito *et al.*, 2012) caused by failed failure of the tongue embryogenesis process (in the fourth to eight weeks of gestation) (Bommarito *et al.*, 2016), whereas microglossia and

hypoglossia refer to abnormal smallness of the tongue (Salles *et al.*, 2008; Tubbs *et al.*, 2016). The first case of hypoglossia was first described in the eighteenth century by de Jussieu (1718/1719) as aglossia (Meundi *et al.*, 2013). The association of tongue anomalies with malformations of limbs was noted by Kettner (1907) and the first report of a true case of Aglossia – Adactylia syndrome was described by Rosenthal (1932) (Sultan Olmez-Gurlen, 2016). In 1971, Hall (1971) classified OLHS into 5 major type (Table 1).However, there are cases reported in the literature in which aglossia was also associated with other congenital deformities as hypodactyli, partial anodontia, rudimentary ear, and conduction deafness (Kalaskar *et al.*, 2010; Kantaputra and Tanpaiboon, 2003; Higashi and Edo, 1996). Therefore, Kalaskar *et al.* 2016suggested modification for type IB Hall's classification (Table 2) (Hall, 1971).

The tongue is the most mobile organ of the body and is associated with coordinated activities such as sucking, swallowing, chewing, and phonation, and it plays an important role in facial growth, dentofacial deformities (Bommarito *et al.*, 2016), affecting the development of jaws and alignment of teeth (Kumar and Chaubey, 2007). It works mainly as an oral arch stabilizer and molder, and in the tongue's absence, the mandibular dental arch becomes atresic (Bommarito *et al.*, 2016). Distraction osteogenesis (DO) is a biological process of new bone formation between bone segments that are gradually separated by incremental traction (İşeri and Malkoç, 2005) by use of rigid external distraction (Sultan Olmez-Gurlen, 2016). This procedure was used in 1905 by Codivilla (Codivilla, 1994) and was later popularized by the clinical and research studies of Ilizarov (Ilizarov *et al.*, 1969) in Russia. Mandibular symphyseal distraction osteogenesis is an alternative approach to correct mandibular transverse discrepancies and it was first applied by Guerrero (İşeri and Malkoç, 2005). The aim of this study is to describe the use of a tooth-borne distraction device with a Hyrax-type screw as alternative procedure to expand the mandibular by bone-borne distraction device in a clinical case which falls under Type I B according Hall's Classification (1971) (Hall, 1971).

CASE PRESENTATION

A girl 9 years and 4 mouths reported with chief complaint of her difficulty to chew and she was diagnosed with congenital aglossia which was classified by Hall[9]as type I-B (Table 1) or Type I B1 according Kalaskar *et al.*, (2016) (Fig 1). The mother was 29 years at birth did not attend any prenatal consultation. The birth was normal but postnatal period was associated with suckling and swallowing problems. The first attempt at breastfeeding revealed the tongue abnormality and she was initially fed via nasogastric tube, but she rejected this after 2 months and subsequently had difficulty adjusting to bottle feeding. The mother gave a history of intake of some medication and reported to have contracted syphilis during pregnancy. She was unable to remember the names of the all medications, except Benzetacil® (Penicillin G Benzathine Intramuscular).Patient's family history for congenital abnormalities was negative and she was her third daughter. In the third year of life, her weight and height were below normative values due to difficulty swallowing. During the physical exam, the patient showed interposition of the lower lip between the anterior maxillary and lower teeth. In lateral view, the facial profile was strongly convex, with the severe eversion of the lower lip and regular nasolabial angle (Fig 2). The temporomandibular joint functioned normally. She had no abnormalities in the head, ears, eyes, nose, or any other part of the body. The intraoral examination shows completely absent of the tongue, poor periodontal status, proclined maxillary incisors, extremely deep hard palate, median palatal groove, severe maxillary and mandibular hypoplasia, and excessive horizontal overjet, probably due to lack of muscular stimulus between the alveolar arches. The maxillary canines, right first premolar, second premolars, the mandibular incisors, left canine, left first premolars and the second premolars were clinically absent (Fig 3-9).In addition, the patient exhibited hypertrophy of the muscles of the floor of the mouth (Fig 1). There was no oronasal communication (Fig 4).The uvula was hypertrophic and the oropharyngeal isthmus was constricted with absence of the palatoglossal arches and it was separate from oral cavity by a thin membrane with a central orifice that permitted feeding and phonation (Fig 1).

