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Editorial

Surgical Strategy for the Treatment of Facial Clefts §

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§ Masterful reading (invited) by Roberto Roddi presented at the University of Turin Medical School on 26 October 2001.

1. Introduction

Craniofacial clefts have an incidence of 1/700 [1]. The cause of facial clefting is still unclear; however, there are multiple genetic and environmental factors which contribute to craniofacial development. Clefting can result from teratogens, agents that disrupt embryo development, such as radiation, maternal infection, chemicals, or drugs [1–3], and chromosomal abnormalities, genetic mutations often linked with craniofacial syndromes [4–8]. Orofacial clefts have great phenotypic diversity. Normal facial growth is directly related to the harmonious integration and interaction of the different growth centers. The arrest of development that can occur during the transformation phase can be responsible for different types of malformations in the craniofacial region [3,9–12].

The absence of fusion between the facial processes during the transformation phase can produce so-called "primary facial clefts" (facial clefts).

This variety of facial clefts can be located:

1. at the junction between the lateral nasal processes and the mid-nasal processes;
2. between the maxillary processes where the palate is formed;
3. between the maxillary and mandibular processes.

The phase of differentiation is characterized by the following phases:

1. formation of the body of the sphenoid;
2. formation of the anterior and medial cranial fossae;
3. reduction of the interorbital distance;
4. union of the two nasal halves;
5. development of the naso-maxillary complex;
6. elongation of the mandibular branch.

A deficit at the level of osteogenesis in the ossification centers is responsible for different types of malformations, which will be defined as "secondary facial clefts" (pseudo-clefts).

Different classifications have been proposed in the past by different authors.

The different types of Tessier clefts are numbered 0 to 14. These 15 different types of clefts can be sorted into four groups, based on their position: [13,14] midline clefts, paramedian clefts, orbital clefts and lateral clefts. The Tessier classification (Figure 1) describes the clefts at the soft tissue level as well as at the bone level, because it appears that soft tissue clefts can have a slightly different location on the face than the bony clefts.

1. Midline clefts

The midline clefts are Tessier number 0 ("median craniofacial dysplasia"), number 14 (frontonasal dysplasia v/d Meulen classification—see further), and number 13 ("lower midline facial cleft", also known as "median mandibular cleft"). These clefts bisect the

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face vertically through the midline. Tessier number 0 bisects the maxilla and the nose and Tessier number 14 comes between the nose and the frontal bone. The Tessier number 13 facial cleft is through the tongue, lower lip and mandible. The tongue may be absent, hypoplastic, bifid, or even duplicated. People with this condition may be tongue-tied.

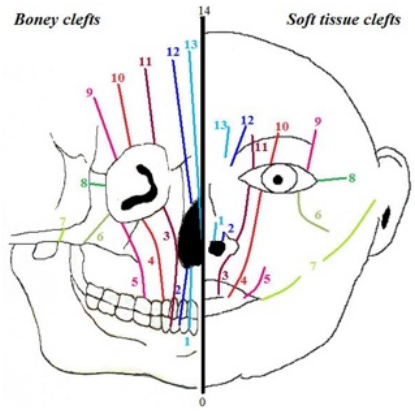


Figure 1. Tessier classification of facial clefts (1976) [14], permission obtained. Tessier classification (left boney clefts, right soft tissue clefts).

2. Paramedian clefts

Tessier numbers 1, 2, and 12 are the paramedian clefts. These clefts are quite similar to the midline clefts, but they are further away from the midline of the face. Tessier numbers 1 and 2 both come through the maxilla and the nose, in which Tessier number 2 is further from the midline (lateral) than number 1. Tessier number 12 is an extension of number 2, positioned between the nose and frontal bone, while Tessier number 13 is an extension of number 1, also running between the nose and forehead. Cleft 12 runs between the midline and the orbit.

3. Orbital clefts

Tessier numbers 3, 4, 5, 9, 10 and 11 are orbital clefts. These clefts all involve the orbit. Tessier numbers 3, 4, and 5 are positioned through the maxilla and the orbital floor. Tessier numbers 9, 10 and 11 are positioned between the upper side of the orbit and the forehead, or between the upper side of the orbit and the temple of the head. Like the other clefts, Tessier number 11 is an extension of number 3, number 10 is an extension of number 4 and number 9 is an extension of number 5.

