Tessier number 30 clefts: surgical correction of a rare malformation

Fendas número 30 de Tessier: correção cirúrgica de uma rara malformação

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RESUMO

Objetivo: As fissuras medianas de lábio inferior e mandíbula (Fissura 30) são anomalias congênitas raras. Há uma ampla variação da gravidade desta malformação. Reavaliamos retrospectivamente pacientes que se submeteram à cirurgia, e propomos um protocolo de tratamento. **Método:** Este é um estudo retrospectivo de 17 pacientes com fissura 30 tratados entre 1997 e 2007. Dados clínicos, como idade, gênero, acometimento de tecidos moles e ósseo da linha média, complicações pós-natal imediatas e procedimentos cirúrgicos foram colhidos. Seguimento a longo prazo e resultados cirúrgicos foram avaliados. **Resultados:** A média de idade foi de 13 anos. O lábio inferior esteve envolvido em seis pacientes, anquiloglossia também em seis, incluindo dois casos que tinham língua bífida. Outros dois casos de língua bífida isoladas foram identificados, assim como dois casos de hipoplasia lingual. Envolvimento cervical foi encontrado em 3 pacientes, sendo um caso de fissura completa de região cervical anterior. Estes pacientes foram tratados com múltiplas plásticas em "Z". A falha óssea mandibular foi encontrada em 14 pacientes. Em cinco pacientes, foi diagnosticada a sequência de Pierre-Robin. Três casos tinham associação de deformidades de membros. Quatro necessitaram de tratamento precoce devido à dificuldade respiratória ou de deglutição. Distração osteogênica foi indicada em 2 pacientes. Reparo isolado da fissura labial foi realizado em 3 pacientes. Uma paciente foi submetida a osteotomia sagital de mandíbula com ressecção do enxerto ósseo prévio. **Conclusão:** Esta representa a maior série de fissura 30 já publicada, acrescendo 17 casos aos 69 anteriormente publicados, e propomos um protocolo de tratamento.

Descritores: Anormalidades craniofaciais. Anormalidades maxilomandibulares. Fenda labial. Mandíbula/anormalidades/cirurgia.

SUMMARY

Purpose: Median clefting deformities of the lower lip and mandible (cleft no. 30) are rare congenital anomalies. There is a broad variation in the severity of this deformity. We retrospectively analyzed patients who underwent surgery from which experience the authors propose a surgical protocol. Methods: This is a retrospective study of 17 patients operated for Tessier number 30 cleft between 1997 and 2007. Clinical data include age, gender, midline involvement of the soft tissue and bone, immediate postnatal complications, and surgical procedures. Long-term follow-up is presented and the outcome of the surgical procedures is evaluated. **Results:** The median age of patients was 13 years. Lower cleft lip was identified in 6 cases. Six patients had ankyloglossia, including 2 who had an associated bifid tongue. Two patients had an isolated bifid tongue, and 2 patients had tongue hypoplasia. The neck involvement was present in 3 patients. Complete cleavage of the neck was seen in only one patient. The cervical cleavage was treated using multiple Z-plasties. A mandible cleft was seen in 14 patients. Five patients also had Pierre Robin Sequence. Three cases had the association of limb malformations. Four patients required surgery in early age due the sleep apnea and feeding. Bone distraction was indicated to 2 patients. Two patients underwent median mandible bone grafting. The isolated cleft lip repair was performed in 3 cases. One patient underwent sagittal osteotomy of the mandible, associated with resection of previous bone grafting. **Conclusion:** This series represents the largest experience in the treatment of Tessier number 30 cleft. We added 17 patients to the 69 cases previously published, and proposed a new protocol to the treatment.

Descriptors: Craniofacial abnormalities. Jaw abnormalities. Cleft lip. Mandible/abnormalities/surgery.

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INTRODUCTION

Median cleft deformities of the lower lip and mandible are rare congenital anomalies. Couronné (1819)[1] was the first author to report this abnormality. The midline cleft of the lower lip was classified by Tessier[2] as a number 30 facial cleft. The role of race and sex remains unknown. Oostrom et al.[3] reviewed the literature and found a total of 65 reported cases, including three cases of their own. Few cases have been published since that time[4,5].

