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To the Lip and Beyond: A Case Report of a Midline Tessier 30 Cleft

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ABSTRACT

Objective: To present the case of a midline Tessier 30 cleft in a baby boy who initially underwent a glossoplasty, cheiloplasty and mentoplasty.

Methods:

Design: Case Report
Setting: Tertiary Government Training Hospital
Patient: One

Result: A 4-month-old boy with a complete midline cleft of the lower lip, alveolus and mandible, and bifid distal tongue that was fused with the floor of the mouth, underwent glossoplasty, cheiloplasty and mentoplasty with subsequent excellent aesthetic outcome and normal oral competency.

Conclusion: Tessier 30 is a rare congenital midline mandibular cleft. Prompt glossoplasty, cheiloplasty and mentoplasty can correct the gross deformity, restore oral competency, and address functional needs such as feeding, swallowing and early speech development. Future bony repair will hopefully complete the reconstruction.

Keywords: *mandible; cleft lip; tongue diseases; jaw abnormalities; reconstructive surgical procedures; Tessier cleft; craniofacial cleft; median mandibular cleft*

Tessier clefts, otherwise known as craniofacial clefts, are extremely rare congenital malformations with an incidence between 1.4 and 4.9 per 100,000 live births, and have adverse functional, psychological, and aesthetic effects on patient's life.[1] In 1976, the French plastic surgeon Sir Paul Tessier classified a nonunion of the bony mandibular symphysis, adherence of the tongue to the midline floor of mouth and median clefting of the soft tissues of the lower lip as principle features of one subtype, the median mandibular cleft (designated Tessier 30).[2] Couronne in 1819 was the first to document an actual case and there are only 69 cases reported worldwide to date.[3] To the best of our knowledge based on a search of HERDIN Plus, the ASEAN Citation Index, the Global Index Medicus - Western Pacific Region Index Medicus

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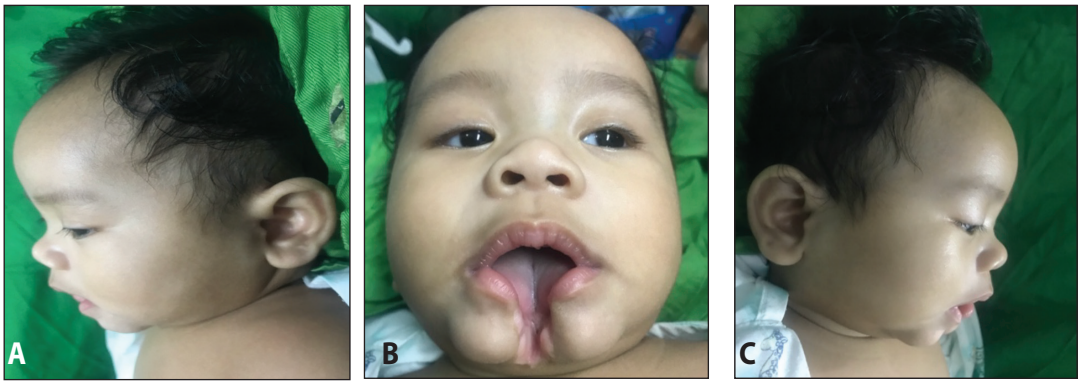

Figure 1. Complete mandibular cleft described as a notch extending from the skin of the lower lip up to the level of mandible. **A.** left lateral; **B.** frontal, and **C**. right lateral views. (photos published in full, with permission)

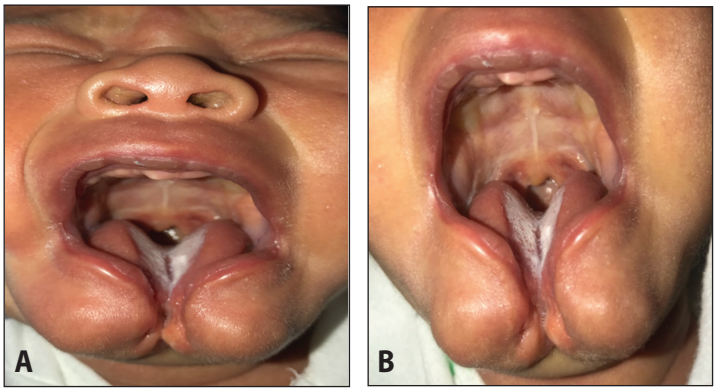

Figure 2. A, B. Oral cavity views revealing bifid tip of the tongue and ankyloglossia. (photos published in full, with permission)

and Index Medicus of the South East Asia Region, there have been no previously published reports in the Philippines. We report what may possibly be the first such case in our country, and the results of our initial soft tissue repair.