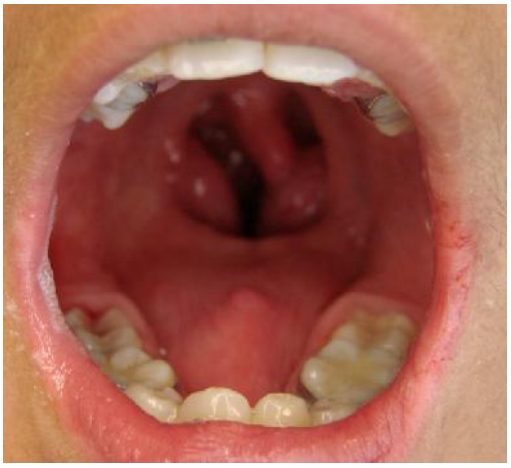

Fig. 1. Total absence of tongue and hypertrophic uvula intraoral photograph

Fig. 2. Pretreatment facial photographs

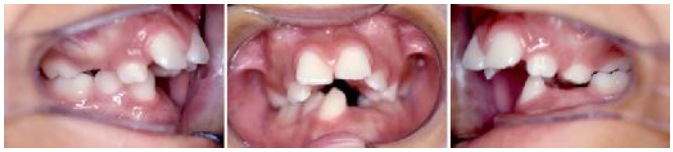

Fig. 3. Pretreatment intraoral photographs

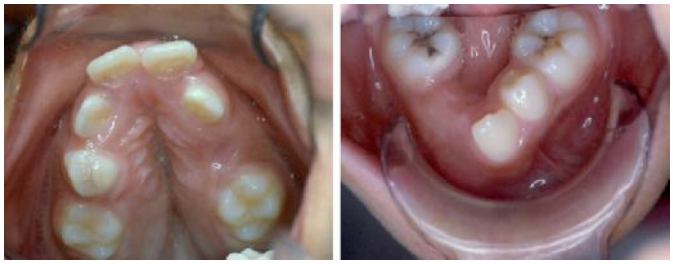

Fig. 4. Pretreatment intraoral photographs

The lateral cephalometric radiography shows a convex bone profile, skeletal Class II discrepancy, with maxillary retrusion in relation to the cranial base, mandible in a severely retruded position in relation to the cranial base, and the proclined maxillary incisors. In the vertical plane of space, all cephalometric measurements indicated a hyperdivergent facial pattern. The pharyngeal air space appeared narrow and the gonial angle was increased (Fig 10 and 11). The panoramic radiograph showed that the 4mandibular permanent incisors were congenitally missing (Fig 12). The case was monitored by the multi and interdisciplinary team, which assessed stomatognathic system functions: breathing, sucking, chewing, swallowing, and speech. The psychomotor development was slightly delayed: speaking at 16 months, and walking at 21 months of age. The speech therapist observed lip tonus and mobility, cheek, mouth floor muscles, and hard palate. Speech development is delayed and slurred. Although she could answer questions, an abnormal speech was characterized by omission of linguoalveolar and linguopalatal phonemes and an altered voice quality. Neurological assessments were below. Fiberoptic nasolaryngoscopy verified nasal cavity hypoplasia, normal pharyngeal orifices and normal tonsils.

