



Rare Facial Clefts

77

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77.1 Introduction

Since ages, congenital deformities were considered evil and wizard, and infants were abandoned to die in isolation. Jean Yperman (1295–1351) valued the congenital origin of the clefts. He additionally characterized the different types of the condition and set out the standards for their treatment. Fabricius ab Aquapendente (1537–1619) and William His of college of Leipzig independently researched and published embryological premise of clefts [1].

Laroche was the first to separate between common cleft lip or harelip and clefts of the cheek. Further qualification was made in 1864 by Pelvet, who isolated oblique clefts including the nose from the other cheek clefts, and drawing on Ahlfeld's work, in 1887 Morian gathered 29 cases from the writing, contributing 7 instances of his own. Morian perceived three unique groups of oblique facial clefts. From that point forward, astounding audits have been composed by Grinberg in 1913, Boo-Chai in 1970, Paul Tessier in 1976 [2] and Millard in 1977 [3].

Craniofacial cleft by definition is “a fissure of the soft tissues that corresponds as a general rule with a cleft of the bony structures.” [1] The greatest research on craniofacial clefts was finished by Tessier and is credited for the formation of the craniofacial surgery for establishing the framework of the advanced craniofacial surgery by fundamentally breaking down facial clefts and portraying facial osteotomies [4].

Craniofacial clefts are significant clefts affecting the face, cranium, or both. These clefts cause distortion of the face and cranium with lacks or abundances of tissue that cleave anatomic planes in a straight fashion [2]. Craniofacial clefts exist in changing degrees of seriousness, and practically every one of them happens along the anticipated embryo-

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logic lines. These clefts can be either complete or incomplete and can seem alone or in relationship with other facial clefts. Seriousness of craniofacial clefts fluctuates extensively, running from a scarcely distinguishable indent on the lip or on the nose or a scar-like structure on the cheek to an extensive partition of all layers of facial structures. Notwithstanding one parted sort can show on one side of the face, while an alternate kind is available on the other side [2, 3].

Craniofacial clefts need comprehensive rehabilitation. Past the physical consequences for the patient, they have monstrous mental and financial impacts on both patient and family, prompting disturbance of psychosocial working and diminished nature of life [4, 5].

Cleft repair is a necessary part of the modern craniomaxillo-facial surgical spectrum and remains a challenge on account of inadequate and contorted tissue (minor to major) at the site of the deformity [6]. The outcomes are additionally impacted by the short and long haul aesthetic (soft tissue and facial skeletal appearance) [7] and useful (occlusal and discourse) outcomes [8]. What's more, the kind of careful fix, the specialist's abilities and the compliance of the patient likewise, affects the stylish [9] and utilitarian [10]outcomes. The real test isn't just understanding the hereditary qualities [11], in addition to plan the standard conventions for the surgery in these phenomenal kinds of clefts [12].

77.2 Incidence

Craniofacial clefts are a lot rarer than the simple cleft lip/palate deformity [13]. The precise occurrence of craniofacial clefts has not been exactly documented in view of their rarity. However, the reported frequency of craniofacial clefts is 1.5–6.0 per 100,000 live births [14]. The occurrence of uncommon craniofacial clefts contrasted with ordinary cleft lip and palate may vary from 9.5 to 34 for every 1000 [15].

Lateral or transverse clefts of the lip are very uncommon and have commonly archived to have a rate of 0.3% of 1.0% of all of the facial cleft deformity spectrum (Boo-Chai 1969; Hawkins et al. 1973; Bauer et al. 1982; Verheyden 1988; Gleizal et al. 2007), or of 0.02% of live births (Kuriyama et al. 2008) [16]. Median clefts of the lower lip are very rare and only 68 cases have been accounted for till date [17].

77.3 Embryology

Successful treatment of innate craniofacial defects depends on an intensive comprehension of the embryologic procedures prompting their development [18]. There are various interesting highlights that plainly recognize craniofacial improvement from the advancement of different tissues in the body.

One of these novel highlights is the double starting point of craniofacial tissues: the skeletal framework and the vast majority of the connective tissues, including veins, begin from a gathering of cells called the cranial neural crest, while the musculature and some parts of the skull begin from mesoderm.

A second one of a kind component is the unit of intricate, complementary tissue interactions between the neuroectoderm, the mesenchyme, and facial ectoderm that drive ordinary advancement.

A third novel element is the extravagantly arranged morphogenetic developments—brought about by both uninvolved cell removal and dynamic cell movement—that characterize head advancement. Any procedure that upsets the rate, the planning, or the degree of these complex cell conduct can result in a craniofacial birth imperfection.

77.3.1 The Initiation of Craniofacial Development

77.3.1.1 Establishment and Fusion of the Facial Prominences (Figs. 77.1 and 72.3)

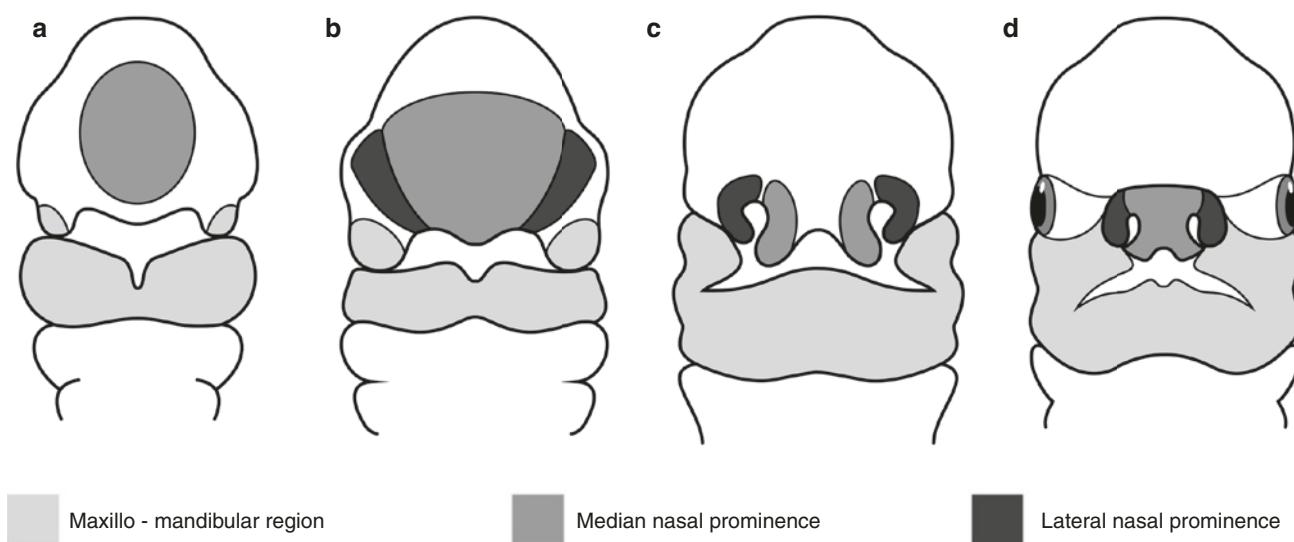
The basic morphology of the face is established between the 4th and 10th week of human development. The face is formed as a result of fusion of the midline frontonasal prominence and three paired prominences, the maxillary, lateral nasal, and mandibular prominences. Each of these prominences is filled with cranial neural crest cells that originated at different positions along the neural tube.

77.3.1.2 The Frontonasal Prominence

The frontonasal prominence gives rise to the forehead, midline of the nose, the philtrum, the middle portion of the upper lip, and the primary palate. Interruptions in frontonasal growth often result in a bilateral cleft lip, where the primary palate frequently “everts.” In the mildest cases, clefts involving frontonasal prominence-derived structures may be limited to a notch in the vermillion border of the lip. In more severe cases, frontonasal clefts involve all of the tissues of the lip, and these cases may most likely occur because of a failure of fusion between the frontonasal and maxillary prominences.

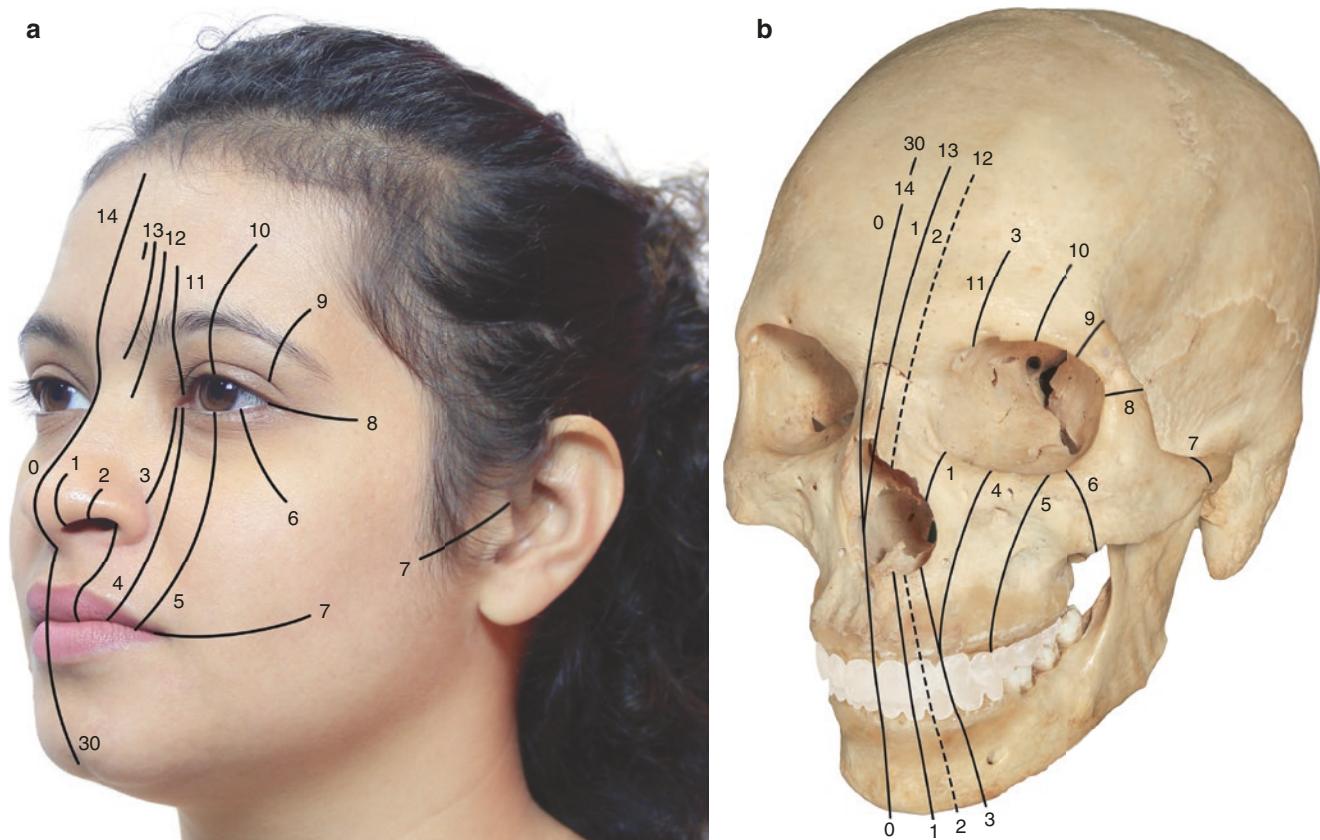
77.3.1.3 The Lateral Nasal Prominences

The lateral nasal prominences give rise to the alae of the nose. Clefts that involve the side of the nose often result from a failure in the fusion between the lateral nasal prominences and either the frontonasal or the maxillary processes.



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Fig. 77.1 (a–d) Embryological representation of fusion of nasal prominences



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Fig. 77.2 (a) Tessier classification for soft tissue clefting. (b) Tessier classification for bony clefting

77.3.1.4 The Maxillary Prominences

The maxillary prominences develop into the upper jaw, the facial halves, the upper lips, and the secondary palate. The nasal passage is divided from the pharynx by the secondary palate, which is formed from the neural crest cells. The palatal shelves first drop vertically and then rotate into a horizontal plane on the dorsal side of the tongue before fusing. The epithelium of the palatal shelves sloughs off, with only the basal layer of epithelium remaining to cover the later palate.

77.3.1.5 The Mandibular Prominences

This develops into the lower jaw and lower lip. Clefts of the lower jaw are very rare, presenting with a wide variety of phenotypes, ranging from a vermillion notch on the lip to a complete cleft involving the anterior mandible, chin, tongue, and lower lip and occasionally involving midline structures of the neck up to the manubrium sterni [19].

77.4 Etiology and Pathogenesis

Development of the head and face contains a standout among the most mind-boggling events among embryonic advancement. Disturbance of this firmly controlled course can result in a facial cleft where the facial primordia fail to meet and

form the suitable structures [20]. The definite instrument of the cause of facial clefts is obscure, yet they are accepted to have a multifactorial etiology including a mix of natural and hereditary causes during embryonic development [21, 22]. In India affiliation and healthful inadequacies in pregnant mothers are the main cause of clefts [23].

The discussion is as yet in contention between the supporters of Meckel who trusted that clefts were brought about by a developmental arrest and Geoffroy St. Hilaire (1832) who felt that amniotic groups were responsible [15].

Fogh-Andersen previously characterized hereditary factors in clefting, which have been affirmed by segregation analysis [23]. Research in molecular genetics have identified genes responsible for rare facial clefts which may be syndromic and also for complex non-syndromic variants [24].

The non-syndromic types of orofacial clefts are likely due to secondary gene-environment interactions [25]. Non-syndromic cleft is a heterogeneous disease entity with candidate clefting loci on chromosomes [1, 2, 4, 6, 11, 14, 17, 19].

Four general classes of natural “cleftogens” have been distinguished to date, as follows [26]:

Radiation. Huge dosages of radiation have been associated with microcephaly.

Infections. The offspring of mothers with toxoplasmosis, rubella, or cytomegalovirus diseases display expanded frequencies of facial clefts.