CASE REPORT

A 4-month-old boy was referred to our outpatient department due to a complete lower lip and mandibular cleft with a bifid tip of the tongue and ankyloglossia. He was born to a 32-year-old gravida 4 para 4 (4103) mother, delivered via Ceasarean section due to failure of fetal descent at a private hospital in Angeles City, Pampanga. No fetal or maternal complications were noted during delivery. The prenatal history was generally unremarkable, with no exposure to alcohol, radiation, or smoking during pregnancy.

However, the mother had experienced abnormal uterine bleeding from the start of the 2nd up to the 5th month of gestation and was maintained on isoxsuprine HCl (duvadilan) and dydrogesterone (duphaston). Moreover, she also had an upper respiratory tract infection during the third month of gestation and was treated with amoxicillin - clavulanate (co-amoxiclav) and carbocisteine (solmux). The rest of the history was non-contributory, and there was no family history of clefting in both maternal and paternal sides.

Physical examination revealed a complete mandibular cleft extending from the skin of the lower lip up to the level of mandible, sparing the upper lip, nose, maxilla, rest of the face and neck. *(Figure 1A-C)* Further examination of the oral cavity revealed a bifid tip of the tongue and ankyloglossia. *(Figure 2A, B)*

The facial computed tomography (CT) scan confirmed a median mandibular cleft with widely separated symphysis menti and midline soft tissue defects of the lower lip, anterior portion of the oral tongue (at the frenulum) and subjacent floor of the mouth. The hyoid cartilage, base of tongue, geniohyoid-genioglossus muscle complex, thyroid gland, maxilla, hard palate and remainder of the oropharynx and larynx were intact. The nasal cavities were patent without evidence of choanal atresia. The orbital structures, ear structures and visualized intracranial structures were also intact. The rest of the maxillofacial structures, suprahyoid neck and visualized infrahyoid neck structures were also unaffected. *(Figure 3A-C)*

Figure 4A illustrates the preoperative extent of the Tessier 30 defect: cleft tongue with bilateral adhesion/ankyloglossia to the floor of the

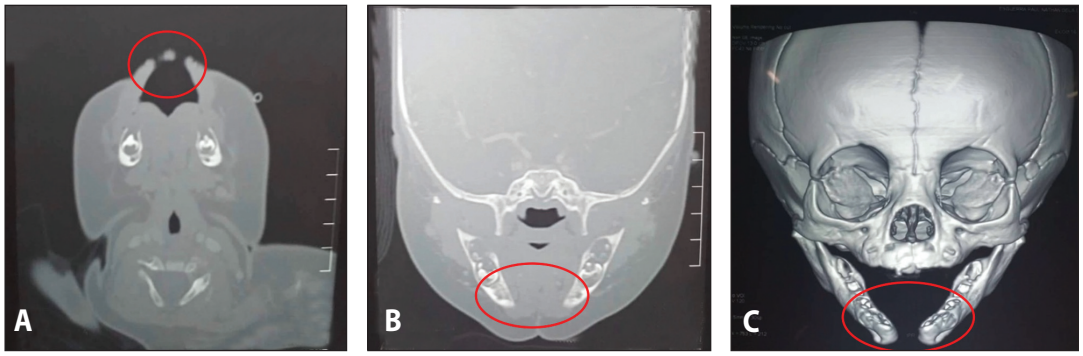

Figure 3. CT scan images **A.** axial view, **B.** coronal view, and **C.** 3D Reconstruction images showing (encircled) median mandibular cleft with widely separated symphysis menti and midline soft tissue defects of the lower lip, anterior portion of the oral tongue (at the frenulum) and subjacent floor of the mouth.

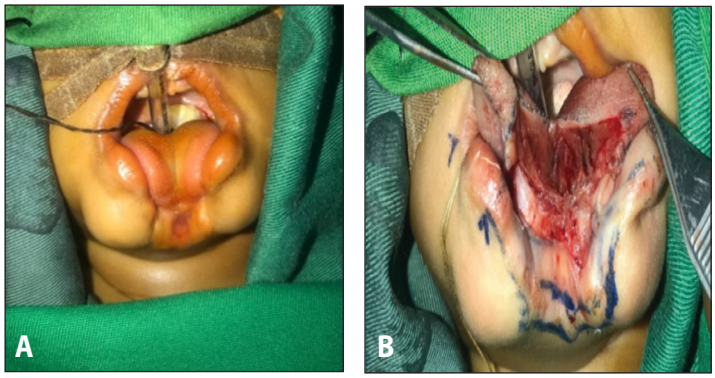

Figure 4. A. Components of Tessier 30 cleft as mentioned with the following defects; cleft tongue with bilateral adhesion/ ankyloglossia to the floor of the mouth on both sides, lower lip clefting, absence of mandibular symphysis and parasymphysis. **B.** Tongue reconstruction with initial excision of midline tongue mucosa, identification of the intrinsic muscles of tongue, release of adhesions from the tip of the tongue on both sides to the floor of the mouth.