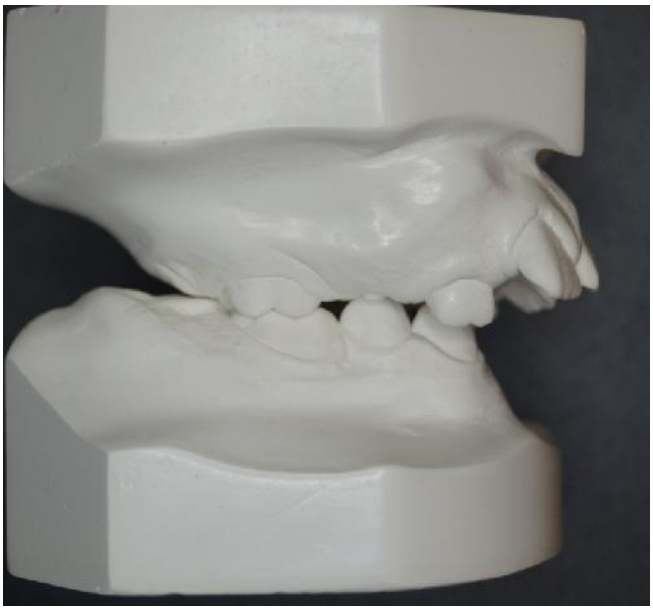

Fig. 5. Pretreatment models photographs

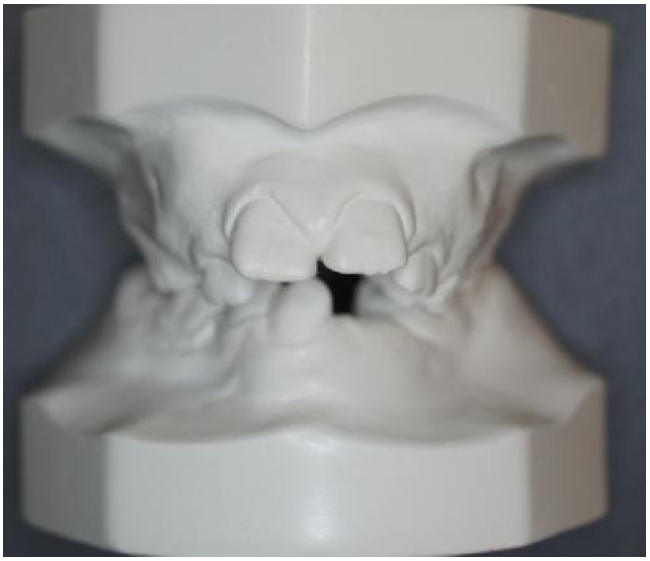

Fig. 6. Pretreatment models photographs

Treatment objectives

Objective of her orthodontic treatment included rapid maxillary expansion and mandibular midline DO to correct maxillary and mandibular transverse deficiencies to improve her quality of life and to provide conditions of the myofunctional adaptation may occur in the course of growth and development of craniofacial complex.

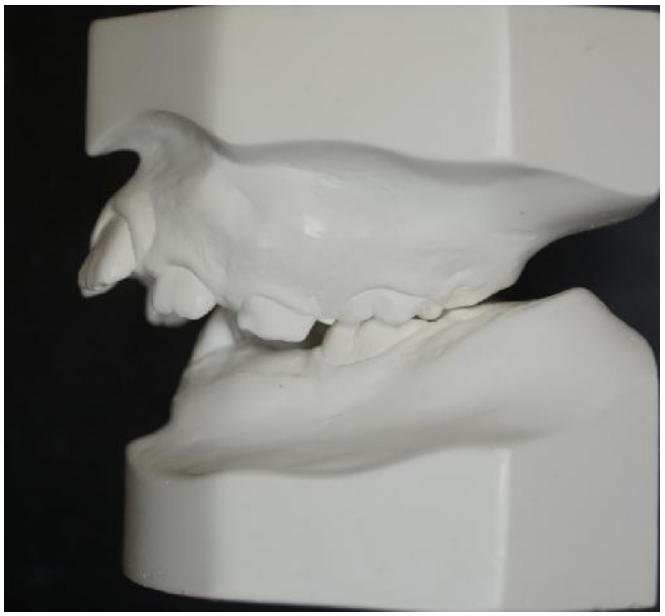

Fig. 7. Pretreatment models photographs

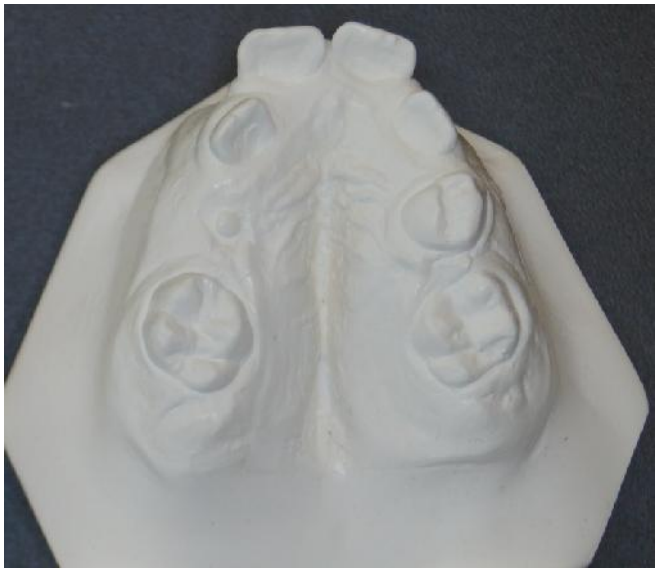

Fig. 8. Pretreatment models photographs

Treatment progress

The orthodontic treatmentwas conducted in two stages.

Phase I: rapid maxillary expansions with tooth-anchored device activated by means of a conventional Hyrax expander with a soldered framework and orthodontic bands on maxillary first premolars and first molars (Fig 13).After the expander was cemented, it was actived 8 turns. Then, her mother was instructed to activate the jackscrew 1 turn (0.25 mm) twice a day until the required expansion was achieved. The degree of expansion was calculated including a general bilateral overexpansion.