Maternal idiosyncrasies. Mothers of children with CLP display an increased rate of phenylketonuria. The oculo-auriculovertebral range has been seen with strange recurrence among neonates with mothers who are diabetic. Numerous examinations have proposed maternal factors, for example, age, weight, and general well-being, as potential reasons for distortions.

Chemicals. Nutrient deficiencies are related with expanded rates of CLP, which might be decreased with vitamin supplementation for expectant mothers. Vitamin A, its subsidiaries, and related compounds, for example, isotretinoin, have been involved in the developments of facial clefts and hemifacial microsomia [26].

Any maternal liquor consumption during pregnancy increases the frequency of orofacial clefting [27]. The impact of maternal smoking also may be responsible [28]. Multiple studies have demonstrated that folate deficiency is related with clefts. Pre-birth folic acid supplementation has shown to diminish this hazard. At present, folic acid supplementation is the main empirical safeguard to diminish the frequency of facial clefting [2].

77.5 Classification

An all-round grouping plan that completely envelops, precisely depicts, and coordinates all the different types of orofacial and craniofacial clefts does not exist [2]. Soemmering (1791), Morian in 1886, Degenhardt (1961), the American Association of Cleft Palate Rehabilitation (AACPR) (1962), and Boo-Chai have made huge contributions in building up a classification [29–32].

77.5.1 Tessier Classification

In 1976, Paul Tessier depicted an anatomical order framework in which a number is doled out to every abnormality based on its position with respect to the sagittal midline [33].

This framework has moved towards global acknowledgement and allows reliable correspondence among clinicians [34].

This classification involves the orbit as the principle reference point. Fifteen areas of clefting have been demonstrated with discussion of their soft tissue and hard tissue involvement [33] (Fig. 77.2a, b).

The numbered clefts relate the soft tissue features to underlying bony involvement.

These have been verified by operative findings and more recently preoperative 3D CT assessments [35]. The clefts are radially distributed around the orbit with the midline 0 cleft named as median facial dysrhaphia [33].

77.5.1.1 Number Zero

The no. 0 cleft is the most widely recognized of the craniofacial clefts [36]. No. 0 and 14 is also called as midline craniofacial dysrhaphia. Clinically, this cleft shows up as an absence of conclusion of the front neuropore [33].

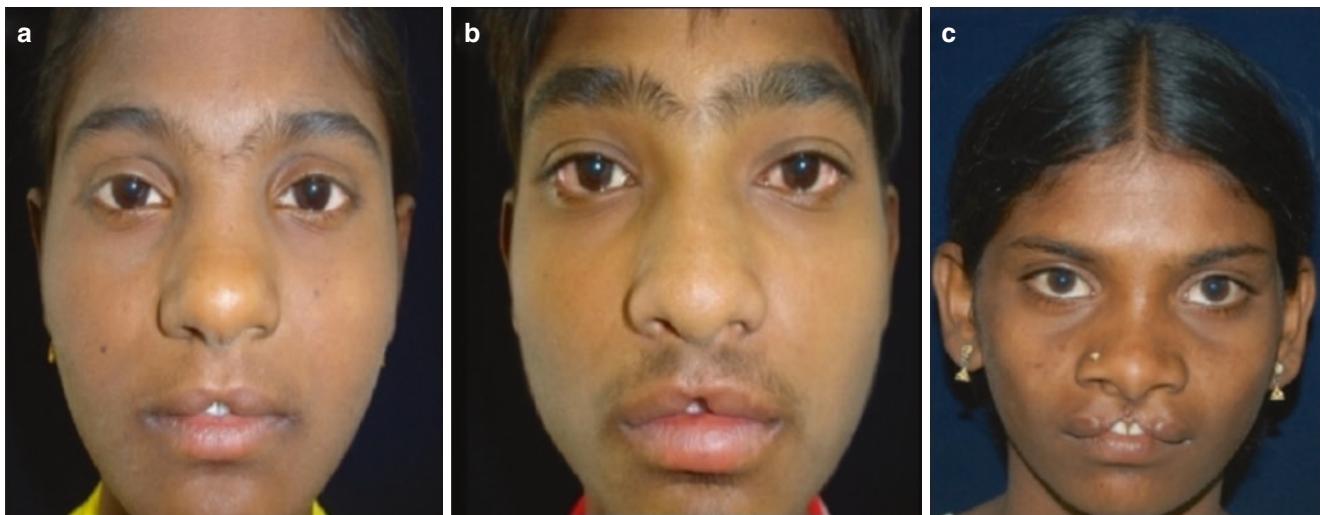
It shows two variants: a true and false middle congenital cleft, with or without related hypo- or hypertelorism [36]. Developmental cause of Tessier no. 0 isn't evident; however, it is realized that midline facial deformities can be followed to a period relating to the third week of gestation [36]. The cleft expresses as a duplication of the crista galli in frontal bone ("skull bifidum" and middle encephalocele), and as nasal septal duplication, and cleft through the columella, maxilla, and lip [33]. This form of cleft may have either a deficiency or abundance of tissue: with tissue agenesis and holoprosencephaly toward one side and frontonasal hyperplasia and inordinate tissue (the hyperplasias) at the opposing end. Midway inconsistencies with normal tissue volume possess the center segment of the spectrum [37].

Features of true midline congenital fissure:

1. Split upper lip with renal duplication
2. Bifid nose with wide columella and a wide furrow in the nasal dorsum
3. Duplication of the nasal septum
4. Overprojected nose due to fibromuscular connection of the alar ligaments and the frontal bone
5. Hypertelorism
6. Low placed cribriform plate of the ethmoid bone
7. Occasional midline encephalocele [38]

Features of the pseudo midline congenital fissure include:

1. Hypotelorism
2. Absence of the philtrum of the lip, premaxilla, nasal septum, columella, and the crista galli [33]
3. Clefting along the entire length of the upper lip
4. Hypoplastic nasal septum
5. Holoprosencephaly
6. Hypoplastic ethmoid bones [37]
7. Widened body of the sphenoid bone with separation of the pterygoid plate [35]



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Fig. 77.3 (a–c) Clinical subclassification of Tessier number 0. (a) Type I—Involving only vermillion not involving the white roll. (b) Type II—Involving vermillion and white roll. (c) Type III—Involving vermillion, white roll, and philtrum



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Fig. 77.4 (a–c) Clinical subclassification of Tessier no. 0. (a) Type IV—Involving vermillion, white roll, philtrum, columella. (b) Type V—Involving columella and tip, supratip, and dorsum of the nose.

(c1, c2) Type VI—Involving columella, tips, supratip, dorsum of the nose and frontonasal area

Millard [39] classified a middle split of the lip as any vertical cleft through the focal point of the upper lip, paying little heed to the degree (Fig. 77.3a–c). Middle clefts have been isolated into two gatherings by Millard and Williams [39]. The principal bunching includes agenesis of the frontonasal procedure, and the second gathering is portrayed as separated to the middle component. The last is related with different degrees of nasal bifurcation and cranial malformations including hypertelorism [40–42] (Fig. 77.4a–c).

(Fig. 77.5). It also widens the area between the nasal bone and the frontal process of maxilla. The soft tissue component involves the dome of the nose and may involve the alveolus and the lip. It may have a cranial counterpart in No 13 cleft. [33]

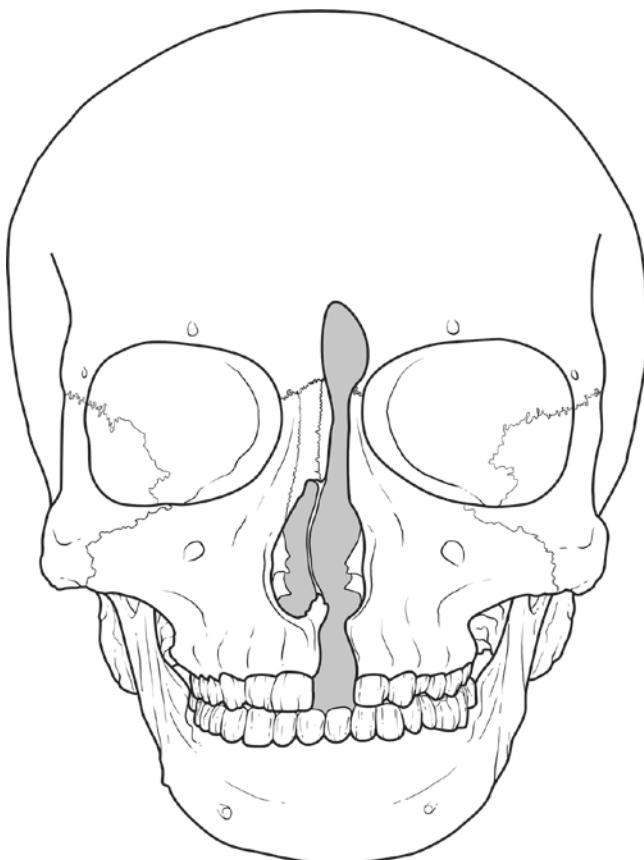
Soft Tissue Characteristics:

Soft tissue characteristics of No 1 cleft include:

1. Alar clefting that produces deviation of the nose to the opposite
2. Vertical ridges and furrows on the nasal dorsum
3. Vertical orbital dystopia and telecanthus
4. Tongue-shaped frontal hairline which is indicative of no. 13 cleft [35]

77.5.1.2 Number 1 Cleft

The number 1 cleft is also called as a paramedian cleft. Skeletally it passes through the frontal bone affecting the olfactory groove along the ethmoid producing hypertelorism



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Fig. 77.5 Tessier 1

Skeletal Involvement:

Skeletal features are as follows:

1. Clefting of the maxilla extending posteriorly as a total cleft of the hard palate.
2. Hypoplastic maxilla.
3. Nasal dorsum is deviated.
4. Asymmetry of the pterygoid plates and increased prominence of the lesser wing of the sphenoid bone.
5. Mild plagiocephaly may also be an associated feature [35].

77.5.1.3 Number 2 Cleft

Tessier no. 2 clefts are found parallel to the midline Tessier No. 0 clefts [33]. The deformation of the middle third of the nostril rim is a characteristic feature of the Tessier 2 cleft. This produces widening of the nasal bridge and flattening of the lateral side of the nose (Fig. 77.6a–e).

Skeletal features of Tessier 2 clefts are as follows:

1. Alveolar dysplasia from the lateral incisor to the pyriform aperture.

2. Palatal cleft may or may not be seen.
3. Deviated nasal septum.
4. A bony indentation is seen near the naso-maxillary suture [35].
5. Dysplasia of the lateral ethmoid region with orbital hypertelorism [43].
6. Cranial base asymmetry.
7. Dislodged medial canthus with intact lacrimal duct [44].

77.5.1.4 Number 3 Cleft

No. 3 is the oculo-nasal cleft (Morian I). This is also called “medial” orbito-maxillary cleft which passes through the lacrimal segment of the lower eyelid. This paranasal cleft occurs obliquely involving the lacrimal groove [33]. The patient may have microphthalmia [35] but anophthalmia is rare [45] (Figs. 77.7a–c and 77.8a–c).

Soft Tissue Characteristics [35, 46]

The important soft tissue features of the Tessier 3 cleft are:

1. Soft tissue hypoplasia of the face in the vertical direction [35]
2. Clefting of the alar base involving the nasolabial groove
3. Alveolar cleft with cleft of the lip
4. Displacement of the medial canthal apparatus and the lacrimal punctum of the lower eyelid
5. No patency of the lacrimal duct into the nose
6. Normal globe with orbital dystopia

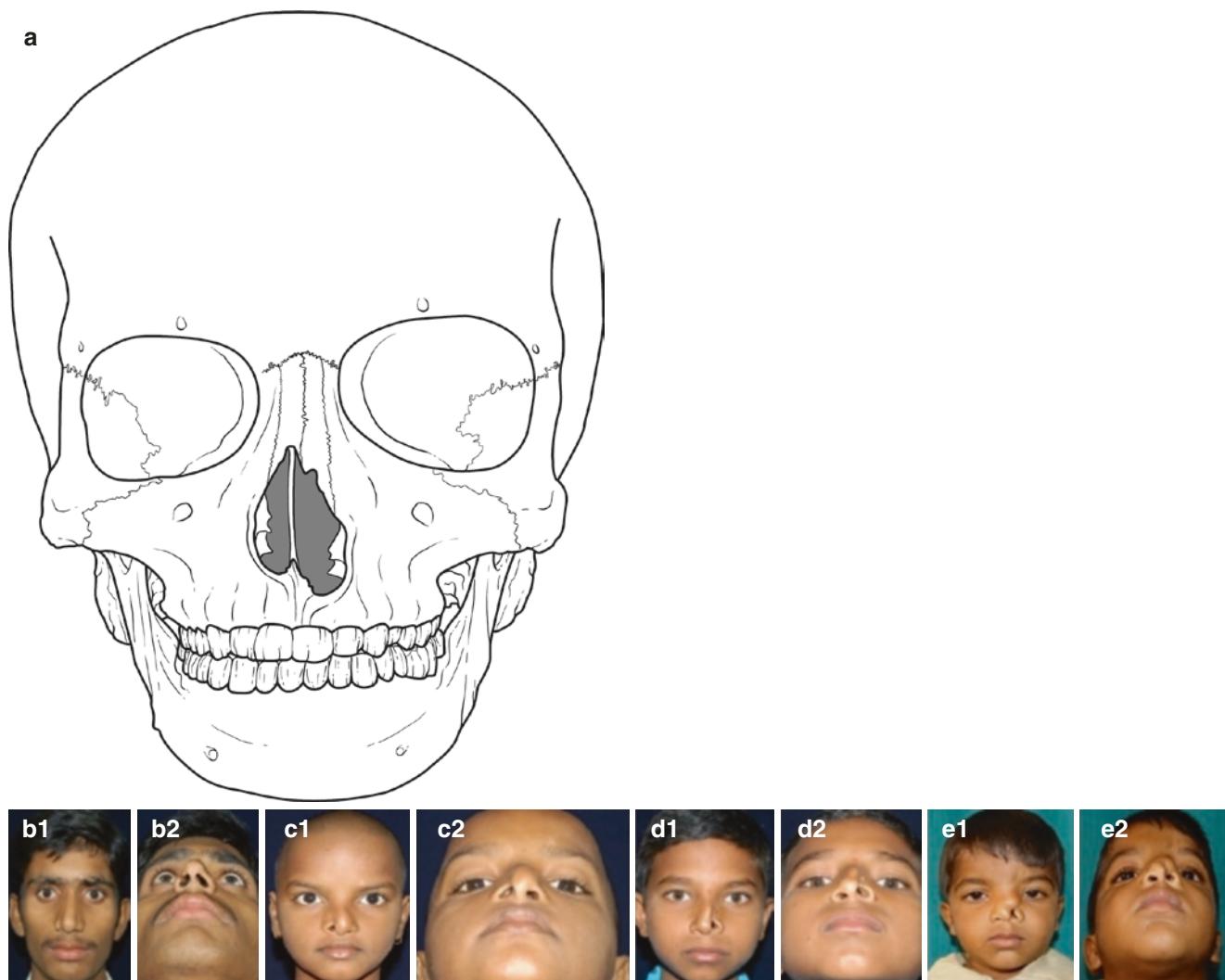
Skeletal involvement include the following features:

1. Absence of the frontal process of the maxilla and medial wall of the maxillary sinus [33]
2. Deviated nasal septum
3. Cleft lip and palate
4. Continuity of the nasal cavity into the maxilla with no bony lateral nasal wall [35]
5. Hypoplastic maxilla
6. Narrowing of the ethmoid body and sinus on the affected side [35]
7. May be associated with a cranial cleft No 10 or 11 [33]

77.5.1.5 Number 4 Cleft

The No 4 Tessier cleft is a rare, complex, and gruesome craniofacial malformation [47]. No. 4 is the oculofacial separated I (Morian II). This is a “focal” orbito-maxillary cleft [33].