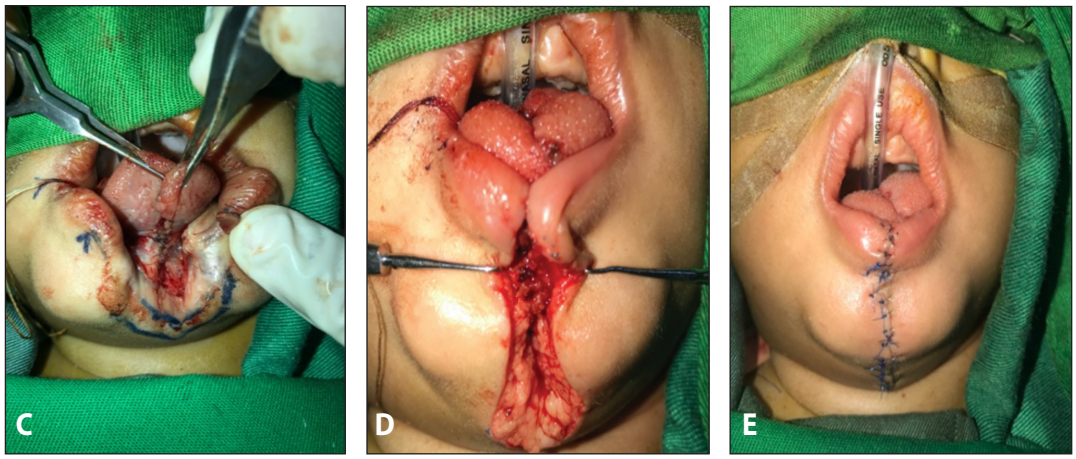

Figure 4. C. Detachment of the tip of the tongue on both sides of the floor of mouth. Mobility was noted, with coaptation of the intrinsic muscles and tongue mucosa **D.** Excision of redundant skin from the lower lip to the submental cleft leaving the non fused bony ends of the mandibular body intact for future reconstruction. **E.** Approximation and coaptation of the repaired tongue, floor of mouth, lower lip and mentum with vicryl and nylon sutures and Z-plasty. (photos published in full, with permission)

mouth on both sides, lower lip clefting, and absence of mandibular symphysis and parasymphysis. We performed a glossoplasty, lower lip cheiloplasty and soft - tissue mentoplasty.

Tongue reconstruction began with initial excision of midline tongue mucosa, identification of the intrinsic tongue muscles, and release of adhesions from the tip of the tongue on both sides to the floor of the mouth. *(Figure 4B)* The tip of the tongue was detached on both sides from the floor of mouth. Tongue mobility was noted with coaptation of the intrinsic muscles and tongue mucosa. *(Figure 4C)* We then excised redundant skin from the lower lip to the submental cleft leaving both non-fused ends of the mandibular body intact for future mandibular reconstruction. *(Figure 4D)* Finally, we approximated and coaptated the repaired tongue, floor of mouth, lower lip and mentum using vicryl and nylon sutures. A Z-plasty was also utilized to lengthen the volume over

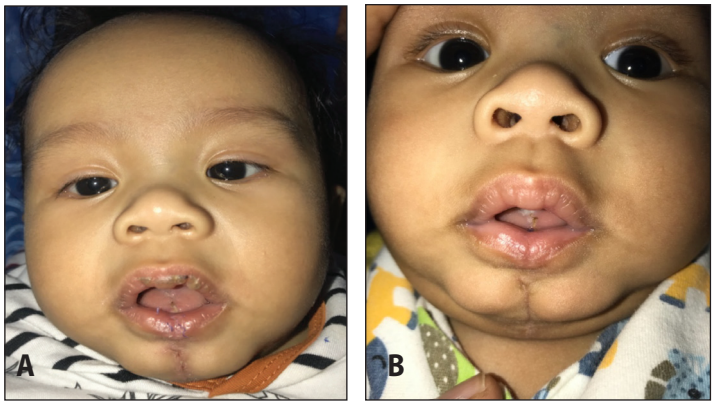

Figure 5. A. One (1) week post operation; and **B.** Two (2) weeks post operation. (photos published in full, with permission)

Figure 6. A. 5 months post operation (age 9 months); **B.** well-healed tongue, floor of mouth, and lip. The baby was thriving with good suckling and swallowing and beginning phonation. (photos published in full, with permission)

the mentum area for adequate closure and better aesthetics. *(Figure 4E)*

There were no postoperative complications. After 2 weeks, there was excellent healing with establishment of oral competence *(Figure 5A, B)* This facilitated suckling and swallowing and bottle feeding was initiated. The child continued to thrive, with beginning phonation after 5 months. *(Figure 6A, B)*