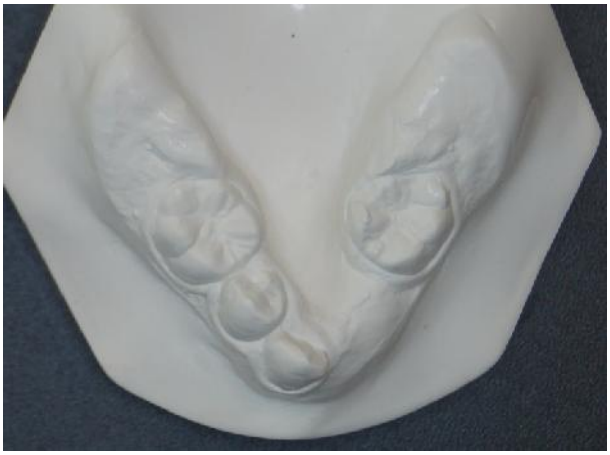

Fig. 9. Pretreatment models photographs

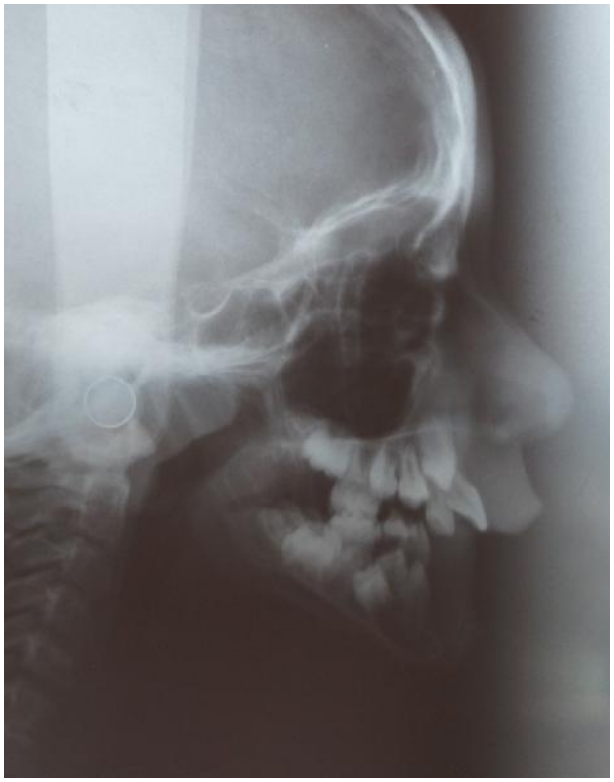

Fig. 10. Pretreatment lateral cephalometric radiograph

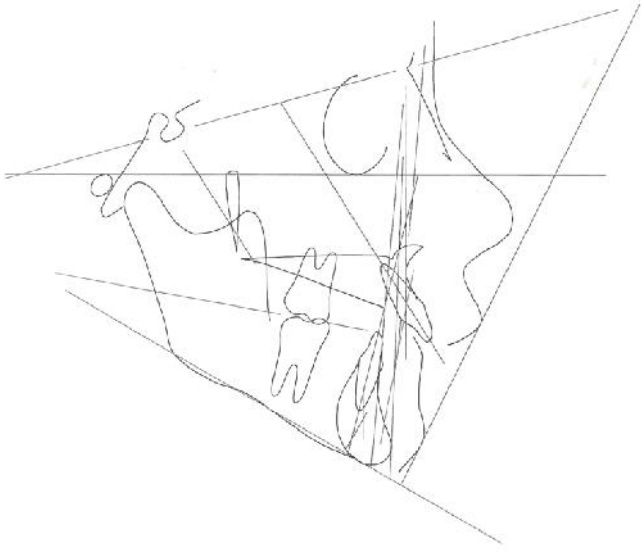

Fig. 11. Pretreatment cephalometric tracing

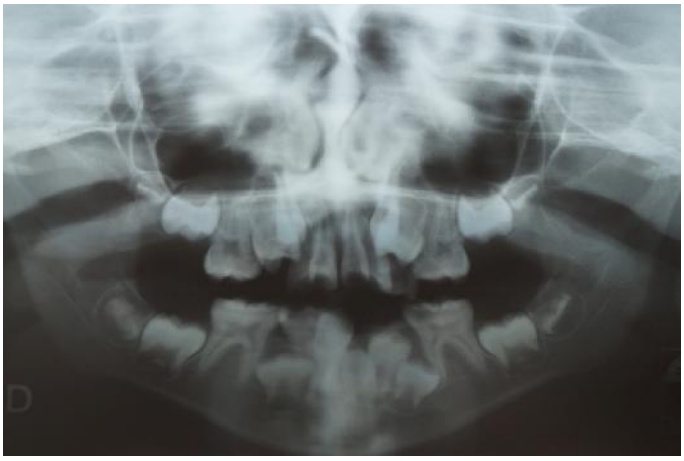

Fig. 12. Pretreatment panoramic radiograph

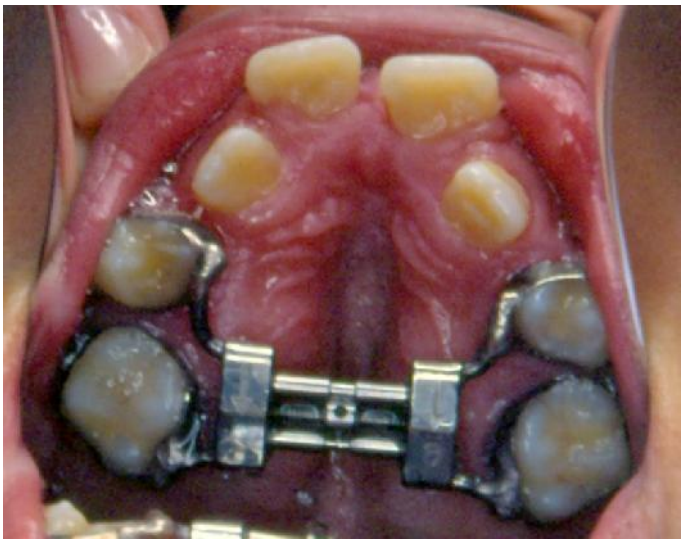

Fig. 13. Photograph of rapid maxillary expansion with Hyrax on phase 1.

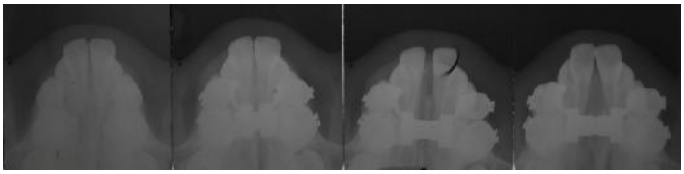

Fig. 14. Progress occlusal radiographs

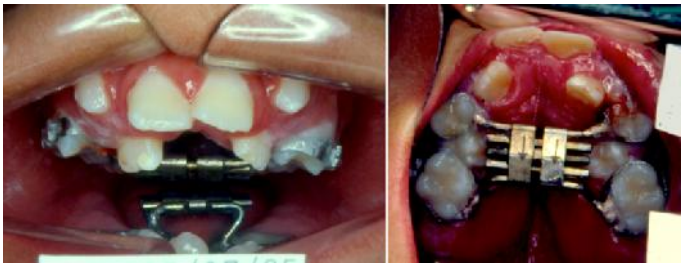

Fig. 15. Progress intraoral photographs of phase 1 and 2 with Hyrax appliance adaptation in lower jaw

After a mean active expansion period of 15 days, the expander was tied off with a ligature wire and it was kept on the teeth as a passive retainer for at least 90 days (3 months). This period of retention allowed for reorganization and reossification of the midpalatal suture after expansion (Fig 14). The amounts of transverse deficiency of the mandible and mandibular length

deficiency were also taken into consideration during the rapid palatal expansion (Fig 15).

Phase II: at the end of the active phase of the rapid maxillary expansion, a mandibular distraction osteogenesis was initiated by expansion with a tooth-borne distraction device with a Hyrax-type screw (Fig. 16). The device was supported by the