



Laurice Ann B. Canta, MD

Department of Otorhinolaryngology
Head and Neck Surgery
East Avenue Medical Center

Cleft Beyond the Lip and Palate: A Bilateral Tessier Cleft

Oblique facial clefts (Tessier Clefts) are severe orofacial clefts beyond the lip and palate that count among the rare congenital malformations “with an incidence of 1.43 to 4.85 per 100,000 births.”^{1,2} The Tessier Classification System devised by Paul Tessier in 1976 assigned specific caudo-cranial numbers to clefts involving the “soft tissue and underlying bones of the mouth, maxilla, nose, eyes and forehead” in relation to the sagittal midline of the face.^{1,3,4} The midline is designated 0 – 14, and adjacent clefts are numbered 1-13, 2-12 and so on, depending on the location and axis of the malformation. We present a patient with a bilateral Tessier 5 cleft.

CASE REPORT

A 4-month-old baby boy was referred to our outpatient clinic due to facial deformity. He had been born full-term via normal spontaneous vaginal delivery to an 18-year-old G1P0 mother, assisted by a mid-wife at a lying-in clinic with no fetal or maternal complications, and no history of trauma or prenatal drug intake (except for amoxicillin for urinary tract infection during pregnancy).

The facial deformity was noted at birth. Neither parent nor any other family member up to the second degree had a similar condition. Although there was no difficulty feeding or developmental delay, incomplete closure of the right eye with reddening of the sclera and frequent tearing when asleep eventually prompted consult.

On physical exam he had a right Tessier facial cleft 5, originating as a lip furrow approximately 5mm medial to the right labial commissure extending upward and laterally as a groove on the cheek, ending at the junction of the middle third and lateral third of lower eye lid with ectropion and downward displacement of the lateral canthus compared with the left side. (Figure 1A) The bony maxillary cleft involved the ipsilateral alveolar ridge and hard palate. (Figure 1B) The soft palate was intact. There was also a mild Tessier 5 on the left, beginning 10mm medial to the left commissure extending superiorly along the nasolabial area ending midway at the level of the ala, mildly involving the alveolar ridge on the same side.

The facial computed tomography (CT) scan revealed a thin bony defect in the right alveolus at the premolar region extending and widening posteriorly to the lateral aspect of the hard palate. (Figure 2A) An apparent thin linear defect was likewise appreciated between the left canine and first premolar mildly involving the alveolar ridge. (Figure 2A) The right skeletal cleft also involved the body of the maxilla lateral to the infra-orbital foramen (Figure 2B), through the infra-orbital rim and the orbital floor. (Figure 2C).

Correspondence: Dr. Laurice Ann B. Canta
Department of Otorhinolaryngology
Head and Neck Surgery
6th Floor, East Avenue Medical Center
East Avenue, Diliman, Quezon City 1100
Philippines
Phone: (632) 928 0611 loc. 324
Fax: (632) 435 6988
Email: eamc_enths@yahoo.com

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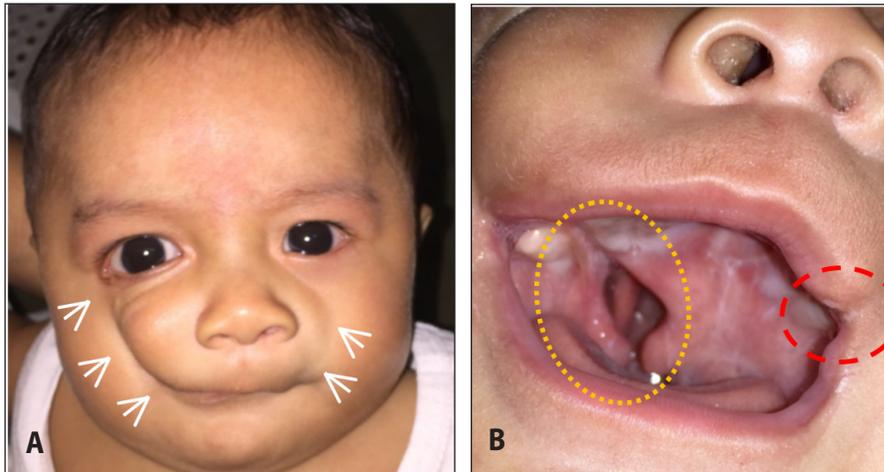