DISCUSSION

A Tessier 30 cleft, also known as median mandibular cleft, is one of the rarest craniofacial anomalies, with no race predisposition, gender predilection or hereditary factors identified.[3] Clinical features and severity of the malformation differ from case to case, ranging from a midline notch to complete clefting of the lower lip with or without bony involvement ranging from notching of the mandible to complete clefting of the mandibular symphysis.[4] Ankyloglossia, fibrotic band in the neck, bifid or total absence of tongue, absence of hyoid bone, hypoplasia of the thyroid cartilage, strap muscles of the neck with bifid or absent manubrium sterni may also be present.[4] Other malformations that may also be present include congenital heart deformities, cleft palate, and face, hand and foot anomalies.[3] Majority of the cases exhibit only partial clefts of the lip and mandible[3] but our patient had clefting of the tongue with ankyloglossia, clefting of the lower lip, and absence of the mandibular symphysis and parasymphysis.

There are several theories on the pathogenesis of Tessier 30 cleft. Embryonically, the mandible develops from the cartilage of the first branchial arches, hence, developmental anomalies of these may result to the failure of neural crest cell development along the normal fusion planes[5] causing hypoplasia of mandibular processes[3] and disruption of mesodermal penetration into the midline mandible.[5] Exposure to teratogenic agents such as retinoic acid during pregnancy have also been implicated, resulting in binding of excessive retinoic acids to retinoic acid-receptors that disrupts the expression of Hox genes responsible for the ability of the cells to direct neural crest cell migration.[6] Another theory suggested that disruption of the SPECC1L gene that is responsible for normal chondrocyte assemblage and molecular signaling processes leads to the inhibition of frontonasal and maxillary process fusion and mandibular prominence convergence.[7] None of these proposed theories could be confirmed in our patient. However, the upper respiratory infection during pregnancy may have contributed to the developmental anomaly in our patient, as specific antibodies against viruses such as influenza may play a role in the development of orofacial clefts.[8]

Soft tissue reconstructive surgery should ideally be performed during the third month of life[3] to restore oral competence and facilitate normal feeding, deglutition and phonation. A Tessier 30 cleft can be treated with one- or two- stage procedures depending on the size of the defect.[9] A one-stage procedure involves simultaneous repair of the soft tissue and bony mandibular defects. Sheckter *et al.* successfully repaired a Tessier 30 cleft with a modified single-stage surgery by infiltrating the non-fused ends of the mandible with human recombinant bone morphogenic protein to facilitate bone growth, avoiding bone graft harvest and allowed simultaneous treatment.[10] A two-stage procedure, on the other hand, involves an early life soft tissue repair with delayed bony mandibular reconstruction. Most of the cases reviewed by Freitas *et al*. involved a two-staged surgery and the techniques did not vary significantly; the general approach to treatment included a release of the tongue, excision of the clefts and primary closure of the defects.[3] Duplication of the tongue can be corrected using a wedge excision or Z-plasty[11] and this Z-plasty technique can also be utilized if the defect extended on the neck.[3] In our case, glossoplasty, cheiloplasty and mentoplasty served as the first stage of reconstruction in anticipation to the functional deficits caused by a Tessier 30 cleft.

Second stage surgery involves mandibular bony reconstruction. Bone grafts harvested from the costa, calvarium or iliac crest are commonly used in the reconstruction[3,5,12] in conjunction with

interosseous wiring or absorbable stainless plates to correct the bony defect.[5,12] Most surgeons begin bony reconstruction at 10 years of age[12] (and this can be delayed up to 12 years in order to prevent mandibular hypoplasia[5] and damage from the developing tooth buds)[3,5,12] although Oostrom *et al*. claim that a carefully planned osteosynthesis at the base of the mandible may prevent harm to the tooth buds, allowing earlier repair of the bony defect and providing better early occlusion.[5]

As of this writing, our patient has good oral competence, is able to feed orally, has full tongue mobility with no swallowing difficulties and is able to speak simple words such as "mama" and "papa." The boy is also at par for age in terms of growth and development. We initially considered stem cell therapy for the repair of the bony mandibular defect, as stem cells have been used in craniofacial reconstruction, often combined with synthetic polymer derived scaffoldings to promote bone growth.[13] However, we reconsidered this option in favor of planning an iliac graft with interosseous wiring or resorbable plates, because stem cell therapy is costly, the cell harvest is invasive, and materials are not readily available locally.[13]

In conclusion, a Tessier 30 cleft is a rare congenital midline mandibular cleft that can be treated successfully with timely glossoplasty, cheiloplasty and mentoplasty. Meticulous coaptation of the muscle layers and adequate release of tongue adhesions provide excellent restoration of oral competence, functional outcomes and ensure better aesthetics. Bony mandibular defect repair may be performed later as a second stage.

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