

craniofacial clefts Rare craniofacial clefts are severe deformities of the face and head that affect both bones and soft tissues. Clefts are formed in utero when normal development of a baby's head and neck are disrupted and parts of the face fail to fuse together, creating facial and/or cranial differences.

Rare craniofacial clefts occur in about 1 in every 150,000 births. These clefts are present in various syndromes, but can also occur unrelated to a syndrome.

Rare facial clefts fall into three main categories:

Clefts through the center of the face, which can create uneven eyes and other defects Oro-nasal-ocular clefts, which involve the mouth, nose and eyes Lateral clefts associated with Treacher Collins syndrome, hemifacial microsomia and other syndromes that are characterized by gross abnormalities of the lower jaw These clefts are named and numbered (0-14) for the anatomy affected and include clefts of the lip, mid-face, orbit (the eye socket), mandible and cranium. Clefts 0 - 7 typically involve the lower face (below the mid axis of the eye), while clefts 8 - 14 involve the upper face (above the mid axis of the eye) and cranium.

In some cases, different types of clefts occur together, with a cleft of the lower face continuing into a cleft of the upper face.

No. 0 — upper median facio cranial cleft: involves the upper lip, nose and forehead

No. 1 — oblique facio cranial cleft: involves the upper lip and nose

No. 2 — oblique facio cranial cleft: involves the upper lip, nose and eyelids

Nos. 3 - 5 — oblique facio cranial clefts: involves the upper lip, mid-face and eyelids

No. 6 — oblique facio cranial cleft: involves the upper lip, mid-face, outside corners of the eyes and temples

No. 7 — oblique facio cranial cleft: involves the mid-face and ear

No. 8 — transverse medial ocular facio cranial cleft: involves the mid-face, inside corners of the eyes and nose

No. 9 — lateral ocular transverse facio cranial cleft: involves the outside corners of the eyes and temples

No. 10 — first branchial cleft anomaly: involves the neck and ear

No. 11 — second branchial cleft anomaly: involves the neck and mouth

No. 12 — third branchial cleft anomaly: involves the neck and mouth

No. 13 — fourth branchial cleft anomaly: involves the neck and mouth

No. 14 — lower median facio cervical and tongue cleft (also known as glossoschisis): involves the lower lip and tongue

Tessier 30 cleft — lower midline facial cleft (also known as the median mandibular cleft): involves the lower lip, chin and mandible

Oblique craniofacial clefts and encephaloceles are each rare conditions, and only a few instances of these findings in combination have been described. Each pathologic entity presents a unique reconstructive challenge. The authors report the case of a male infant who presented with a large right frontoencephalocele and bilateral Tessier number 3 clefts ¹⁾.

¹⁾

Wood BC, Yi S, Oh AK, Rogers GF, Magge SN. Frontal Encephalocele Associated With a Bilateral Tessier Number Three Cleft and Fraser Syndrome. J Craniofac Surg. 2015 Aug 6. [Epub ahead of print] PubMed PMID: 26267579.

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