permanent first molars due the difficulty with device insertion and an expansion screw was placed as anteriorly as possible to avoid impinging the gingiva and mucosa and to facilitate its activation with a key. The modificated mandibular distraction device was placed a day before surgery. An osteotomy was made in the mandibular symphysis and the screw was activated 8 times during surgery. Distraction was initiated 3 days after surgery (latency period to enable callus formation) and the screw was activated twice per day achieving a total distraction of 9 mm at the end of the 14 day distraction period. After this period, the Hyrax screw was stabilized by ligature wire (0,30mm) and the device remained in the mouth for a 90-day period. The lower anterior interdental bone level and lower canines tooth root positions were evaluated via panoramic radiographs before surgery. After new bone formation was radiographically evident (Fig. 17), the Hyrax appliance was remove and fixed orthodontic appliance treatment was initiated, and the leveling and aligning stage was started in both dental arches (Fig. 18). Once the growth phase is completed, the patient is aware that orthognatic surgery can be programmed to correct the anterior posterior discrepancy between the jaws.

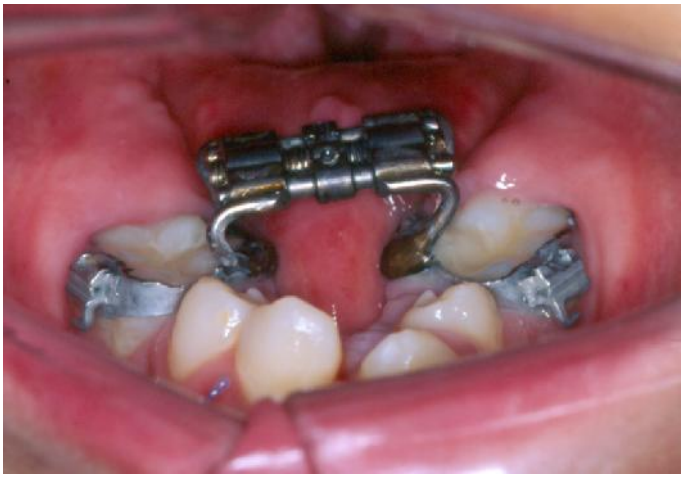

Fig. 16. Intraoral photographs of tooth-borne distraction device with a Hyrax-type screw