There is a broad variation in the severity of this deformity, ranging from a simple notch in the mucosa to complete cleavage of the lower lip involving the tongue, chin, mandible, neck, & manubrium sterni. The tongue involvement may range from a bifid anterior tip with ankyloglossia and adhesion to the mandibular cleft margins, to tongue hypoplasia. The incisor teeth are frequently missing at the mesial mandibular margins. The hyoid bone and thyroid cartilage may be cleaved or completely absent. Thyroglossal tissue may be found in the surgical specimen of the fissura colli medialis[6]. Other malformations may be present, these include: congenital heart deformities, cleft palate, as well as face[5], hand and foot anomalies.

Embryologically, the mandible develops from the cartilage of the first branchial arch, the mandibular process of which is known as Meckel's cartilage. This occurs through ossification of an osteogenic membrane formed from ectomesenchymal fusion at 36-38 days of development. In the mental region, one or two small cartilages appear and endocondral ossification commences in the 7th month in utero forming numerous mental ossicles. The calcification stops at approximately one year of age. Most authors believe clefting results from a failure of fusion of the first pair of branchial arches, or a failure of mesodermal penetration into the midline structures. Monroe believed that the failure of fusion of the first branchial arch may be followed by failure of fusion of all the lower arches[6]. However, Oostrom et al.[3] hypothesized that only one first branchial arch develops during the early embryonic period, and two mandibular processes develop from within this structure. These mandibular processes do not fuse but merge during the late embryological period (after 7 weeks of development). In the same period, there is formation of the lower lip and the alveolar process with enlargement and outgrowth of the osteogenic membrane in each mandibular process, resulting in the formation of the mandible and its symphysis.

Proposed descriptions of the pathogenesis of median clefts of the lower lip and mandible include: A) Hypoplasia of the mandibular processes during the early embryonic period leading to severe clefting of the mandible extending into the neck; or B) during the late embryonic period, resulting in less severe median clefts. The lower cleft lip, ranging from a notch of the mucosa to a complete cleft, can be explained by insufficient outgrowth of the lip, most probably due to incomplete merging of the mandibular processes. The bifid tongue can be explained as a persistent intermandibular groove, similar to a merging defect of the tongue. The mandibular cleft is explained as impairment of the outgrowth of the osteogenic centers of the definitive mandible, resulting in absence of its symphysis.

A retrospective analysis of this series of patients with Tessier number 30 clefts was performed. The clinical findings regarding: the lip, teeth, alveolus, mandible, cervical clefts, associated deformities, and surgical procedures were reviewed. We also propose a surgical protocol to approach these cases.

METHODS

This is a retrospective study of 17 patients who underwent surgery for correction of Tessier number 30 clefts over a ten year period (1997–2007). Age, gender, midline involvement of the soft tissue and bone, immediate post natal complications, surgical procedures, and post operative course was analyzed. Long term follow-up was included to evaluate the outcome of the surgical procedures.

RESULTS

Seventeen patients with Tessier number 30 clefts were treated by the Assistance Center for Cleft Lip and Palate (CAIF) in Curitiba, Brazil, as well as the Plastic Surgery Section of São Paulo University, in Sao Paulo Brazil. The median age of the patients at initial presentation was 13 years (range 2 days to 30 years of age) (Table 1). The average treatment period lasted 4 years, with a range of 1 month to 12 years. There was one mortality in the series including a 3 month old, who died 1 month after surgery, from aspiration pneumonia (Table 2).

Lower lip clefts were identified in 6 cases: 2 complete clefts, 2 incomplete and 2 with only a small notch in the vermillion. Six patients had ankyloglossia, including 2 who had an associated bifid tongue. Two patients had an isolated bifid tongue. Two patients had tongue hypoplasia. Four patients had a double lower frenulum and 1 paramedian lower frenulum was also noted. Neck involvement was present in 3 patients with fissura colli medialis. A complete cleavage of the neck was seen in only one patient.