It may range from a unilateral notch paramedially to large bilateral tissue defects extending from the mouth to the eyes with huge bony fissures [47] (Fig. 77.9a–f).



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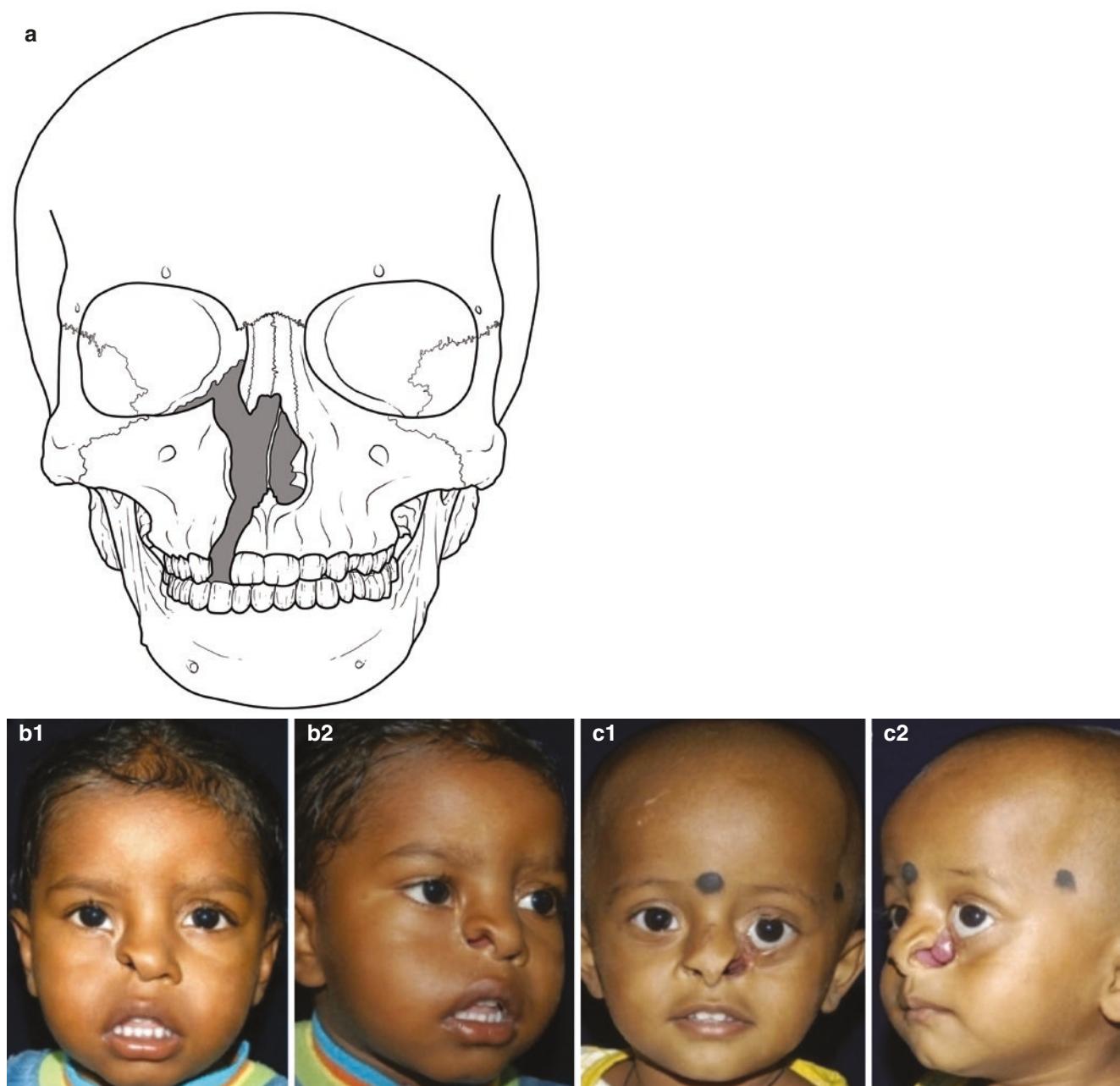
Fig. 77.6 (a–e) Tessier 2 clefts ranging from microform defect to more severe

Soft tissue characteristics include:

1. Clefting extending halfway between the philtral crest and the labial commissure
2. Extreme degree of vertical tissue deficiency extending from the labial cleft to the ocular separation in the lower eyelid [35]
3. Decreased oro-ocular distance
4. Microphthalmia with exposure keratitis secondary to eyelid deficiency [47]
5. Dystopia and inferior placement of the involved globe

Skeletal characteristics are enumerated below:

1. Bony cleft starting caudally between the lateral incisor and the canine proceeding cephalically medial to the infra-orbital foramen onto the orbital floor.
2. Prolapse of the orbital contents into the maxillary sinus due to the orbital defect.
3. Naso-lacrimal duct is intact, but the inferior lacrimal canaliculi may be hypoplastic or absent [47].
4. This may be associated with a cranial cleft no 12.



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Fig. 77.7 (a–c) Tessier 3 cleft with ocular involvement

77.5.1.6 Number 5 Cleft

The Tessier No 5 cleft is a very uncommon malformation and is also referred as the oculofacial cleft II (Morian III). This lateral orbito-maxillary cleft gets through the medial third of the lower eyelid [33] (Fig. 77.10a, b).

Soft tissue involvement demonstrates:

- Clefting immediately medial to the commissure which courses along the cheek lateral to the ala of the nose, ending in the lateral half of the lower eyelid.

- The globes are usually normal but may occasionally show microphthalmia [35].

Skeletal involvement:

- The alveolar cleft starts lateral to and travels lateral to the infra-orbital foramen and ends in the lateral part of the orbital floor. There may be associated hypoplasia of the maxillary sinus [48].

Fig. 77.8 (a–c) Tessier 3. (a) With bilateral involvement (b) without ocular involvement (c) with oral involvement

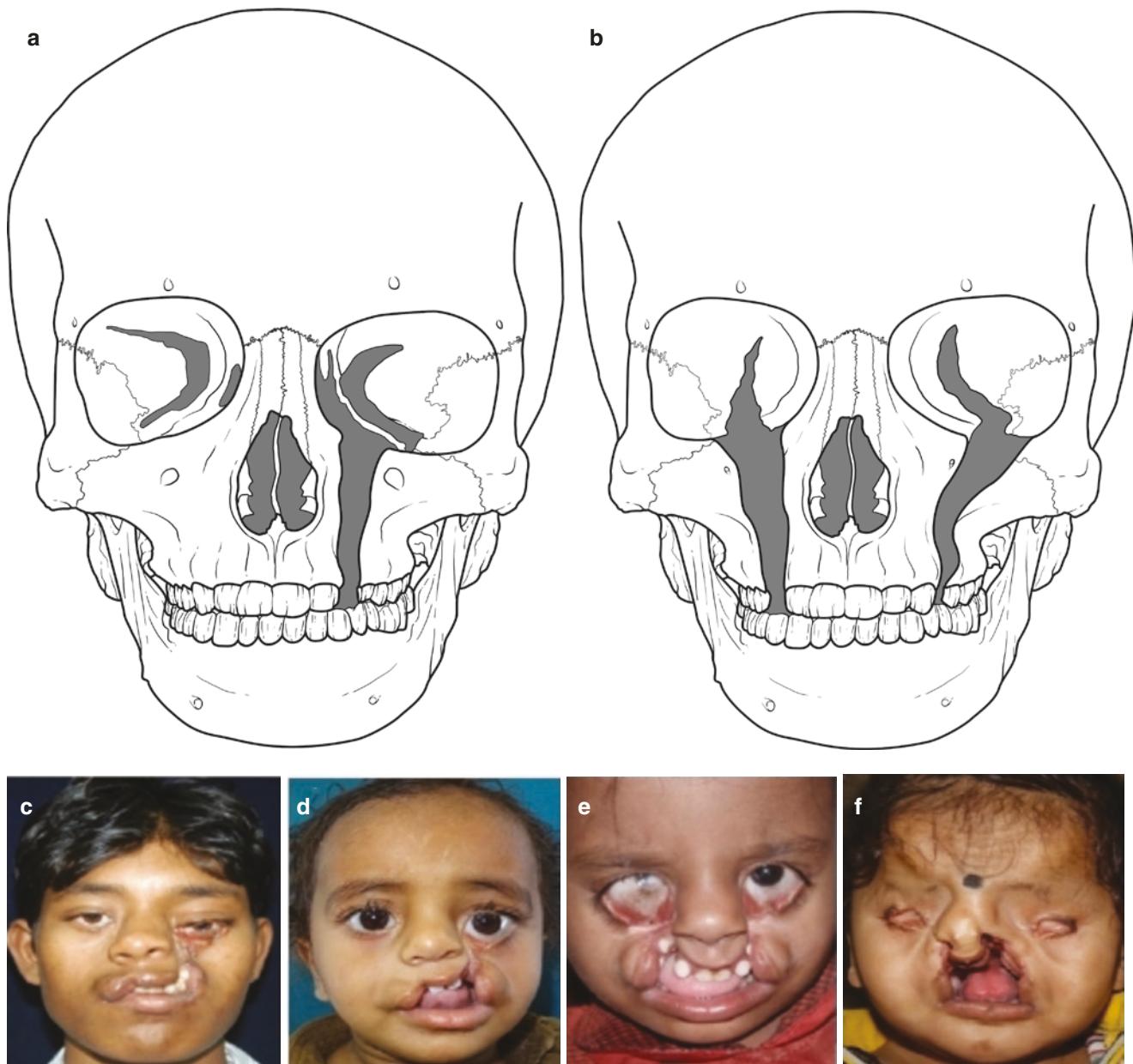


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2. Vertical orbital dystopia due to prolapse of the orbital contents.
3. Thickened lateral orbital wall and abnormal greater wing of the sphenoid bone.
4. Cranial base may be generally normal [35].

77.5.1.7 Number 6 Cleft

This is otherwise called as zygomaticomaxillary cleft which may form an incomplete variant of the Treacher Collins syndrome (Fig. 77.11a, b). It was named as maxillozygomatic dysplasia by Van der Meulen [33, 49].



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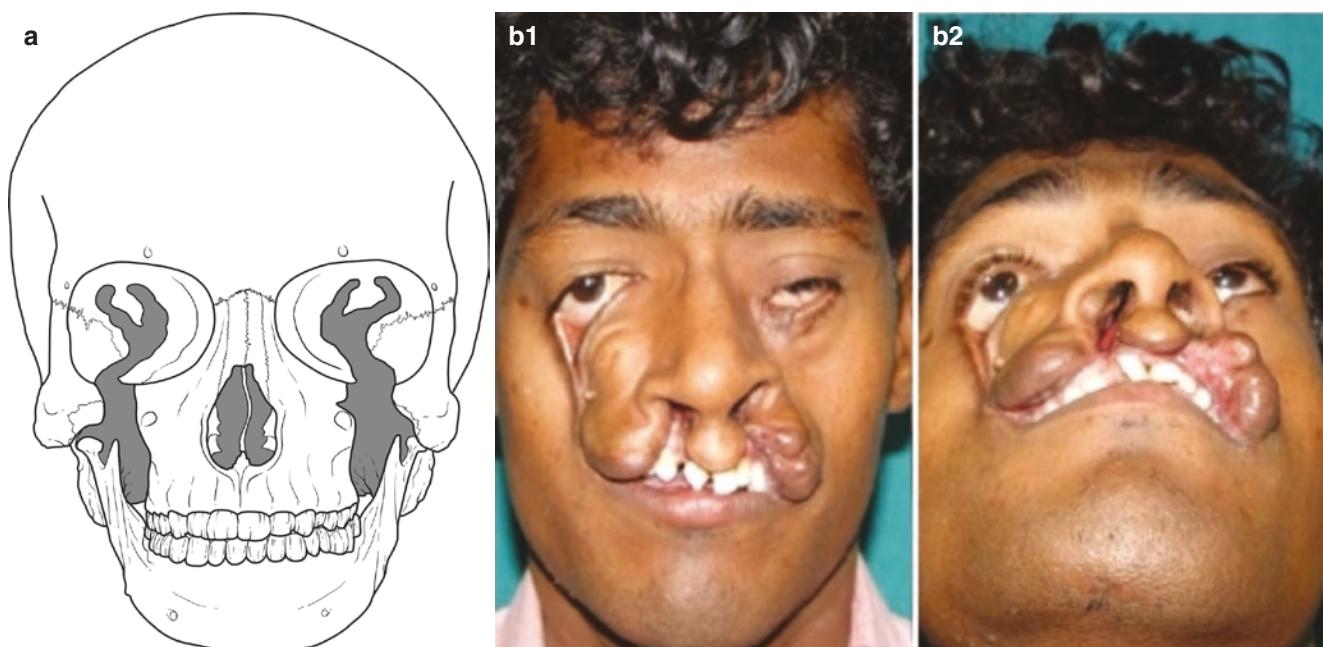
Fig. 77.9 (a–f) Tessier 4. (a, b) Skeletal Tessier 4. (c, d) Unilateral Tessier 4. (e, f) Bilateral Tessier 4

Soft tissue involvement of the No 6 cleft is detailed below:

1. Vertical cleft furrow extending from the oral commissure to the lateral aspect of the lower eyelid.
2. This also involves the zygomatic eminence pulling the lateral aspect of the palpebral fissure down with inferior displacement of the lateral canthus [33].
3. This gives the appearance of an anti-mongoloid slant and lower lid ectropion.
4. Colobomas may be seen in the lateral lower eyelid region [33].