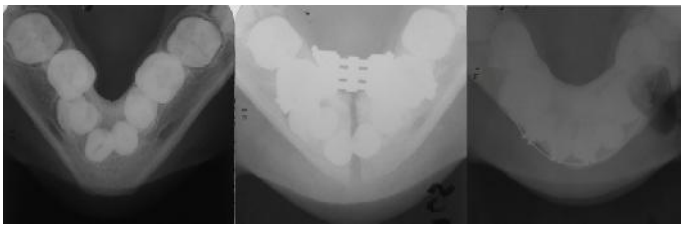

Fig. 17. Progress occlusal radiographs

Surgical procedure

Mandibular distraction osteogenesis was performed with the patient under general anesthesia. The incision was made through the mandibular labial and buccal mucosa and sulcus extending from premolar to premolar avoiding the mental nerve. A second incision was made through the submucosal tissue and periosteum (Fig. 19). After the muscle was transected, dissection was directed obliquely, poster inferiorly through the mentalis muscle until establishing contact with the mandibular symphysis. A full-thickness mucoperiosteal flap was elevated from medial to lateral. Soft tissues were detached from the planned osteotomy site at the crest of the alveolar ridge, and the symphysis was completely degloved, including the inferior border. The osteotomy was performed with a reciprocating sawstarting at the inferior border of the mandible and extending to the interdental space between the apices of the mandibular canines under saline irrigation (Fig. 20).