Figure 1A. Right facial cleft originating medial to the right labial commissure extending upward and laterally, ending at the lateral third of lower eye lid and a mild Tessier 5 cleft on the left beginning medial to the labial commissure going superiorly and laterally as furrow ending midway on the cheek at the level of the ala (arrows). Note ectropion and downward displacement of the lateral canthus on the right compared with the left side; **B.** Palatal component of right maxillary cleft (dotted circle) with left facial cleft beginning medial to the lateral commissure (dashed circle). Photos published in full, with permission.

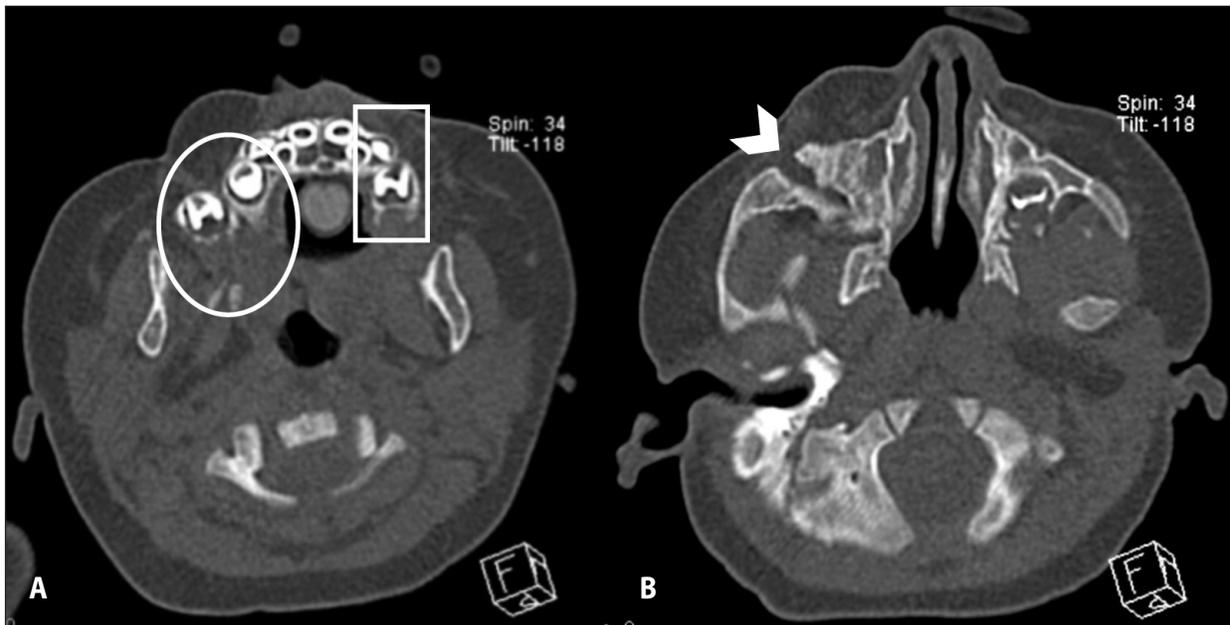
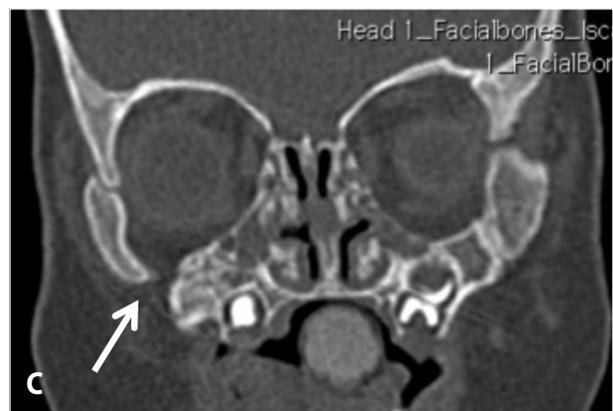