Skeletal involvement is as follows:

1. A bony split is seen along the zygomaticomaxillary suture separating the maxilla and the zygoma [33].
2. Usually there is no alveolar cleft.
3. The maxilla may be shorter antero-posteriorly.
4. The cleft enters the orbit at the lateral orbital floor.
5. The zygoma is hypoplastic with change in the cheek contour.
6. The anterior cranial fossa is narrow but sphenoid is normal [35].



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Fig. 77.10 (a, b) Tessier 5



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Fig. 77.11 (a, b) Tessier 6. Skeletal Tessier 6. (a) Unilateral Tessier 6. (b) Bilateral Tessier 6

77.5.1.8 Number 7 Cleft

This is a temporo-zygomatic cleft and is the most well-known of all the craniofacial clefts [35] (Figs. 77.12 and 77.13a–d). It may occur along with hemifacial microsomia.

Soft tissue features include:

1. Soft tissue clefting from the oral commissure to the pre-auricular hairline.
2. It may range from a mild broadening of the oral commissure with pre-auricular skin tags to a complete fissure ending in a microtic ear [35].
3. There may be deficiency of the ipsilateral tongue, soft palate, and muscles of mastication.

4. There may be absence of the parotid gland and duct.
5. There may be external and middle ear abnormalities [50–52].
6. Abnormal pre-auricular hair form in hemifacial microsomia and Treacher Collins disorder [35].

Skeletal qualities include [35, 53, 54]:

1. Skeletal cleft involves the pterygo-maxillary junction.
2. Hypoplasia of the posterior maxilla, mandibular ramus, coronoid, and condylar process with occlusal canting [33].



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Fig. 77.12 Tessier 7



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Fig. 77.13 (a–d) Subclassification of Tessier 7. (a) Oro-aural Tessier 7. (b) Bilateral Tessier 7. (c) Tessier 7 with cleft lip. (d) Tessier with ear tag

3. Hypoplasia of the zygoma with displacement and deformation. In extreme cases the arch may manifest as a little stump alone.
4. There may be true orbital dystopia in severe cases.
5. There may be a tilt of the cranial base [35].

77.5.1.9 Number 8 Cleft (Fig. 77.2)

This fronto-zygomatic cleft situated at the lateral canthus forms the equator of the Tessier craniofacial cleft sphere. It is a part of the zygomatico-frontal dysplasias [49]. The number 8 cleft seldom occurs in isolation and usually occurs as a part

of other craniofacial clefts. It corresponds to the cranial occurrence of the No 6 cleft. In bilateral occurrences it is associated with numbers 6 and 7. This shows features similar to the Treacher-Collins and the Goldenhar's disorder with the former showing more skeletal deformities and the latter soft tissue ones [55].

Soft tissue features include [56, 57]:

1. Cleft extends from the lateral canthus to the temporal region.
2. They may present with dermatoceles and colobomas with absence of the lateral canthal apparatus.
3. Abnormal hair patterns may be present between the temporal area and the lateral canthus.
4. Globe may show eye bulbar dermoids.

Skeletal involvement includes [58, 59]:

1. Bony clefting at the fronto-zygomatic suture [58].
2. Features of the Goldenhar's or Treacher-Collins syndromes may be seen with the zygoma being hypoplastic or missing along with the lateral orbital wall [59].
3. The palpebral fissure shows a lateral descent due to the absence of the zygoma.

77.5.1.10 Number 9 Cleft

No. 9 is a form of the upper "lateral" orbital cleft. The clefting is seen in the lateral third of the upper eyelid and the lateral supra-orbital angle (Fig. 77.14). It is the cranial extension of the number 5 facial cleft [52]. There may be a deficient greater sphenoid wing in this type [60].

Soft tissue involvement shows:

1. Anomalies of the lateral third of the upper eyelid and the eyebrow.
2. Distortion of the lateral canthus.
3. Severe cases show microphthalmia.
4. The superolateral bony deficiency in the orbit may be the cause of lateral displacement of the globes.
5. Anterior displacement of the temporal hairline with temporal projection.
6. Palsy of the 7th cranial nerve with temporal and eyelid signs may be common.

Skeletal involvement:

1. Cleft extending through the superolateral aspect of the orbit.
2. The greater wing of the sphenoid may be distorted affecting morphology of the orbital wall.



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Fig. 77.14 Tessier number 9

3. Hypoplasia of the pterygoid plates.
4. Decrease in the antero-posterior dimensions of the anterior cranial fossa [35].

77.5.1.11 Number 10 Cleft

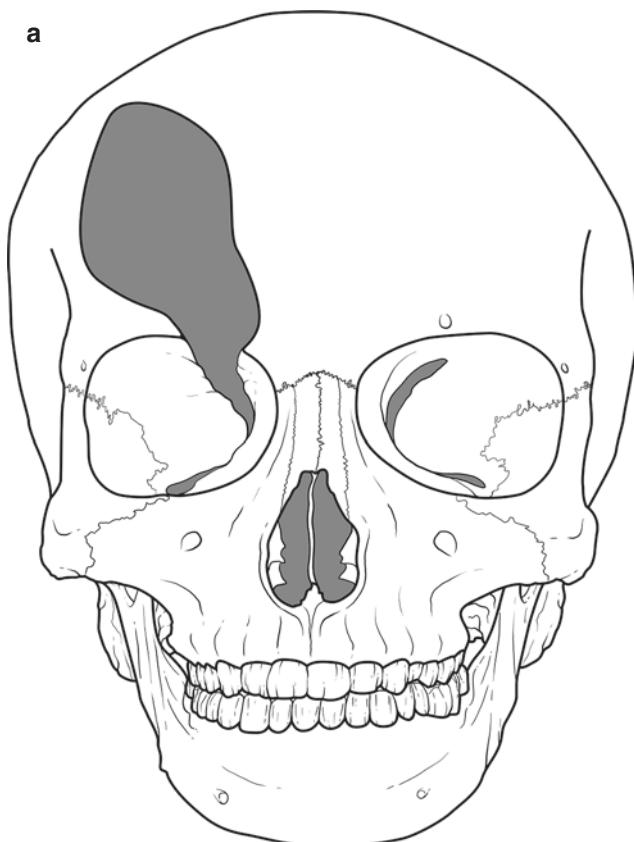
No. 10 is an upper "central" orbital cleft with the cleft happening in the middle third of

the supra-orbital edge, lateral to the supra-orbital nerve (Fig. 77.15a, b) [33]. This is the cranial counterpart of the No 4 cleft. Both present with similar ocular deformities and may show colobomata of the iris [33].

This cleft causes a large defect in the frontal bone [46].

Soft tissue features include:

1. Elongated palpebral fissure
2. Amblyopia with inferior and laterally displaced eye [35]
3. Eyebrow which is deficient medially and is more dispersed laterally [46]
4. Occurrence of frontal encephalocele involving the middle third of the frontal bone and the orbital roof [35]



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Fig. 77.15 (a, b) Tessier number 10

Skeletal involvement:

1. Cleft defect on the lateral aspect of the supra-orbital rim which may involve an encephalocele.
2. Orbital deformity with a latero-inferior rotation.
3. Orbital hypertelorism may be a feature in severe cases with distortion of the anterior cranial base [35].

77.5.1.12 Number 11 Cleft (Fig. 77.2)

No. 11 is the upper “medial” orbital cleft. This shows coloboma of the medial third of the upper eyelid, with stretching of the eyebrow [33]. This cleft is often associated with the facial cleft No 3 [35]. Van der Meulen incorporated this deformity in his frontal dysplasia group [49].

Soft tissue involvement includes:

1. Coloboma of the upper eyelid in the medial third
2. Disturbance in the upper eyebrow with a tilt toward the frontal hairline
3. Tongue-like projection of the frontal hairline [35]

Skeletal involvement:

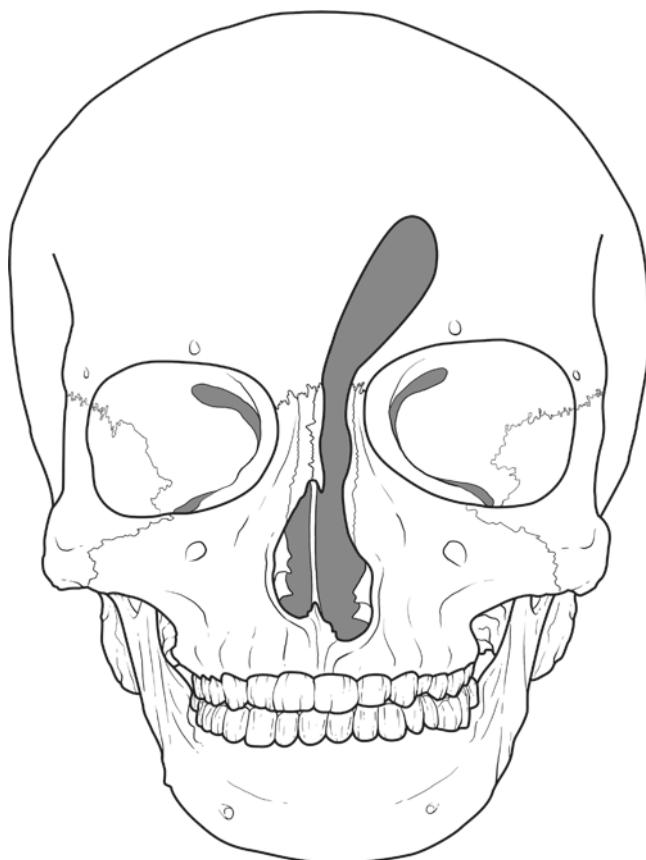
1. Frontal bone defect medial to the supra-orbital nerve and lateral to the ethmoid and lacrimal bones
2. Bony defect of the medial orbit
3. Fronto-ethmoid encephalocele with resultant orbital dystopia
4. Deficiency of the lacrimal bone with associated lacrimal stenosis [46]

77.5.1.13 Number 12 Cleft

The No 12 cleft shows deficiencies in the ethmoid labyrinth and the glabella [46]. This cleft is usually found medial to the medial canthus [35] (Fig. 77.16).

Soft tissue characteristics include:

1. Soft tissue dehiscence medial to the medial canthal apparatus [33]
2. Lateral displacement of the medial canthus with aplasia of the medial eyebrow



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Fig. 77.16 Tessier number 12



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Fig. 77.17 Tessier number 13

3. Presence of a V-shaped frontal hairline in the paramedial region of the forehead [35]

Skeletal involvement:

1. There is a flattened widening of the affected bone.
2. May present with grades of hypertelorism or telecanthus.
3. Normal sphenoids with mild hyper-pneumatization of the frontal sinus.
4. Obtuse fronto-nasal angle.
5. Low incidence of associated encephaloceles.

77.5.1.14 Number 13 Cleft

The number 13 cleft is the cranial counterpart of the paramedian facial cleft 1.

It is situated between the nasal bone and the frontal process of the maxilla passing through the frontal bones and along the olfactory groove [36] (Fig. 77.17).

Soft tissue characteristics include the following:

1. This occurs medial to the eyebrow which remains undivided [36].
2. The cleft produces a paramedian widows peak [35].
3. The cleft may descend and pass through the intersection of the nasal skin and alar skin and results in an alar cartilage cleft [46].