Mandibular clefts were seen in 14 patients, five of which had sleep apnea or feeding difficulties after birth. These were considered to represent Pierre Robin Sequence (micrognatia, glossoptosis and airway obstruction). Three patients had associated median upper cleft lip, and 1 patient had a bilateral upper alveolar cleft (in the same position as a common bilateral cleft lip, however without upper lip soft tissue involvement). Limb malformations were present in three cases, these were classified as either Richieri-Costa Syndrome or Hanhart Syndrome (Table 1).

It was necessary to perform surgery at an early age in four patients, secondary to sleep apnea and feeding. When distraction osteogenesis was possible, it was used to achieve better position of the tongue base in 2 patients. The other 2 patients were submitted to median mandible bone grafting to ensure the continuity of the mandibular arch.

Isolated cleft lip repair was performed in 3 cases. Z-plasty was utilized in the mucosa, followed by muscle repair and a straight line closure for the skin. Orthodontic wase used in 7 patients to align the teeth and prepare for orthognatic surgery in 4 cases, or to improve tooth implant osteointegration in 3 cases. An 18 year old patient underwent sagittal osteotomy of the mandible and resection of previous bone grafting to improve occlusion and the patients profile (Figure 1).

Table 1 - Clinical findings in Tessier number 30 cleft.

Case	Age	Gender	Lip	Frenulum	Mandible	Tongue	Teeth	Neck	Chest Wall	Other
1	5	M	notch	paramedian	no	no	no	no	no	no
2	26	F	no	adhesion tongue-lip	clefted	bifid	absence 11,12,21	no	no	upper alveolar cleft
3	25	F	cleft	double	clefted	bifid	absence 11,21	fissura colli medialis	no	no
4	30	M	cleft	double	no	no	no	no	no	no
5	0	M	cleft	double	clefted	bifid	no evaluated	fissura colli medialis	no	no
6	25	F	cleft	adhesion tongue-lip	clefted	bifid	absence 11,21	fissura colli medialis	clefted	no
7	6	M	no	adhesion tongue-lip	partial clefted	no	absence 11,12,21, 22	no	no	no
8	1	M	no	adhesion tongue-lip	clefted	no	no evaluated	no	no	Richieri Costa syndrome
9	0	M	no	adhesion tongue-lip	clefted	no	no evaluated	no	no	limb deformities (Hanhart)
10	0	F	no	adhesion tongue-lip	clefted	no	no evaluated	no	no	no
11	13	M	no	no	clefted	no	absence 11	no	no	median upper cleft lip
12	13	F	no	no	clefted	no	absence 21	no	no	median upper cleft lip
13	20	M	no	no	clefted	no	absence 11,12,21, 22	no	no	median upper cleft lip
14	8	M	no	no	clefted	hypoplasia	absence 11,12,21	no	no	no
15	8	M	no	no	partial clefted	hypoplasia	no	no	no	Richieri Costa syndrome
16	24	F	no	no	partial clefted	no	absence 11,21	no	no	no
17	17	M	notch	double	no	no	no	no	no	no

Figure 1 - Patient with complete cleft involving lower lip, mandible, chin, and the cervical region, accompanied with the lack of hyoid bone, thyroid cartilage, and manubrium sterni. (**A**) Photograph at 2 month of age, before the thoracic, cervical and lip repair in the first stage and costal graft in a secondary procedure performed in another institution. (**B**) Patient at 15 years of age, with median scar and previous double cervical Z-plasties, during bilateral bidirectional mandible distraction. (**C** and **D**) Patient at 23 years of age, during sagital split mandible osteotomy, showing a gap between the incisors and resection of the costal graft. (**E** and **F**) Two-year follow-up after orthognatic surgery.