Figure 2. CT Scan A. Axial view showing the thin defect between the right first molar and second premolar extending and widening posteriorly to the lateral aspect of the hard palate (oval) and an apparent thin linear defect between the left canine and first premolar mildly involving the alveolar ridge (rectangle); **B.** Bony defect in the right maxilla lateral to the infraorbital foramen (arrowhead); and **C.** Coronal view revealing the defect in the right maxillary bone at the inferolateral orbital wall and orbital floor (arrow).



The patient was referred to the Department of Pediatrics – Genetics Clinic for further evaluation, and karyotyping at the National Institutes of Health ruled out any chromosomal disorder. Colleagues from the co-managing Department of Ophthalmology prescribed ophthalmic drops and advised lid taping while waiting for definitive surgical management.

DISCUSSION

Facial clefts may be described as “a gap (hypoplasia), misshapen face (dysplasia), interruption or deficiency in the continuity of a soft tissue (coloboma) or fissure in the soft tissue, bone or a combination of both.”¹ These craniofacial defects may be classified under the Tessier Classification which designates numbers to the site of the cleft. Based on this classification, the patient in this report has a bilateral Tessier 5 but an incomplete or mild one on the left.

The etiology of Tessier clefting especially Tessier 5 cleft is not well understood because it cannot be explained by genetics or known processes of craniofacial fusion and embryologic development.^{2,3,5-7} Factors that may contribute include interplay of the environment, like exposure to radiation or infection, intake of teratogens, vitamin or folate deficiency and metabolic or hematologic disorders.^{1,7} Some postulate that it is developmental and might be due to primary stop of development,^{1,5} neurovascular insufficiency^{1-3,5} and amniotic bands.^{2,3,5,6} Cannistra and others concluded that these defects can occur late in development when they observed that agenesis of the maxillary nerve in a 24-week-old fetus resulted in Tessier No. 5 Cleft.²

Another theory states that facial clefts are due to genetic aberrances and that they are usually associated with congenital syndromes.^{1,2,6,7} A study by Gfrerer *et al.*⁶ demonstrated that facial clefts can be traced to disruption in the human genome specifically to SPECC1L which encodes a cytoskeletal protein and its deficiency leads to failure of cell adhesion and migration. It is hypothesized that this genetic sequence is required for normal chondrocyte assemblage and molecular signaling processes leading to the fusion of frontonasal and maxillary processes and convergence of mandibular prominences.⁶ The many theories proposed with regard to the origin of orofacial clefting attests that this condition has no single cause; Tessier 5 cleft may be due to genetic disorder, neurovascular agenesis or amniotic bands.

We are not aware of any existing surgical protocol for repairing Tessier 5 Clefts. Nevertheless, the objectives of surgery for this type of orofacial cleft are to reconstruct the lower eyelid and reposition the lateral canthus, repair the labiomaxillary cleft, and “reconstruct the orbital floor, the malar bone, and the body of the maxilla with bone grafts.”^{5,8} Most patients also need serial surgical reconstruction to correct significant scarring and persistent facial deformity due to lack of development of the facial structures.^{1,5,8,9}

Commonly used surgical procedures include z-plasty, transposition or advancement flaps, canthopexy, commissureplasty and bone grafting.^{1,5,8} Since Tessier 5 clefts involve the eyelid region with ectropion and corneal exposure, reconstruction of the lower eyelid must be undertaken as soon as possible to prevent keratitis. A lateral tarsal strip is the traditional technique however in severe loss of lower eyelid tissue an advancement flap may be utilized which also has a more favourable scarring.^{5,8} Multiple z-plasties can redirect and correct the cheek fissure and labial cleft in a Tessier 5 cleft.^{1,2,5,8} There should be enough vertical length and tensionless closure in the area between the lower eyelid and upper lip to prevent asymmetry and displacement of eyelid and vermilion border when scar maturation occurs.^{8,9} When there is no viable soft tissue bulk left, free flaps or regional flaps and tissue expanders may be used.^{5,8} Bone grafts from the iliac crest, ribs and calvarial bone are used reconstruct the orbital floor, malar bone and body of the maxilla.^{5,8} Although bone failure rate is unpredictable, this is recommended to restore skeletal continuity, prevent globe prolapse and achieve facial symmetry.