Skeletal involvement:

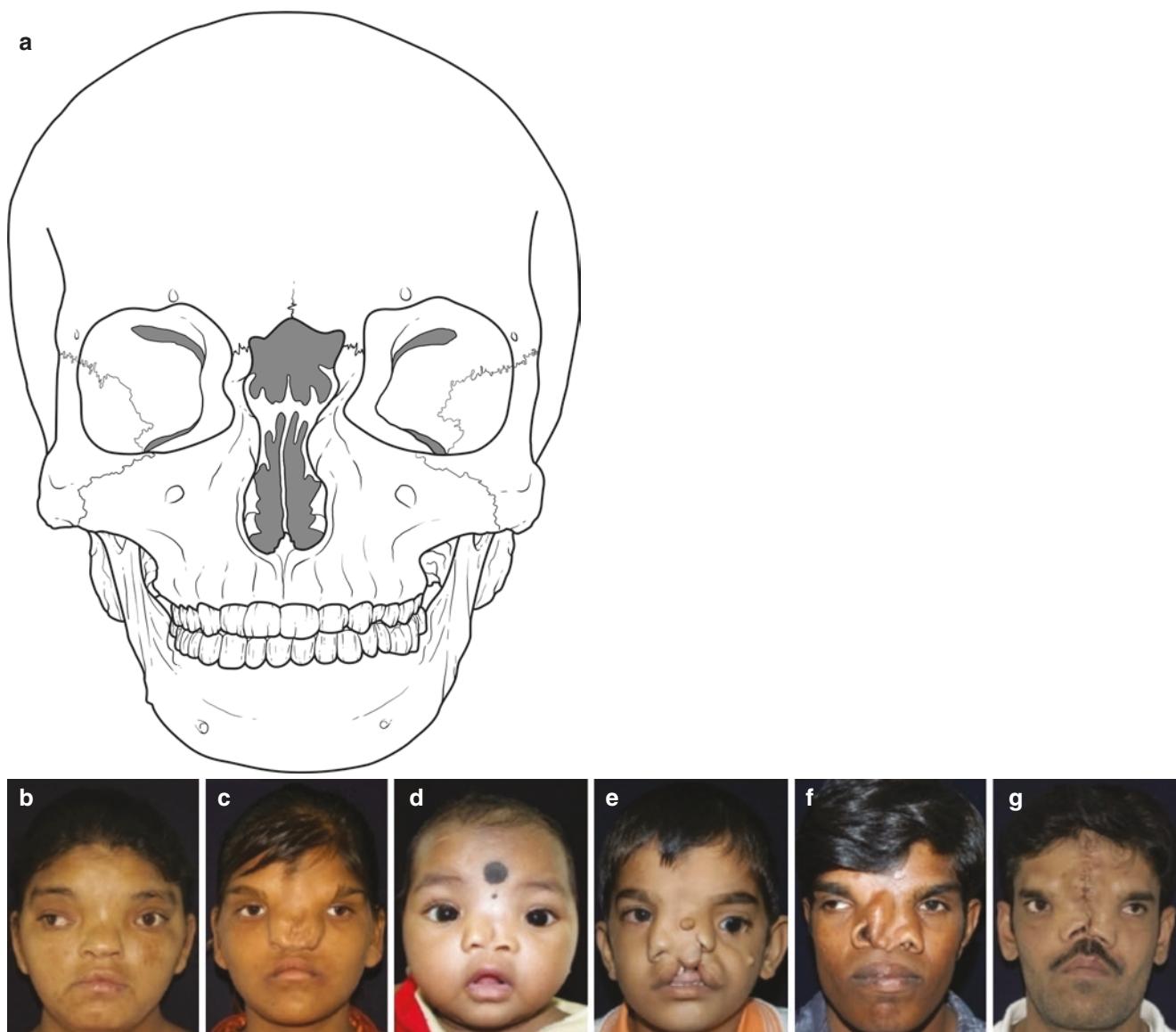
1. Deformities of the medial aspect of the cribriform plate [46].
2. There may be olfactory groove widening with associated widening of the cribriform plate and the ethmoid sinus with resultant hypertelorism.
3. Paramedian frontal encephalocele may cause inferior displacement of the cribriform plate with orbital dystopia.
4. Unilateral and bilateral types of the number 13 cleft exist with bilateral demonstrating the most extreme forms of hypertelorism [4].

77.5.1.15 Number 14 Cleft

No. 14 is the cranial congener of the cleft no. 0, which is the median craniofacial dysraphia. The terms frontonasal and frontonasoeethmoid dysplasia were utilized by Van der Meulen for this group of deformities [49] (Fig. 77.18a–g).

Soft tissue characteristics:

1. Severe lateral displacement of the medial canthal apparatus bilaterally.
2. Glabellar flattening.
3. The periorbita including the eyelids and eyebrows is generally normal.



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Fig. 77.18 (a–g) Tessier number 14 with variable degree of nasal duplication and hypertelorism

4. Midline projection of the frontal hairline [35].
5. There is distortion of the nasal capsule and the developing forebrain stays in a lower position.

Skeletal characteristics:

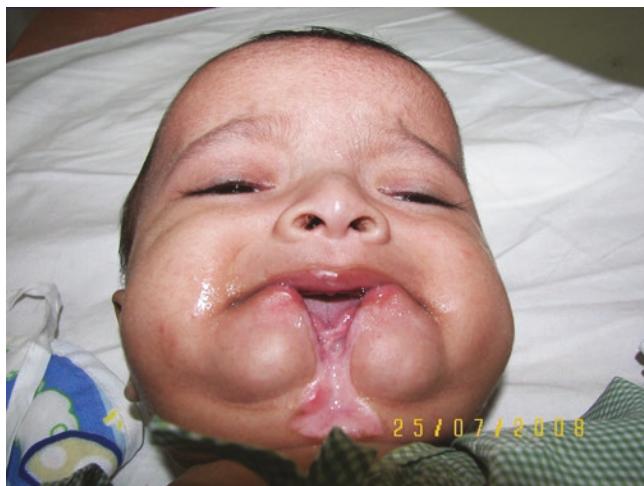
1. Severe hypertelorism.
2. Frontal encephalocele may be seen herniating through the midline frontal bone defect.
3. Caudal flattening of the frontal bone is flattened with flattened glabellar region
4. Absence of frontal sinus pneumatization.
5. The crista galli and the perpendicular plate of the ethmoid are bifid, and there is an expanded separation between the olfactory grooves [35].
6. The crista galli and ethmoids are widened with caudal dislodgement.
7. There may be a shortening of the middle cranial fossa.
8. The anterior cranial fossa is tilted upward, producing a harlequin eye disfigurement on plain radiographs [35].

77.5.1.16 Number 30 Cleft

Tessier 30 cleft otherwise known as lower midline facial cleft or median mandibular cleft is a rarity (Fig. 77.19). Median cleft of the lower jaw was first described in 1819 by Couronne [30].

It is generally constrained to a deformity in the soft tissue of the lower lip.

In its severe form, it may groove or split the mandibular symphysis and at times involve the midline structures of the



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Fig. 77.19 Tessier no 30

neck including the hyoid bone, the thyroid, and even the strap muscles. The anterior part of the tongue may be bifid, showing ankyloglossia [54], with one case of absence of tongue being reported [55].

77.6 Treatment of Craniofacial Clefts

Surgery for facial deformities involves the best balance of art and science. The restoration of the malformed anatomy requires artistic creativity, while the science lies in the reestablishment of impaired function [57].

From a careful perspective, even microform clefts might deform. Contingent upon the level of distortion, an arranged strategy has been viewed as the treatment of choice [58]. Due to their multifaceted nature; the individual level of cleft involves successful reconstruction and the rehabilitation in practically every one of the cases request multistep and multi-proficient procedure [59]. Besides cautious examination, imaging methods are important to evaluate the individual level of skeletal inclusion. For correct determination current imaging frameworks seen in systems, for example, CT, MRI, and 3D CT, permit better preoperative comprehension of the issue and planning of the surgeries. Analysis ought to be founded on a classification relying upon the site and types of defects (morphology) which helps in foundation of a legitimate treatment plan [60] (Figs. 77.20, 77.21, and 77.22a–e).

Institutionalized treatment plans are not constantly conceivable in light of the assortment of craniofacial clefts and dimensions of seriousness. Be that as it may, core values are useful in deciding the best possible planning and stages for restorative surgery [30]. The Australian Craniofacial Unit Treatment Protocol, which is a pioneering center with international acclaim, has recommended the following:

- Early repair of the soft tissue defects and preventing exposure keratitis
- Orthodontic intervention interceptive orthodontics and speech therapy for the school-going years (4–10 years)
- Reconstruction of the orbital floor, orthognathic surgery, and rhinoplasty to be done after growth completion [30]

77.6.1 Tessier No. 0–14 Cleft

Literature reveals that the midline Tessier 0 and 14 clefts are among the most common encountered [5], while the combination of 0 and 14 is the most common combination of non-isolated clefts [61].

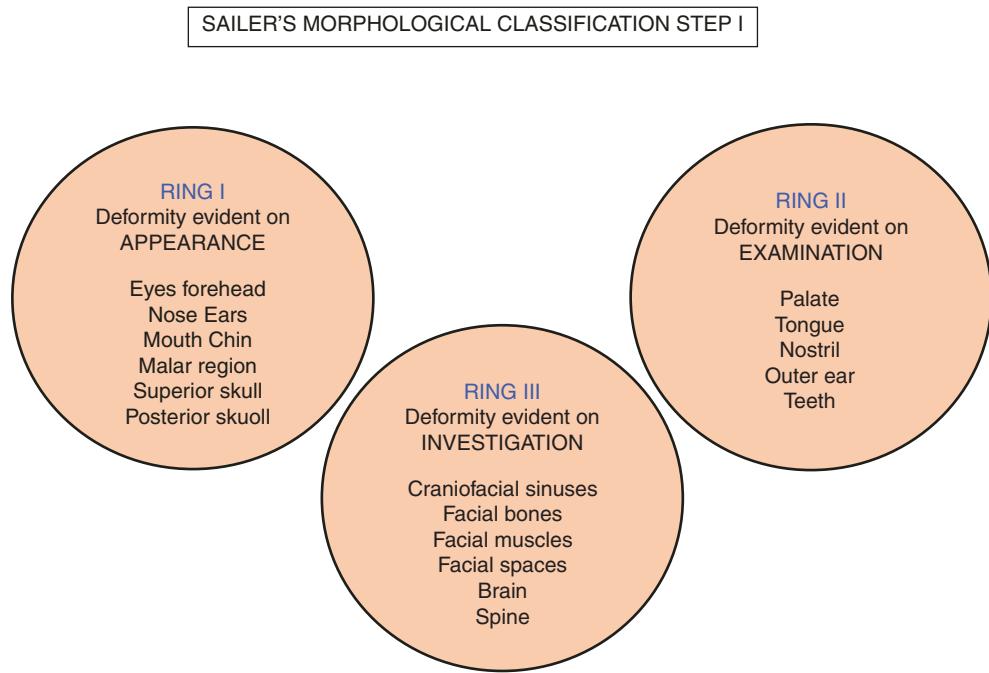
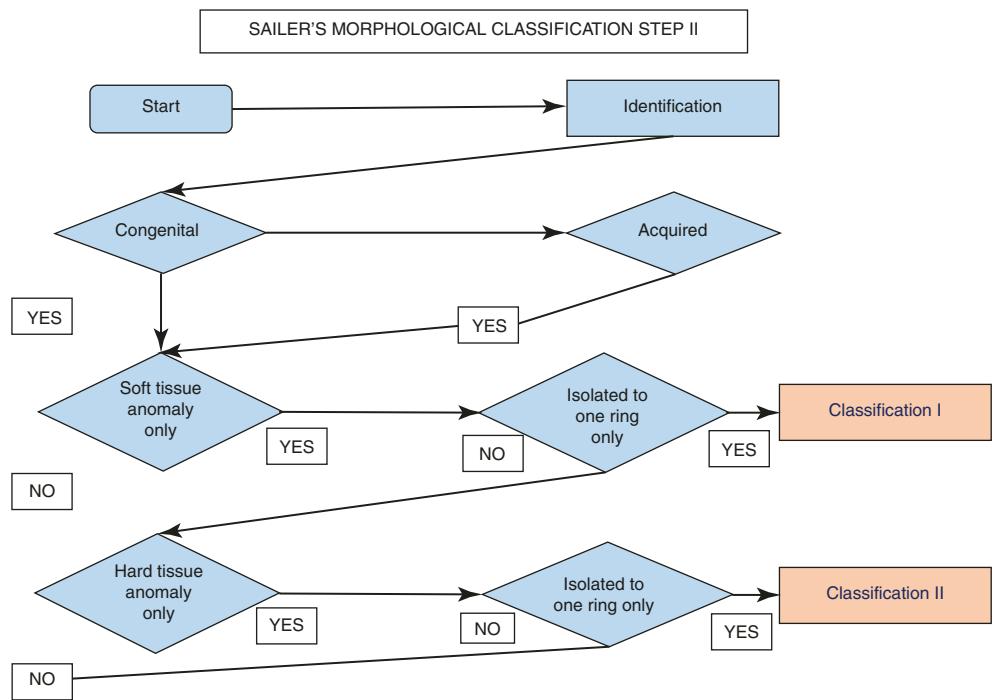
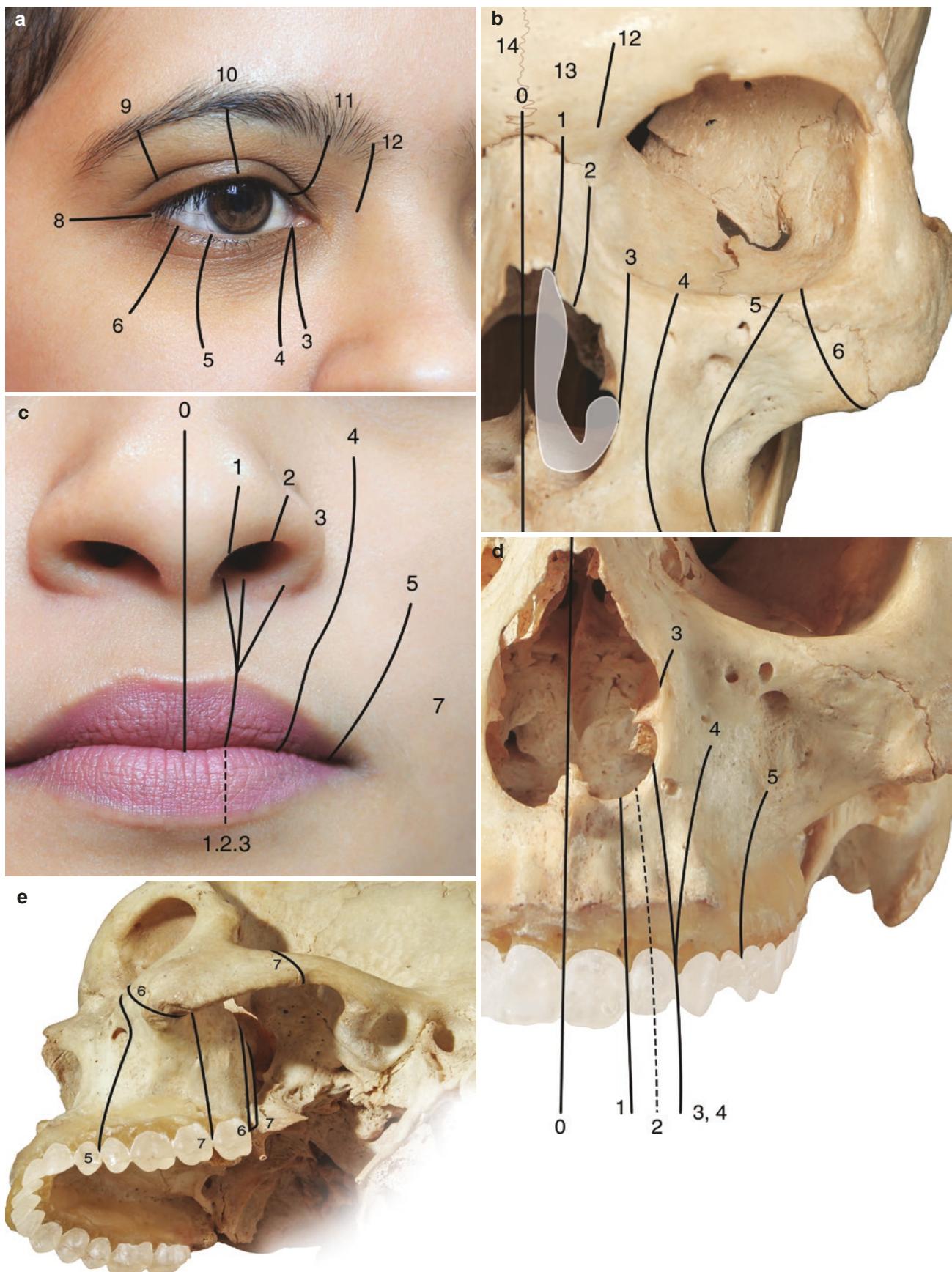