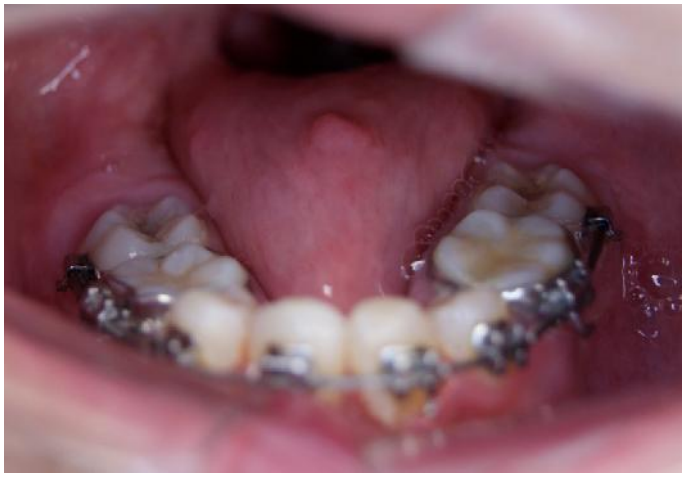

Fig. 18. Photograph illustrating dental leveling

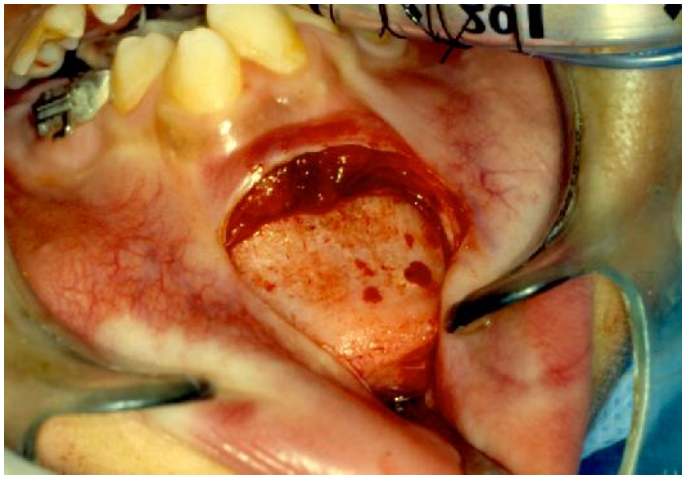

Fig. 19. Photograph illustrating surgery phase

Fig. 20. Photograph of reciprocating saw

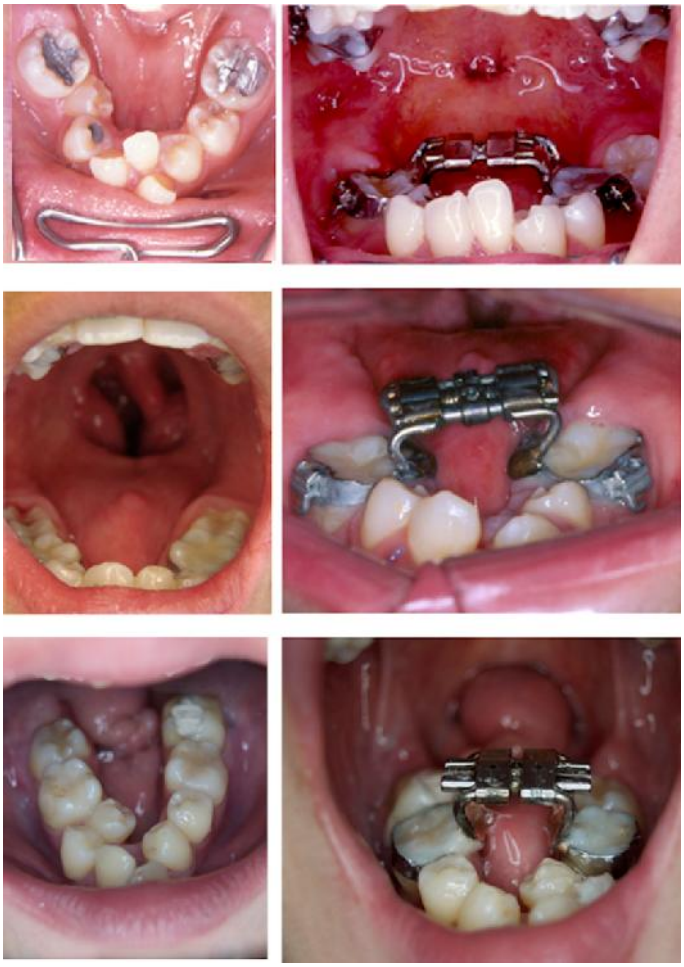

Fig. 21. Frontal close up view: intraoral initial photograph of lower arch and tooth-borne mandibular distractors with a Hyrax-type screw used in 3 clinical cases

The oscillating saw is safer and easier to use than a surgical bur and has the advantage of obtaining a more regular and thinner osteotomy line. At the final osteotomy stage, the screw of the mandibular expansion appliance was activated 8 turnsto assure that the interdental bone incision was complete and that the margins of the osteotomy site were separated. The submucosal tissue was sutured with interrupted 3/0 chromic sutures and the mucosa with interrupted 4/0 chromic sutures. The incision was closed with absorbable sutures, and an antibiotic and a nonsteroidal anti-inflammatory drug were prescribed for 5 days. After surgery, the chin was stabilized by a bandage to prevent muscle pull.

RESULTS

Maxillary and mandibular bones were successfully expanded and the osteogenesis of the distracted region was good. Proportional movement occurred in mandibular arch due to the screw position, which was placed as anteriorly as possible, and during the distraction phase, mandibular bone segments were expanded in parallel movement assessed from distraction sites on the right and left sides and bone formation was visible on serial occlusal radiographs that was taken at the beginning, before a week and immediately after the 90-day consolidation and retention period (Fig 17). Orthodontic treatment was initiated after observing radiographic evidence of bone healing by the lower and upper arch leveling and aligning.The stability of these treatments is often unpredictable, and the functional and esthetic rehabilitation of patients with congenital malformations is difficult, and their development depends on the involvement of a multidisciplinary team dedicated to full rehabilitation objectives to achieve good quality of life for patients and their families.

Table 1. Hall's Classification: Syndromes of Oromandibular and Limb Hypogenesis[6]

Type I	A) Hypoglossia
	B) Aglossia
Type II	A) Hypoglossia-Hypodactyly
	B) Hypoglossia-Hypomelia (Peromelia)
	C) Hypoglossia-Hypodactylomelia
Type III	A) GlossopalatineAnkylosis (AnkyloglossumSuperius Syndrome)
	B) With Hypoglossia
	C) With Hypoglossia-Hypodactyly
	D) With Hypoglossia-Hypomelia
	E) With Hypoglossia-Hypodactylomelia
Type IV	A) Intraoral bands and fusion
	B) With Hypoglossia
	C) With Hypoglossia-Hypodactyly
	D) With Hypoglossia-Hypomelia
	E) With Hypoglossia-Hypodactylomelia
Type V	A) Hanhart Syndrome
	B) Charlie M. Syndrome
	C) Pierre Robin Syndrome
	D) MoebiusSyndrome
	E) Amniotic Band Syndrome

Table 2. Modification of type I B Hall's Classification[1]