For this patient, the initial procedures that can be done include lateral canthopexy and repair of ectropion using lateral tarsal strip or advancement flap from the superolateral side to elevate the lateral canthus (Eutrapio S. Guevara, Jr., expert opinion presented at the PSO-HNS Inter-hospital Grand Rounds, August 30, 2017) and address the most immediate problem which is corneal exposure. Great attention must be given during this stage to prevent disproportionate stripping or advancement leading to a narrower palpebral fissure compared to the left side (Eduard M. Alfanta, personal communication, August 29, 2017). (*Figure 3A*). Either side of the cheek can be repaired using multiple z-plasties to redirect and correct the continuity of the cheek fissure. (*Figure 3B*).

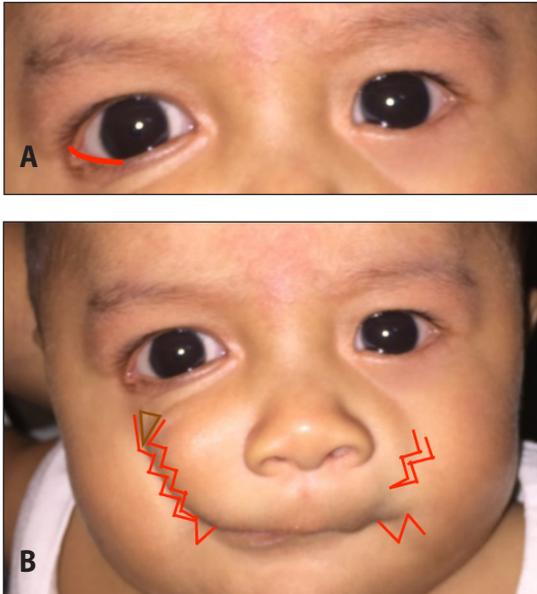


Figure 3. Proposed surgical reconstruction for the eyelid and cheek fissure: **A.** Lateral canthotomy either through lateral tarsal strip or advancement flap from the superolateral area; and **B.** multiple Z-plasties to correct the cheek fissure and redirect the labial commissures and vermillion border. Photos printed in full, with permission.

As mentioned previously, there is no single surgical plan for Tessier Clefts and although most patients with Tessier 5 cleft need serial and multiple surgical reconstructions to correct significant scarring or persistent facial deformity, a pleasing and balanced aesthetic result is achievable. Aside from surgical management, proper advice for post-operative monitoring, therapy and rehabilitation may also be recommended to achieve the best aesthetic outcome for this kind of congenital deformity.

The incision and dissection should be brought down to the subperiosteal plane since the infraorbital nerve is spared of the bony cleft and can be preserved. Soft tissue dissection should be made along the dermal fat and subcutaneous plane to allow layered closure. The periosteum and all soft tissues can be carried medially to cover the bony defect. Another z-plasty might be used to reconstruct and correct the upper lip commissures and vermillion border. Since the soft palate is intact, there would be no immediate problem in feeding and language development. An alveolar moulding or feeding plate can reshape the developing alveolus and palate and eventually narrow the gap of the cleft (Eduard M. Alfanta, personal communication, August 29, 2017). Second stage reconstruction with palatoplasty and bone grafts or use of a 3-D printed bioresorbable medical grade Polycaprolactone scaffold such as Osteopore™ for the infraorbital and maxillary bony cleft may be postponed until conditions are optimal. Factors such as bone growth and maturation must be taken into consideration to decrease the need for serial reconstruction and to come up with a better outcome as the pediatric patient grows. (Eutrapio S. Guevara, Jr., expert opinion presented at the PSO-HNS Inter-hospital Grand Rounds, August 30, 2017).

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