Tessier Clefts and Hypertelorism

Ryan Winters, MD

KEYWORDS

• Tessier cleft • Hypertelorism • Craniofacial surgery • Congenital

KEY POINTS

- Tessier's classification of craniofacial clefts denotes their position on the skull and face relative to the orbit.
- It provides no information regarding the severity of the cleft, only the location.
- Orbital hypertelorism refers to true increased distance between the bony orbits.
- Surgical repair must be tailored to the individual cleft based on severity and structures involved.

The rarity, complexity, and great variety of craniofacial clefts have all contributed to the difficulty in establishing a concise yet comprehensive classification system for these anomalies. In 1887, Morian proposed a basic schema with the infraorbital foramen as the reference.[1,2] Morian type I clefts existed in the space between the infraorbital foramen and the facial midline, and type II existed lateral to the infraorbital foramen. Clearly significant ambiguity exists within each of these 2 categories regarding structures involved in the clefting process, and a more precise classification system was needed. Boo-Chai subdivided the oro–ocular clefts described by Morian in an attempt at further refinement, and Karfik was the first to attempt classification by embryologic origin in 1966.[2,3]

In 1976, Tessier proposed a classification system based on his personal experience with 336 patients,[4,5] and the resultant ordered numbering system has greatly facilitated communication between reconstructive surgeons. Tessier's system has gained widespread acceptance and is now the most consistently used method of describing craniofacial clefts in the literature. It is centered on the orbit, with clefts assigned a number in a counterclockwise rotation. Facial clefts are numbered from 0 to 7, with 0 a midline facial cleft, and the cranial clefts are numbered 8 to 14, with 14 being a midline cranial cleft. Midline mandibular cleft is assigned number 30 (**Fig. 1**). Each of these clefts may involve both soft tissue and bone, and the number does not provide information regarding severity of tissue involvement, merely the location on the face and/or skull.

The remainder of this article explores the various Tessier clefts in more detail, followed by a discussion of orbital hypertelorism (an abnormally widened distance between the bony orbits), which may be present in association with some craniofacial clefts.

CLEFT DESCRIPTIONS

What follows is a description of each cleft type, with the bony and soft tissue manifestations discussed in further detail. Within each cleft type there is a spectrum of severity, both of soft tissue and bony involvement.

TESSIER 0

Tessier 0 cleft is a true midline facial cleft, which may be accompanied by a Tessier 14 cleft (extension of the midline cleft to the cranium), with a

Disclosure: The author has no conflicts to disclose.
Surgery Service Line, Sunrise Health System and Sunrise Children's Hospital, 3201 South Maryland Parkway, Suite 300, Las Vegas, NV 89109, USA
E-mail address: ryan.winters@gmail.com

Facial Plast Surg Clin N Am 24 (2016) 545–558
http://dx.doi.org/10.1016/j.fsc.2016.06.013

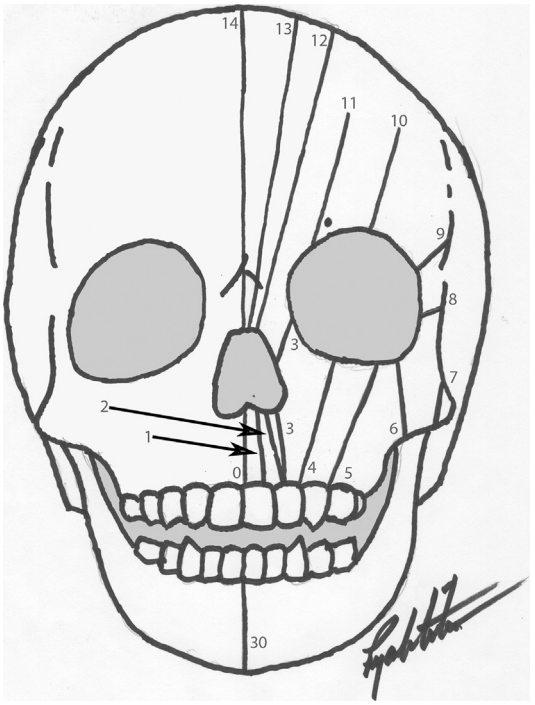

Fig. 1. Clefts numbered according to Tessier.