Fig. 77.20 Sailer's morphological classification step I

Fig. 77.21 Sailer's morphological classification step II





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Fig. 77.22 (a–e) Anatomical classification of facial, craniocalvarial clefting

The most commonly adapted protocol for Tessier no. 0–14 is modified from surgical protocol for midline Tessier 0–14 craniofacial clefts (David 2006) (Fig. 77.23).

As per Sailer's morphological classification, Tessier 0–14 clefts involve:

- (i) The medial orbital zone
- (ii) The zone of the maxilla, oral cavity, and lips
- (iii) The zone of the nose

Treatment is generally formulated based on the degree of involvement of these zones (Figs. 77.20 and 77.21).

77.6.2 The Orbital Zone and Skull Bone Defect

77.6.2.1 Resection of Encephalocele

An encephalocele is a herniation of a part of the cerebral matter through a deformity in the skull. This may contain meninges (meningocele), or meninges, cerebrum, and ventricle (meningoencephalocystocele) [61].

Tessier no. 0–14 is most generally connected with fronto-ethmoidal gathering of encephaloceles group which can be subdivided into nasofrontal, nasoethmoidal, and nasoorbital types.

The objectives of repair involve:

1. Meticulous repair of skin deformities. This helps prevent contamination and desiccation of brain tissue.

2. Water-tight dural repair with removal or invagination of the non-functional brain tissue which is present extra-cranially.
3. Complete craniofacial reconstruction of skeletal components. Care taken to prevent appearance of a long nose [62].

77.6.3 Orbital Hypertelorism

Hypertelorism is the most common indication for major craniofacial correction in a Tessier 0–14. Treatment strategies are varied and range from medialization of the medial wall of the orbits to total repositioning of the orbital and facial bipartition [63]. Hypertelorism is a physical finding in many craniofacial malformations, which is characterized by an increase in interorbital distance. It may be a part of a syndrome but it is not a syndrome by itself.

In 1924, Greig called orbital hypertelorism as “ox-eyed” and also coined the term “ocular hypertelorism” [64]. The more accurate term of “orbital hypertelorism” to denote true lateralization of the orbital complex was coined by Tessier in 1972 [65]. Tessier classified hypertelorism into three degrees based on the interorbital distance [65, 66] (Fig. 77.24).

77.6.3.1 En Bloc Osteotomies (Fig. 77.25a–d)

Radical mobilization of the orbits to correct increased interorbital distance is one of the most challenging procedures in craniofacial surgery [67].

Paul Tessier was the first to perform orbital mobilization using a trans-cranial approach [53]. This surgical intervention was planned to eliminate undue risk to the

Fig. 77.23 Surgical protocol for midline Tessier 0–14 craniofacial clefts

Adapted Surgical Protocol for Midline 0–14 Tessier Craniofacial Clefts (David, 2006) [16, 17]

Birth to 1 year

- Preserve the essential functions of the airway, feeding, sight, and hearing
- Repair any encephaloceles
- Craniosynostosis correction by fronto-orbital advancement at 3–6 months

5 years

- Orbital box osteotomy once the tooth buds can be reliably preserved
- As an alternative, a medial faciometry (facial bipartition) can be performed slightly later
- Possible use of tissue expanders beneath the zygomatic periosteum in extreme hypertelorism
- Temporary nasal reconstruction, as this will not be repeated later

5 to 10 years

- Orthodontic management is commenced when enough permanent teeth are present
- Alveolar bone grafting is carried out to correct the more extensive defects in the maxilla that may be present

10 years until the completion of growth

- Maintenance of the orthodontic therapy with retainers
- Further preservation of the eyes and hearing if required

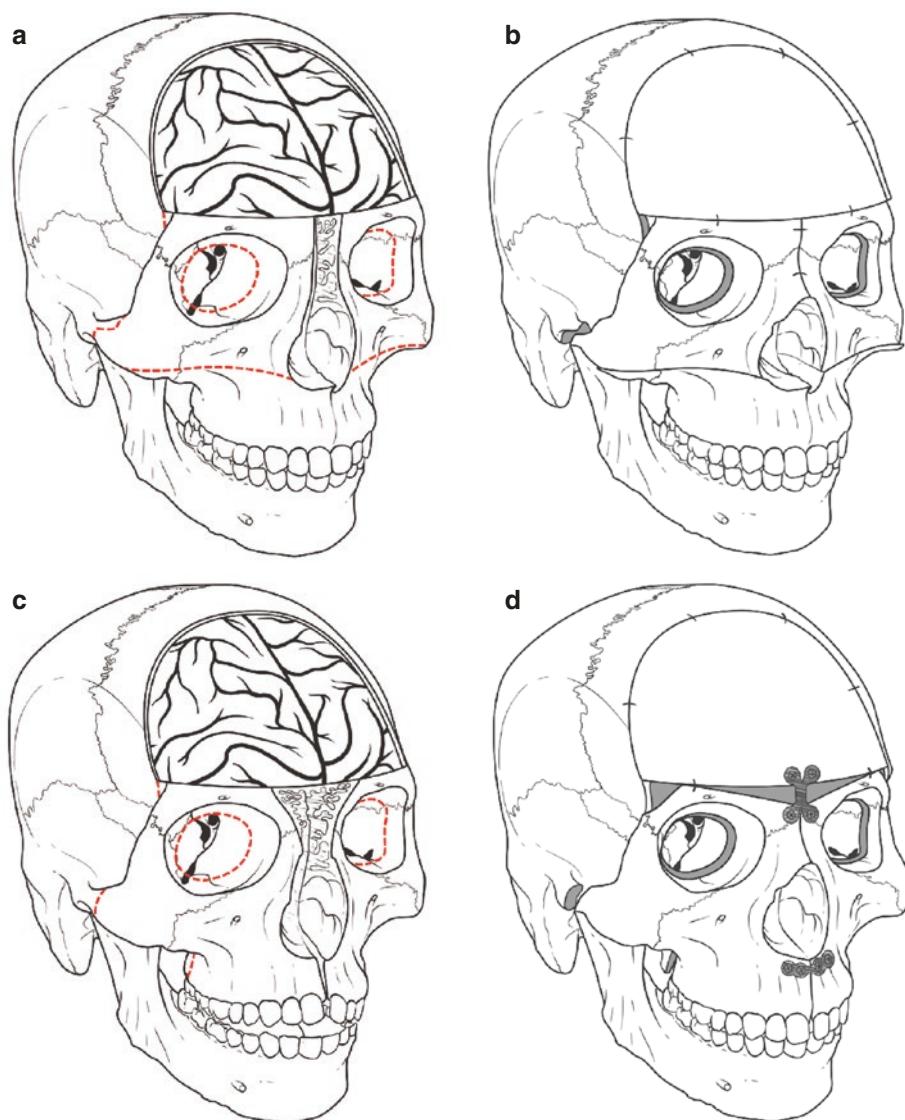
Completion of growth

- Orthodontic preparation for midface surgery
- Midface surgery performed at any level, with or without bone grafting/mandibuloar surgery depending on need
- After establishment of the facial platform, definitive nasal reconstruction is performed
- Further secondary surgery is performed if necessary

| HYPERTELORISM | | |
|---------------|----------|----------------------------|
| | Degree | Interorbital distance (mm) |
| I. | Slight | 30 to 34 |
| II. | Moderate | 35 to 39 |
| III. | Severe | 40 + |

Fig. 77.24 Hypertelorism

optic nerve [65]. Converse described in 1968 preservation of the olfactory nerves by performing subcranial U-shaped osteotomy [68]. Schmid described the extra-cranial circumferential orbital osteotomies for medializing the orbits [68], while Jacques van der Meulen described the facial bipartition in 1983 [69]. Medialization of the medial orbital walls and hemifacial rotation do not interrupt mid-facial growth and thus were performed before age 5 in the majority of patients. Orbital translocation causes growth disturbances and thus is to be performed after attaining skeletal maturity [63].



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Fig. 77.25 En bloc osteotomies (a–d)



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Fig. 77.26 Stereolithographic models—simulation

Pre-op Evaluation

Pre-op evaluation for orbital hypertelorism includes the following:

External facial measurements

Dental casts

Posteroanterior x-ray cephalometry [70]

Modern aids like computed tomographic scan

Three-dimensional imaging and stereolithographic models [71] (Fig. 77.26)

Facial Bipartition

The facial bipartition procedure involves the mobilization and rotation of the entire midface in a monobloc fashion (Fig. 77.27a, b). The osteotomy involves the supra-orbital rims first. It is then continued along the lateral orbital walls in a similar fashion to the LeFort 3 osteotomy and is dropped caudally to involve the dentate segment also in the mobilized segment. The intra-orbital circumferential osteotomies are performed using a combined trans-cranial and trans-facial approach once the hemifacial segments are mobilized, the intervening ethmoids are resected creating space for the upper face to be rotated medially. This causes the maxillary dentate segment and the palate to have a lateral rotation increasing the transverse dimension of the face and correct-

ing the palatal crossbite of the upper posterior teeth. This procedure can also be combined with an advancement of the midface complex by combining it with a LeFort 3 type modification [72].

77.6.3.2 Box Osteotomy (Fig. 77.28a–d)

Correction of orbital hypertelorism done using a box osteotomy may include corrections of associated nasal deformities. Bone and cartilage grafts may be necessary to provide nasal framework. Skin grafts may be required for nasal coverage and may be accomplished by local flaps. The box osteotomy is generally preferred when the dental occlusion is normal.

Van den Elzen et al. (2011) advocated waiting until after age 10 (after eruption of permanent dentition) to perform orbital box osteotomy. Monasterio and Taylor [61] supported the use of orbital translocation after skeletal maturity and have stated that early intervention retards midfacial growth. Tessier (1973) noted that neuro-ophthalmological benefit was increased by correcting hypertelorism at 3 years and psychosocial benefit was heightened by operating at 6 years, before schooling begins, or after 12 years to preserve the dentition. Marchac et al. (1999) also described the use of box osteotomies after 12 years of age.

77.6.3.3 Spectacle Osteotomy (Fig. 77.29a–e)

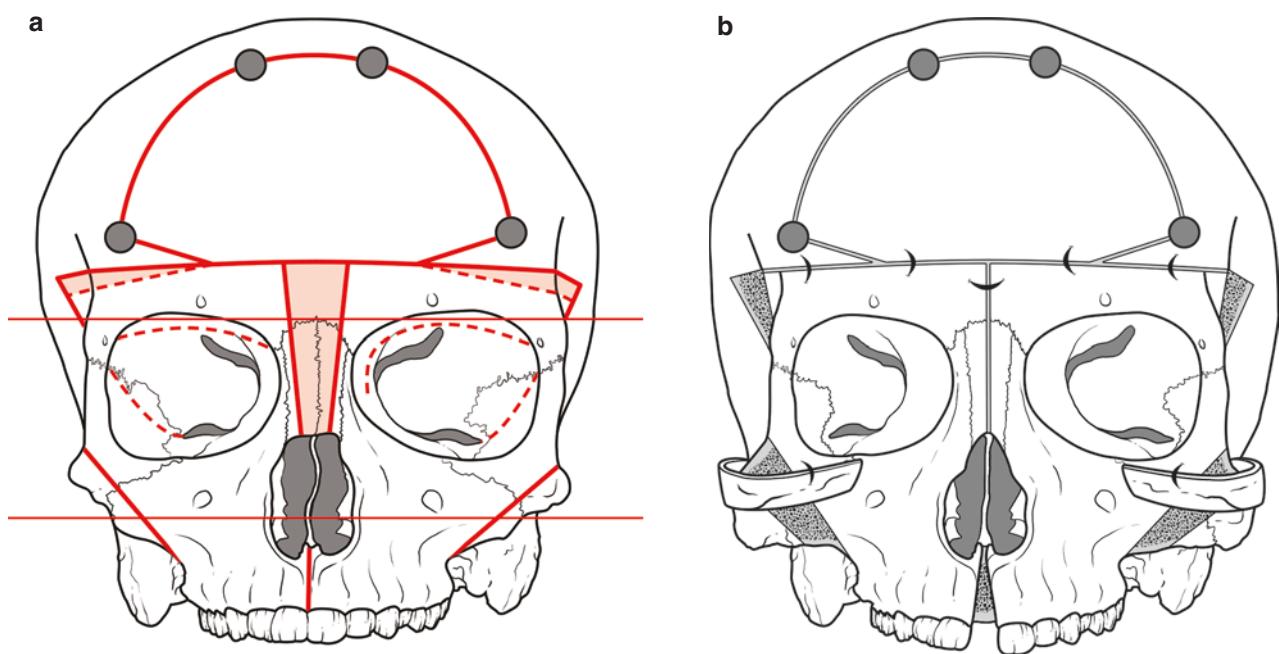
The spectacle osteotomy is done by performing the trans-frontal craniotomy and preservation of frontal bandeau along with trimming of the bone in the periorbital region and around the pyriform aperture with *medialization* of the orbits.