Hall's Classification	Modified subtypes	Clinical features	References
Type I B (aglossia)	Type I B.1	Isolated aglossia	Salles F et al[4], Gupta S[2].
	Type I B.2	Aglossiawithadactyli	Nevin NC et al[5], Purohit et al[11]
	Type I B.3	Aglossia with hypodactyli (mental retardation, cardiac defect, anodontia, hypothyroidism)	Preis et al[12], Kantapura and Tanpaiboon[3]
	Type I B.4	Aglossia with rudimentary ear (deafness)	Higashi and Endo[10], present case

DISCUSSION

There is considerable overlap between these syndromes gathered under the term OLHS, with a marked variability of face and limb anomalies as well as other additional malformations (Brockmann *et al.,* 2009), but severe micrognathia is observed in all the cases of OLHS, due to the osseous defect in the mandibular midline region, and occasionally involves the premaxilla (Meundi *et al.,* 2013). Aglossia is an extremely rare congenital malformation (Kalaskar *et al.,* 2016) that has been associated with jaw deformities (Gupta, 2012) since the muscular pressure contributes in development and shape of jaws and establishment of occlusion of teeth (Kumar and Chaubey, 2007). It also affects facial aesthetics, and hence psychological, sexual, and social development (Gupta, 2012). The association with difficulty in performing functions, especially suckling after birth, has mostly resulted in death of the newborn within 3 days (Salles *et al.,* 2008). Taste perceptions are not severely compromised, probably due to presence of taste buds in the mucosa of the floor of the mouth (Gupta, 2012). The chewing, swallowing, and speech adaptation is made possible by the hypertrophy of the muscles of the floor of the mouth, especially the mylohyoid (Nayyer *et al.,* 2015), due to constant stimulation during swallowing (Gupta, 2012) from early life. This helps the patient in elevating the floor of the mouth to touch the palate, which then mimics the absent tongue and helps in phonation, mastication, and swallowing (Gupta, 2012). The hypertrophied uvula helps in closing the oropharyngeal opening and forces the air through the nasal passage making the articulation of nasalized sounds possible (Gupta, 2012; Salles *et al.,* 2008) and the speech therapy improves phonetics (Tubbs *et al.,* 2016).

The adaptive mechanisms developed by these patients to compensate for absence of the tongue can be used in rehabilitation of patients who have acquired aglossia due to other causes like surgery for malignancy and trauma (Gupta, 2012). Few studies have discussed possible treatments of aglossia (Salles *et al.,* 2008) whose consequences involve several conditions that cannot be treated with a single procedure. Distraction osteogenesis is a process of growing new bone by mechanically stretching preexisting vascularized bone tissue (Nanjappa *et al.,* 2011) and the mandibular widening with DO is an alternative treatment modality (İşeri and Malkoç, 2005) for these patients with severe transverse mandibular deficiencies. The use of a tooth-borne distraction device with a Hyrax-type screw, in 3 clinical cases, as alternative procedure to expand the mandibular by symphysis osteogenesis shown in Figure 21. The desired mandibular change in shape can be achieved by selecting and controlling the force vectors during active distraction (Malkoç *et al.,* 2006). According Conley and Legan 2003 (Conley and Legan, 2003), the tooth movement into regenerate bone is recommended after a period of 3 months (Gökalp, 2006). Furthermore, Bell *et al.* 1997 reported in an animal study that intramembranous and endochondral bone formation was obtained by a 6 mm distraction, which was maintained in fixation for 60 days. Mandibular transverse deficiency generally becomes manifest with maxillary transverse deficiency and the RME is a common orthopedic procedure to treat maxillary transverse deficiency (Gökalp, 2016) in growing subjects undergoing immature skeletal development when the force applied to the teeth and maxilla exceeds the limits needed for tooth movement (Nakamato *et al.,* 2002). Aglossia sequelae involves a multidisciplinary approach with the participation of professionals in the areas of nutrition, psychology, speech and hearing, general dentistry, orthodontics, and maxillofacial surgery is needed to treat the problems of these patients (Salles *et al.,* 2008). Interceptive treatment with this type of mandibular expansion appliance associated with maxillary expansion offers new possibilities for arch development restoring their form and function that were severely deformed secondary to a lack of the normal stimulus provided by a growing tongue (Cappellette and Oliveira, 2013). Mandibular tooth-borne distractor is a practical and noninvasive clinical approach involving new bone formation between bone segments that are gradually separated by incremental traction. Proportional movement was obtained both in dentoalveolar and basal bone regions. However, the maintenance of the results achieved depends on the use of a retainer in a definitive manner.

Conclusion

The treatment of sequelae of aglossia is possible and the use of a tooth-borne distraction as alternative procedure to expand the mandibular by symphysis osteogenesis was efficient and improved the tongue tonus, phonetic, nutrition, chewing, and occlusion allowing a better development of adaptive mechanisms giving the patient a better quality of life. However, they are patients who require constant care with the participation of a multidisciplinary team due to lack of proper stomatognathic function.

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