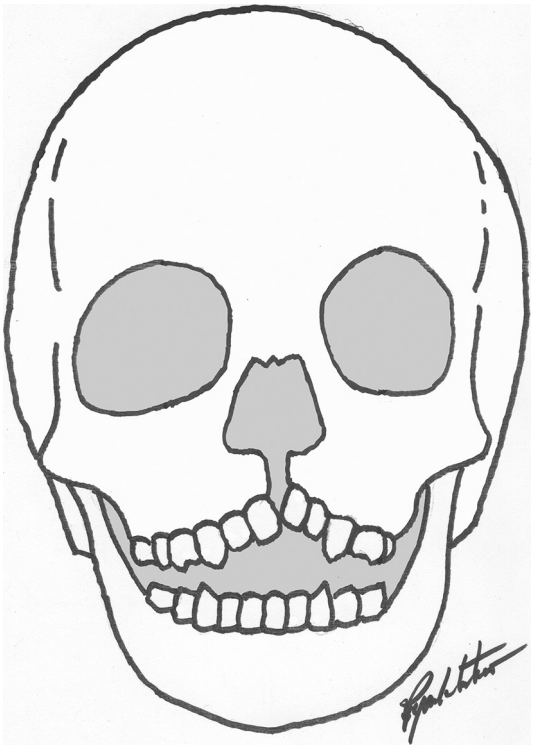

Fig. 2. Tessier 0 midline facial cleft.

resultant variable degree of hypertelorism. This is among the more common of the "atypical facial clefts," although it is still very rare, with a incidence reported as 1 in 1 million live births.[6]

Soft Tissue

The subtlest manifestation is a broadening of the philtrum, with a typical bifid nasal tip and columella, which are widened with a central concavity. In more complete cases, a true midline cleft lip may be present.[4,6] The nose will thus appear shortened, and the nasal ala laterally displaced and the alar base widened.

Bone

The midline cleft alveolus appears between the central incisors (**Fig. 2**), producing a characteristic sloping alveolar ridges toward the cleft bilaterally, described as keel shaped.[2] This will typically create an anterior open bite deformity from vertical deficiency of the maxilla in the region of the midline cleft. Involvement of the nasal septum is variable, and ranges from mild thickening of the septal cartilage and thickening and flattening of the maxillary crest, to lateral displacement of the nasal processes of the maxilla, to true duplication of the septal structures and significant lateral displacement of the nasal bones.[7]

In severe cases, where the cleft extends superiorly or exists in conjunction with a Tessier 14 cleft, the ethmoid sinuses are volumetrically enlarged and prolapsed inferiorly and laterally. There may be widening of the floor of the anterior cranial fossa and hypertelorism. The sphenoid sinus may prolapse anteroinferiorly, although the body of the sphenoid bone is characteristically normal.[2,8] The pterygoids may be displaced somewhat laterally.

TESSIER 1

A Tessier 1 cleft is similar to a "typical" cleft lip. This is a paramedian cleft in the Cupid's bow, extending superiorly to the dome of the alar cartilage or even to the medial aspect of the brow. Extension beyond the medial orbit/brow denotes a Tessier 13 cleft, which may exist simultaneously.[2,9,10]

Soft Tissue

A paramedian cleft lip is present, extending above the lip into the dome of the alar cartilage. The lower lateral cartilage and alar dome are cleft, with a short, wide columella.[2,10] The lateral remnant of the ala and lower lateral cartilage may be atrophic, curled, and deviated away from the cleft margin. Extension of the cleft into the upper lateral cartilage and nasal sidewall or paramedian nasal dorsum ranges from subtle furrow in the soft tissue