A lateral canthopexy is an integral part of this procedure [48]. The medial canthi should be recognized and anchored using trans-nasal wires. At times this is performed with a mini-plate anchor for orienting the wires in the right direction [72]. The plate is secured to the thick nasal bone, and the lower hole is kept at the level of the lacrimal crest. The canthi are independently fixed to these holes bilaterally utilizing steel wires (Also refer Chap. 79 on craniofacial syndromes) [73].

77.6.3.4 Soft Tissue Management

The midline cleft lip notch can be successfully treated by holding fast to three noteworthy standards (Fig. 77.30a, b):

1. Excision of the constrictive band on the lip
2. Approximation of the split orbicularis oris muscle at the midline
3. Mucosal lengthening using “Z”-plastics [36, 42, 74–76]



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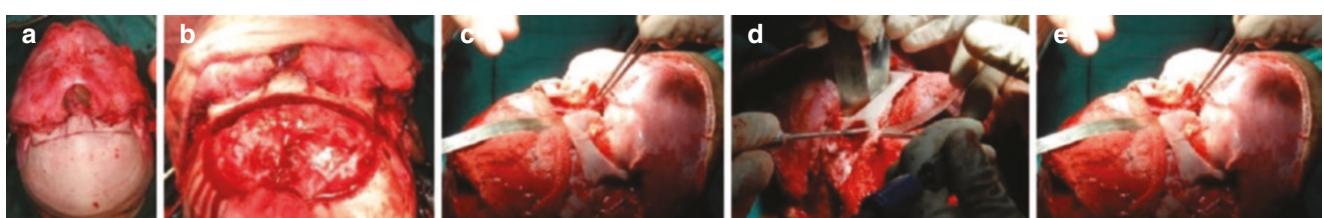
Fig. 77.27 (a, b) Facial bipartition



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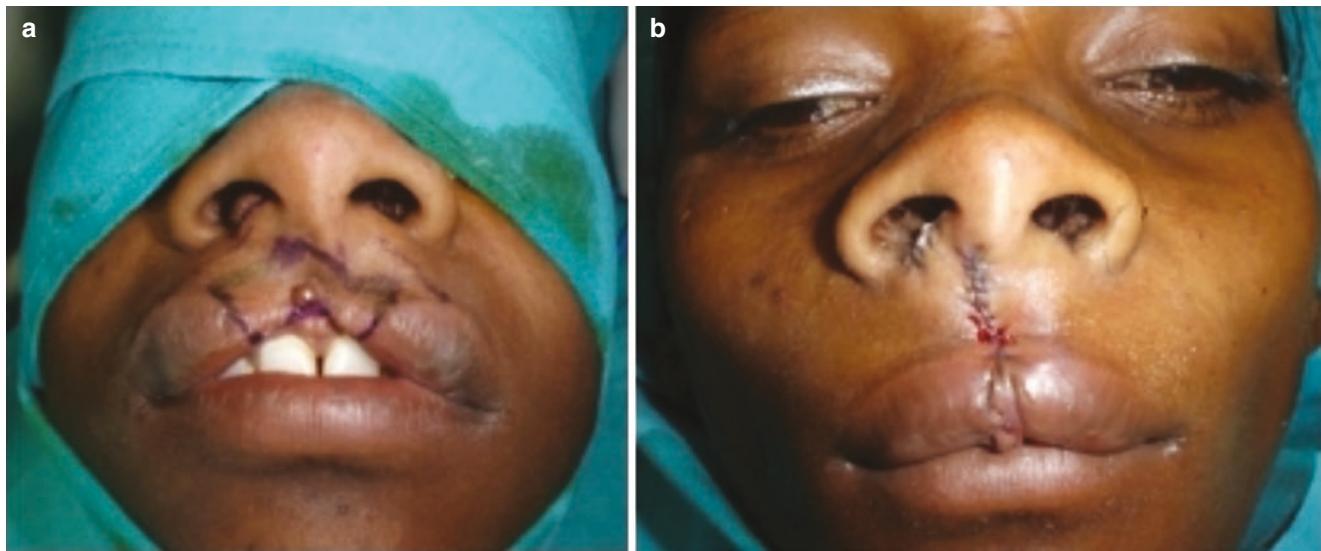
Fig. 77.28 (a–d) Box osteotomy—skin incision for the intracranial correction of the orbital hypertelorism consists of bicoronal incision, transfrontal craniotomy sparing the frontal bar, periorbital osteotomy,

block bone removed near pyriform area, calvarial bone graft, miniplate fixation and closure



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Fig. 77.29 (a–e) Spectacle osteotomy



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Fig. 77.30 (a, b) Reconstruction of Cupid's bow and philtrum



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Fig. 77.31 (a, b) Resection of the excess skin of the nasal dorsum and closing in the midline

In managing the 0–14 cleft, the clinician should be able to diagnose if it is a true or a pseudo occurrence. In cases of a true midline cleft, care should be taken to reconstitute the important components of the lip, external nose, and nasal septum. Resection of excess skin also needs to be planned and performed [36] (Figs. 77.31a, b and 77.32a, b).

Nasal Clefts

Nasal clefts include Tessier no. 2 and Tessier no. 3 clefts.

Principles of surgical management (Fig. 77.33). Because of their rarity and extreme variability, it is not surprising that *standardized* methods of correction have not been established for patients with nasal clefts. Therefore, each case



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Fig. 77.32 (a, b) Reconstruction of the lip and resection of the excess skin of the nasal dorsum and closing in the midline

Principles of Management of Complex Nasal Clefts

- Precise Clinical and Radiographic Diagnosis
- Physical examination
- CT/MRI

Inventory of Nasal Components

- Skin
- support

 - Cartilage
 - Bone

- Lining
- Staged Reconstruction
- First things first
- Do not burn bridges
- Replace Like with Like
- Re-establish Nasal/Facial Esthetics

Fig. 77.33 Principles of management of complex nasal clefts

must be approached on an individual basis using the basic principles of nasal reconstruction as a foundation.

The fundamental principles and techniques of nasal reconstruction are to assess what is present, what is missing, utilization of available tissue, and importing required tissue as indicated by the deformity.

Inventory of Nasal Components

Each component of the nose, skin, lining, and support (bony and cartilaginous), must be evaluated in a quantitative and qualitative manner. The quality and quantity of external skin and nasal lining are evaluated for deficiency or excess. The integrity of the cartilaginous framework, nasal septum, has to be assessed.

Staged Reconstruction

Patients with complex craniofacial clefts frequently require multiple staged reconstructive procedures. Planning of these procedures must be tailor-made to each patient. Surgical stages must be planned and executed with all subsequent surgical sessions clearly in mind.

Replace Like with Like

Whenever possible absent or deformed tissue should be replaced with identical or similar autogenous material.

Bony defects may be present along the cleft from the alveolus to the orbit. It is essential to reestablish a stable anatomic bony base to ensure adequate support for the soft tissue reconstruction of the nose and orbit. Autogenous split calvarial, rib, and iliac graft are the general preferences.

Cartilage

Free cartilage grafts alone may often suffice to correct small deficiencies of the alar rim, nasal rim, nasal dorsum, or tip. Larger defects of the upper and lower lateral cartilages can be replaced using conchal or septal cartilage grafts. Thin bone from the perpendicular plate of the ethmoid can be used to replace upper lateral cartilage.

Small composite chondro-cutaneous grafts from the concha can be used to replace small defects if additional lining is needed. For larger cartilage defects in the area of the upper laterals, septal hinge flaps can be used. Reconstruction of lower lateral cartilage can be accomplished using conchal cartilages or chondromucosal grafts from nasal septum.

Nasal Lining

Adequate lining to the nose is critical to achieve a functional and predictable aesthetic result. This is also essential to reestablish vascularity to the underlying tissue and to minimize contracture of the soft tissues. Local turnover flaps are useful for small defects, while larger defects may require vascularized tissue transfer.

Skin

Small nasal clefts are amenable with local tissue transfer in the form of rotation flaps from the lateral nasal area. Onus in simple nasal clefts is given to the reestablishment of the alar rim contour [68]. The use of z-plasty techniques and composite grafts helps in achieving additional symmetry [77].

Downward rotation of the cephalically displaced alar rim is the first step in this process. This may require the use of a back cut to rotate the alar margin caudally. The triangular defect produced by the alar rotation is then filled by a transposition flap from adjacent areas having excess skin [78, 79].

Descriptions of a nasal dorsal rotation flap (Figs. 77.34a–d and 77.35a, b) for a Tessier 2 cleft and a brow-eyelid-nasal transposition flap for a Tessier 3 are predictable techniques [80].



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Fig. 77.34 (a–d) Design for bilateral Tessier no. 3 cleft—nasal dorsum rotational flap



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Fig. 77.35 (a, b) Nasal dorsum rotational flap

The nasal dorsum rotational flap design to correct Tessier no. 2 clefts improves alar symmetry at the cost of reduced nostril size. A notch on the affected ala is a major problem to handle in this technique [80, 81].

The forehead-eyelid-nasal transposition flap (Fig. 77.36) technique involves the use of an inter-eye-brow-forehead flap which is pedicled on the tissue of the nasal bridge. The rotation achieved by the flap gives both the alar and nostril symmetry while reducing the need for revision [80].

Reestablish Nasal/Facial Aesthetics

Optimal reconstruction of the nose should be based on the principle of aesthetic subunits as described by Gonzalez-Ulloa and Castillo (1954) and refined by Burget and Menick



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Fig. 77.36 Forehead-eyelid-nasal transposition flap

(1985). Superior results are better achieved by replacing the entire subunit of the nose rather than simply patching the defect. The achievement of the nasal proportions should be in conjunction with facial proportions though there are many aesthetic and surgical limitations because of associated severe craniofacial deformities. Definitive nasal reconstruction may need to be delayed or staged as explained, until optimal canthal, orbital, and maxillary relationships are obtained.

77.6.4 Tessier No. 4 (Fig. 77.37a, b)

Tessier no. 4 cleft is a rare, complex malformation which has severe implications on both the soft tissue and skeletal structures of the face. As a general rule, priority needs to be given for reconstructing the soft tissue envelop, and the skeletal repair should in the form of osteotomies or bone grafting procedures need to be deferred until the school-going years. This is due to the fact that early intervention to the bony skeleton may hinder the development of the midface and associated (Resnick and Kawamoto 1990; Kawamoto and Patel 1998) [76].

An important clinical indication that necessitates emergency intervention is exposure keratitis of the cornea and resultant blindness. This depends on the gravity of the cleft deformity. In narrow cases of Tessier 4 cleft, the clefting is more toward the medial side of the orbit which retains a large bony component of the lateral orbit. This lends support to the globe and enables reasonable competence of the upper eyelid to cover the cornea. In such instances the early surgical intervention may be avoided [82]. On the other hand in severe cases, there is total absence of the orbital floor causing the globe to sink downward with the cornea facing upward due to a lax supporting structure. This prevents the upper



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Fig. 77.37 (a) Incision for bilateral Tessier no. 4 (b) unilateral tessier 4 on left side with tessier 7 on right side

eyelid from being able to protect the cornea, with the resultant problem of exposure keratitis and permanent corneal damage. Such instances produce an emergency, and a repair of the orbital support system is mandated. In cases where an early intervention is not possible due to medical and anesthetic implications, emergency intervention at least in the form of a tarsorrhaphy is performed within 2–4 days of birth.

While treating severe deformities like the Tessier no. 3 and Tessier no. 4 clefts, a “split method” of management can be utilized to handle the anatomical regions individually as given below [83].

The affected regions were divided into three segments:

1. Lid segment
2. Lip segment
3. Nasomalar segment

77.6.4.1 Managing the Lid Segment

An ectropion like deformity is commonly present. This can be managed using conventional techniques. They include (a) a back-cut release of the lower eyelid to cheek junction and medial advancement of the lower eyelid with medial canthopexy, (b) lower lid resection with inferior layered tarsal strip, or augmentation of the tarsal plate with spacer grafts. However in cases where there is inadequate tissue locally, tissue must be imported into the lower eyelid for optimal results [83]. Some of the commonly used flaps for this include the median forehead flap and the nasolabial flap. However, free vascularized tissue transfer techniques also may be adopted [84].

Numerous techniques have been described for the management of the Tessier 4 cleft. But most of them result due to the interdigitating scars produce a sub-optimal outcome [33] (Tessier 1976; Kawamoto 1990). Longaker et al. (1997) proposed the superiorly based nasolabial flap that was transposed to the lower eyelid. Though this technique had an advantage cosmetically, it had that limitation that it was useful only for mild forms of the cleft. The cheek advancement technique described by Van der Meulen [4] had the advantages of being useful even for wider clefts and favorable scars along aesthetic facial subunits. Van der Meulen likewise depicted that it was significant for improving the scarring caused by anchoring the cheek flap firmly to the pyriform aperture [85]. However, despite the options available, it is better to understand that a single flap may be insufficient at times to reconstruct the eyelid [86]. To overcome these challenges, the use of tissue expanders and free vascularized transfers have also been advocated [87, 88].