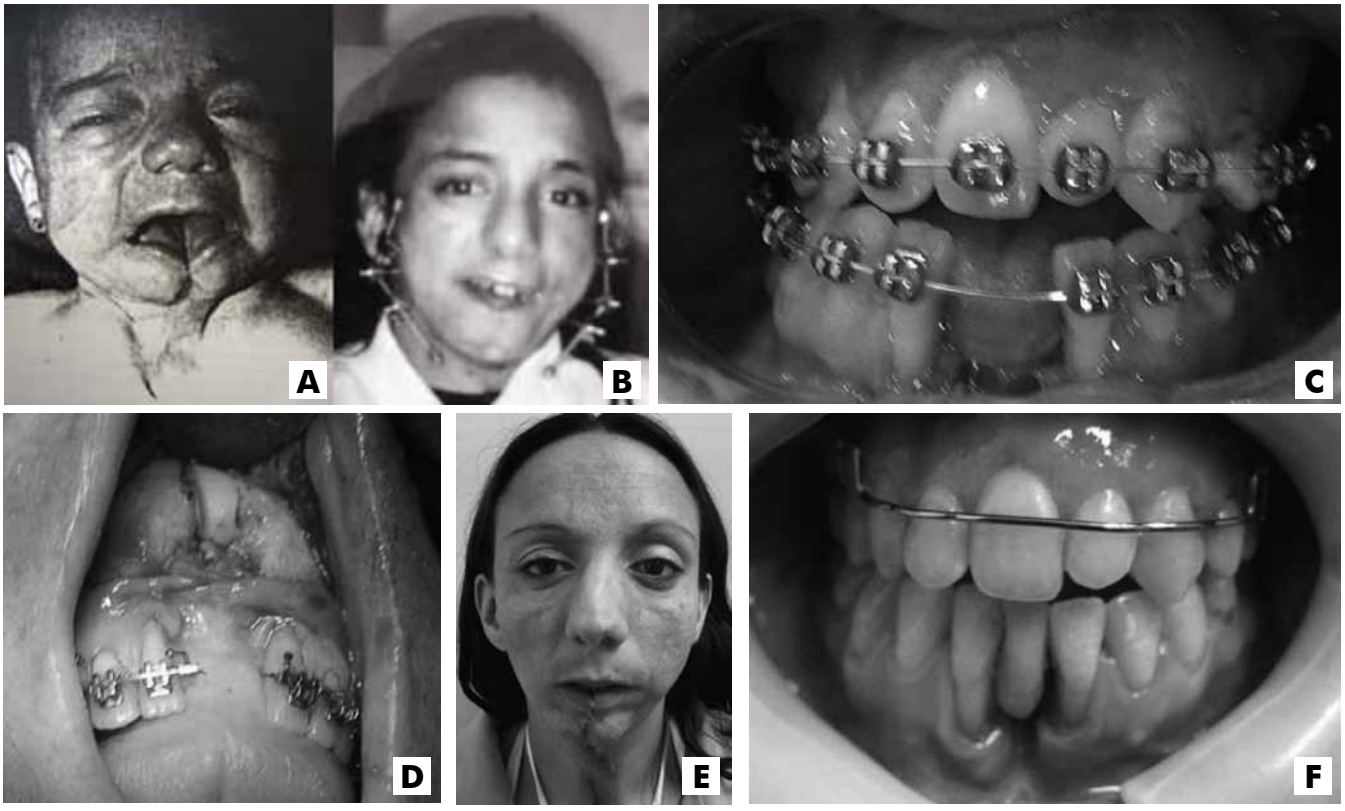

Table 2 - Description of the treatment and the outcome.

Case	Surgery	Orthodontia	Follow-up (years)
1	no	no	5
2	no	yes	10
3	no	no	0
4	lip repair	no	1
5	lip repair	no	1
6	lip repair+ Zplasty + bone graft + distraction + orthognatic	yes	12
7	distraction + tongue release	no	6
8	bone graft	no	1
9	no	no	0
10	distraction	no	died
11	no	yes	6
12	no	yes	10
13	no	yes	4
14	tongue release	Yes	3
15	no	no	1
16	no	Yes	4
17	lip repair	No	1

DISCUSSION

Midline clefts of the lower lip are rare malformations usually associated with clefts of the mandible. No hereditary factors or gender predilection has been identified. Clinical presentation varies from a mild notching of the lower lip to complete midline cleavage of the inferior face, including bifurcation of the mandible, tongue and neck. In this series, we present 3 cases with major neck involvement, including *fissura colli medialis*. However, the majority of cases exhibit only partial clefts of the lip and/or mandible.