The Veau III method of bilateral lip repair can be used for correcting the lip component. The last area to be addressed is the naso-malar junction [89].

77.6.5 Tessier No. 5–9 Cleft (Fig. 77.38a, b)

The primary outcomes that are needed for the correction of these clefts include:

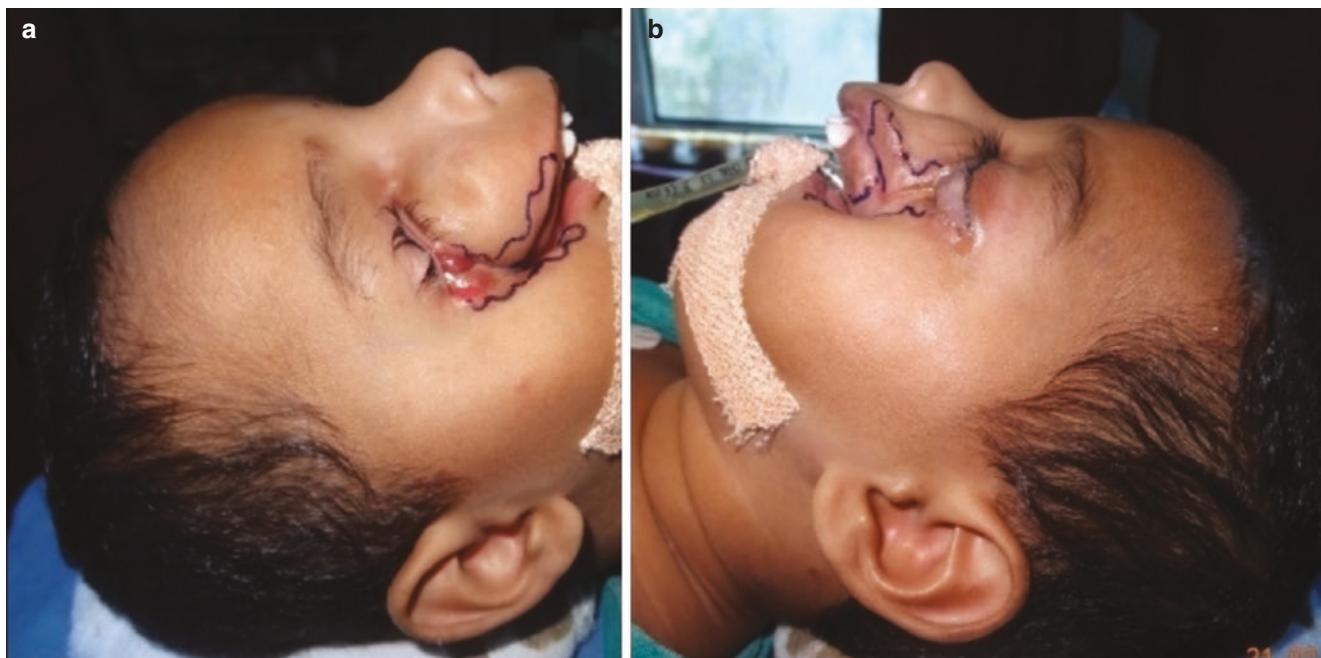
- Lower eyelid reconstruction
- Lateral canthal repositioning
- Repair of the labio-maxillary cleft
- Skeletal continuity restoration (this includes the orbital floor) using bone grafts

The principles to be borne in mind in the management of the Tessier 5–9 spectrum have been modified by different surgeons: Tessier (1971) advocated early correction of orbital dystopia – prior to 1 year of age. In all forms of facial clefts that affect the ocular region, priority is given to the preservation of vision by early interventions to prevent exposure keratitis of the cornea. Pre-surgical care includes use of ophthalmic ointments or temporary tarsorrhaphy procedures [48, 89]. Kara and Öcsel (2001) have reported the use of multiple z-plasties for the soft tissue reconstruction as early as 8th day after birth.

77.6.6 Tessier No. 7

Lateral facial cleft or Tessier no. 7 cleft is generally unilateral in presentation, though may also occur bilaterally. The bilateral form produces more gross clinical appearance with the face being amphibious in nature with an expanded mouth and infero-laterally placed commissures.

The condition is often related to syndromes of the first and second branchial arches. The cleft involves the skin, mucosa, and muscles of the oral sphincter (i.e., the orbicularis oris and the buccinator) [89]. In rare instances the cleft may involve the masseter [90]. This is a condition which may be concurrent with a lot of other deformities mandating a very thorough investigation in any patient exhibiting the classical feature of macrostomia [89].



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Fig. 77.38 (a, b) Tessier no 5–9 management

The surgical objectives for the management of the lateral facial cleft have been detailed as follows [90] (Fig. 77.39a–d):

1. Reconstituting the oral sphincter by reconstructing the orbicularis oris and the buccinator
2. Identification and reattachment of the other muscles of facial expression and the masseter in cases
3. Reconstruction of a natural and symmetrical commissure bilaterally
4. Attaining lip symmetry
5. Closure of the defect in layers with the mucosa, muscle, subcutaneous layer and skin [91]

Numerous procedures for the repair of macrostomia have been described in literature; [16, 17, 92, 93, 96, 97] Onizuka 1965; Boo-Chai 1969; Mansfield and Herbert 1972; Skoog 1974; Talukder 1980; Kaplan 1981; Bauer et al. 1982; Fukuda and Takeda 1985; Verheyden 1988; Yoshimura et al. 1992; Torkut and Coskunfirat 1997; Ono and Tateshita 1999.

Maximal care is exercised on the repair of the involved muscles with an overlapping technique [94, 95]. In severe

cases other muscles of facial expression like the buccinator and the risorius also need to be repaired (Ono and Tateshita) [93]. This ensures normalization of both form and function of the perioral region [94].

The skin repair consists of two parts: repair of the cheek wall and commissuroplasty. Commissuroplasty denotes the reconstruction of the skin mucosa junction at the corner of the lips. Care is taken to design the repair in such a way that there is no associated contracture in the post-surgical period. There are various descriptions for the commissural repair. The triangular flap repair of Ono and Tateshita [93]. This procedure produced better aesthetics due to the triangular design of the flaps. Kawai et al. in 1998 described an inferiorly based triangular flap taken close to the lower lip to minimize the re-expansion of the commissure in the long term.

Eguchi et al. used the vermillion square flap method for commissuroplasty [96].

Other popular techniques described for the skin repair of the lateral lip element include to direct linear suturing, Z-plasty [94], and W-plasty [96]. The purpose of a Z-plasty is to break the long linear scar. But it is seen that the use of multiple Z-plasty or W-plasty procedures violates the relaxed skin tension lines. Therefore, a single Z-plasty with vertical limb lying in the nasolabial crease is preferred [90].



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Fig. 77.39 (a–d) Tessier no. 7. (a) Diagram demonstrating abnormal oral aperture with distorted anatomy of orbicularis oris. (b1, b2) Clinical presentation of patient with macrostomia. (c) Incision marking. (d) Angle grafting

77.6.6.1 Bone Grafts

The establishment of normal contour of the soft tissues is never complete without the much needed skeletal support system. The skeletal reconstruction of craniofacial clefts essentially needs the use of bone grafts to fill and bridge both minor and major defects. The consolidation and take of the bone graft in the recipient site happens in two important steps: (i) the bony union of the joints between the grafted bone and the native bone and (ii) graft remodeling and creeping substitution [97]. Bone grafting can be performed at any age and is generally combined with soft tissue reconstruction [97].

Repairs performed in the early years of life can be accomplished with bone stock from the rib or iliac crest. However,

the quality of the bone supplied by the ilium is better suited for this work. The cortico-cancellous graft material can be placed as such or carried over titanium cribs. It is important to note that the general consensus for the use of non-vascularized grafts is for defects less than 6 cm [98].

- Iliac crest grafts are the most favored with a good stock of available cortico-cancellous bone. Generally the anterior iliac crest is preferred as the posterior iliac crest requires a change of position to prone and may not accommodate simultaneous work from two teams. Donor site morbidity rate for anterior iliac crest grafts is around 23% and much less for posterior iliac crest. Complications include post-operative pain, iliac or acetabular fractures or instability,

persistent hematoma, herniation of abdominal contents, vascular injury, lateral femoral cutaneous nerve injury, and contour defects along the iliac crest [17].

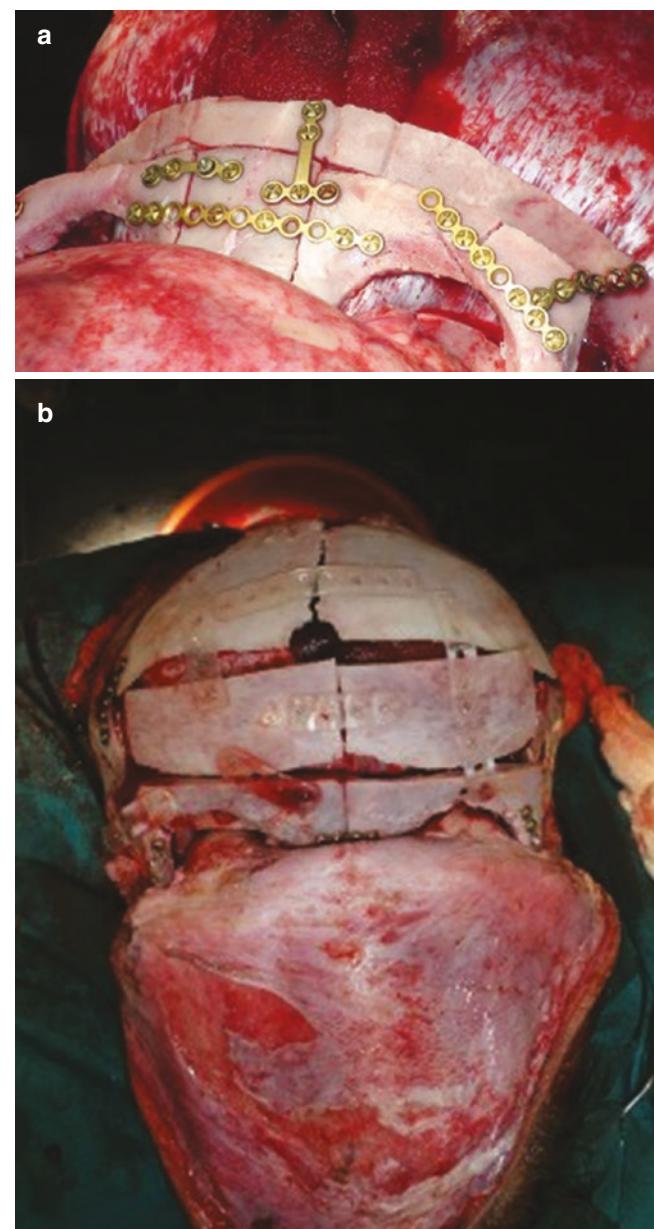
- Occasionally the use of alloplastic materials like Medpor may also be used (Fig. 77.40).
- Calvarial bone is another preferred site and can be freely available when the surgery involves cranial procedures and trans-cranial approaches. It has the lowest rate of resorption among all the bone grafts making it a preferred choice for facial and orbital reconstruction (Fig. 77.41a, b). The parietal calvarium can safely provide up to 8×10 cm of the bone for harvesting [99]. The temporo-parietal region provides more curved bone suitable for orbital or malar reconstruction. Generally the grafting is performed in strips so that there are no gross fractures and later the strips are joined together to reconstruct larger defects. Calvarium is harvested in three main forms: partial-thickness outer cortex, full-thickness outer cortex, and bicortical grafts. Partial-thickness outer cortex grafts are ideally harvested from children between 4 and 8 years of age, using an osteotome to produce a curling sheet of the bone.
- Calvarial grafts are safe in adults and are the standard indication when simultaneous craniotomy is performed. A bicortical graft is harvested and the graft is split. The inner cortex is used for reconstruction, while the outer cortex is replaced to fill the donor site defect and contour.
- Complications of calvarial grafting include contour defects of the donor site, graft fractures, and rarely dural tears while harvesting bicortical grafts. Dural tears mandate a thorough and formal dural repair to prevent further complications. Intra-cranial hemorrhage is a very rare complication during this procedure but has been documented [100].



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Fig. 77.40 Angle Medpor implant

- Grafts of ample volume can also be harvested from the anterior tibia both in the form of cancellous bone or in the form of cortical strip from the anterior tibial plateau.
- Rib bone or costochondral grafts can be harvested from ribs 5 to 7 which may be used as full-thickness or split rib grafts. Costochondral grafts are extensively used in the reconstruction of ascending mandibular ramus and condylar defects. Site-specific complications include postoperative pain, injury to the pleura with associated pneumothorax, hemothorax, or pneumohemothorax. Occasionally it may cause exaggerated facial asymmetry due to overgrowth.



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Fig. 77.41 (a, b) Calvarial bone graft



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Fig. 77.42 Distraction osteogenesis in the correction of asymmetry

Defects which are larger and demand more complex reconstruction may not be amenable to bone grafting alone and may require well-designed osteotomy techniques [82, 101] or at times, even distraction osteogenesis (introduced by McCarthy and co-workers) [102] to form better contour of the involved skeletal framework and improve function (Refer Chap. 87 on Distraction osteogenesis) (Fig. 77.42).

77.7 Conclusion

The uncommonness of craniofacial clefts has made the accumulation and complete anatomic documentation of this extensive arrangement troublesome. Preoperative and post-operative CT examinations with 3D reproductions will improve the understanding of these complex deformities. The test of managing these monstrous deformities still challenges the skill and experience of many a craniofacial surgeon [35].

This is an effort to bring a comprehensive account on the varied presentation and management techniques employed in the management of craniofacial clefts of the head and face. We also emphasize the utilization of the brilliant and time-tested diagnostic and surgical principles detailed here to establish new protocols for the comprehensive management of these deformities. Moreover we also advocate that more standardization along with structured investigation and planning methods should be utilized to come to sensible yet efficient treatment models that can be utilized by all for the treatment of this unfortunate set of people.

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