Historically, treatment of this anomaly has not varied greatly. Most authors have released the tongue and closed the lower lip in the early stages of life in the majority of patients[4,7]. This is a logical approach, since the lip and tongue are necessary to suck, swallow and phonate. Most studies report a simple "V" excision of the cleft with direct closure of the lip and the use of a Z-plasty if the cleft extends into the neck[6,8]. Millard et al.[7] proposed a Z-plasty of the released lingual frenulum, and a midline incision vertically to the buccal sulcus. The fibrotic union of the muscle is excised except in the normal chin dimple position, and the muscle is sutured together across the cleft. The excess mucosa and vermilion is trimmed. The mento-sternal band should be resected and multiple Z-plasties performed in the skin and platisma, aiming to obtain a good contour of the cervico-mental angle[9].

The ideal timing for treatment of the mandibular cleft remains undetermined[4]. Millard et al.[10] reported a mandible repair using a rib graft, wiring and vitallium splint at 8 years of age. In patients without respiratory disturbance, the mandibular defect closure is frequently delayed until 10-12 years of age, in order to prevent damage of tooth buds during mandibular surgery[4,10]. However, careful unicortical osteosynthesis of the lower border of the mandible may prevent harm to the tooth buds if surgery is performed in the early period[3,11]. Sherman and Goulian[12] treated median mandibular bone defects at 20 months of age successfully. Moreover, osteosynthesis may be advantageous for mandible development, resulting in important functional improvements in speech, and eating and improved occlusion. The technique consists of subperiosteal dissection through an incision in the lower buccal sulcus, preserving the continuity of the mucosa. The exposed bone ends are freshened with a burr, and a bone graft is used to reconstruct the defect. The donor material may be rib, calvarium, or iliac crest. Screws, miniplates or wire may be used to fix the graft into position A one month old patient in this series had extrusion of a fixation plate 3 months after surgery, possibly due to very small mandible segments or lack of oral soft tissue for coverage. Ichii et al.[4] performed bone cleft reconstruction in 2 stages: a mandibular fixation at 6 years of age and an iliac graft 1 year after. We prefer single stage reconstruction at an early age, avoiding multiple procedures and reaping the benefits of improved speech eating and occlusion early on.

Our surgical protocol is as follows, in patients without respiratory distress or feeding problems, the continuity of the mandible is assessed. If the mandible is contiguous, distraction may be used for contour improvement. If the mandible is not contiguous, we recommend bone grafting with a costal graft at an early age under 2 years of age. Soft tissue deformities of the lower lip tongue and neck should also be corrected at an early age (3 months) as indicated.

Patients with Pierre Robin Sequence associated with mandibular clefts should be approached differently. If the continuity of the mandible is preserved, a nasopharyngeal airway may be utilized initially until better neuromuscular development occurs. If this conservative approach is inadequate, mandibular distraction may be indicated. In one case, we performed bilateral mandible distraction, which resolved the respiratory distress 24 hours after the procedure. Another patient underwent bilateral mandibular distraction, but this surgery was complicated by aspiration pneumonia resulting in death 30 days after the procedure. Patients with cleft mandible may undergo mandibular bone grafting with a costal graft. If respiratory distress continues, mandibular distraction may be considered. Two patients underwent costal bone grafting, neither required early distraction. One of them underwent distraction at age 13 years, then, 7 years later, resection of the bone graft, midline fixation using miniplates, and sagittal split osteotomy of the mandible. This was indicated to correct under bite and improve the profile, after a long orthodontic treatment.

CONCLUSION

In summary, this series represents the largest experience in the treatment of the Tessier number 30 cleft. 17 new patients are added to the 68 cases previously published. A new therapeutic protocol, which uses sleep apnea and the presence of a complete cleft of the mandible to dictate treatment is presented. Patients with Pierre Robin Sequence associated with mandibular clefts should undergo mandibular bone grafting; and, if the respiratory distress does not resolve, distraction should be considered. For patients with preservation of the mandible, a nasopharyngeal airway should be utilized initially, and mandibular distraction considered if respiratory distress does not resolve.

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