4. Lateral clefts

The lateral clefts are the clefts which are positioned horizontally on the face. These are Tessier numbers 6, 7 and 8. Tessier number 6 runs from the orbit to the cheek bone. Tessier number 7 is positioned on the line between the corner of the mouth and the ear. A possible lateral cleft comes from the corner of the mouth towards the ear, which gives the impression that the mouth is bigger. It is also possible for the cleft to begin at the ear and run towards the mouth. Tessier number 8 runs from the outer corner of the eye towards the ear. The combination of the Tessier numbers 6, 7 and 8 is seen in Treacher Collins syndrome. Tessier number 7 is more related to hemifacial microsomia and number 8 is more related to Goldenhar syndrome.

Van der Meulen reported a morphogenetic classification of craniofacial malformations in 1983 [15] (Figure 2).

The identification of facial clefts according to both chronological and topographical bases allows us to state that all malformations can be classified and correlated to the shapes of the craniofacial region of certain stages of embryonic life. Van der Meulen classification divides different types of clefts based on where the development arrest occurs in the embryogenesis. A primary cleft can occur in an early stage of the development of the face (17 mm length of the embryo).

Figure 2. Van der Meulen classification of facial clefts (1983) [15].

Van der Meulen was the first author to use the term "Dysplasia" to define the various types of craniofacial malformations. The localization of each malformation was named using the term of the area of development involved (facial and bone processes). The spectrum of facial dysostosis is dominated by clefts that have their origin in the internasal structures: nasal, naso-maxillary, maxillary and malar (or zygomatic).

The developments arrests can be divided into five different location groups:

1. internasal;
2. nasal;
3. naso-maxillary;
4. maxillary—the maxillary location can be subdivided into median and lateral clefts;
5. malar dysplasia/or zygomatic (Treacher Collins Syndrome).

1. Internasal dysplasia

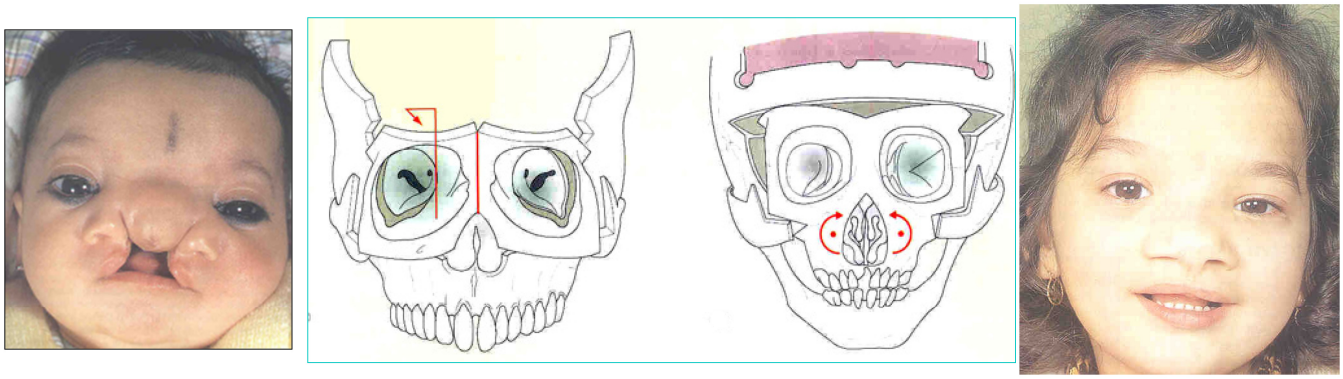
This type of malformation is also known as median facial cleft, or facial cleft No. 0 according to Tessier's classification. A large spectrum of malformations can be observed in this category, and the severity of the examples reported can be classified using sequential logic. One of the most extreme forms of this malformation syndrome is represented by the bifidity of the tip or the back of the nose, sometimes associated with the median cleft of the lip or the labial filter and Cupid's arch and the duplication of the labial frenulum. The other extreme is represented by those forms of monstrousness with separation of the two halves of the nasal valves and associated with extreme forms of hypertelorism, including all other forms of fronto-nasal-ethmoidal dysplasia (photo 2) and occasionally a trans-sphenoid encephalocele with associated herniation of the pituitary gland. The pre-maxilla is always present, contrary to what is often reported in the literature. However, the pre-maxilla may have suffered a delay in its growth, and may therefore be bifid, like the other portions of the nose. The maxilla often shows a typical vessel keel deformation, and the incisors can be rotated upwards at the two halves of the alveolar processes. A median cleft palate may also be present and often extended upwards towards the lamina cribrosa in the shape of an inverted V. Abnormalities during the closure phase of the anterior neuropore and the persistence of epithelium in the internasal area can lead to the formation of cysts or fistulas. Figure 3. Treatment of internasal dysplasia [16].

At the level of the facial skeleton:

- resection of abnormal or excessive tissues;
- correction of hypertelorism by medial rotation of the orbits;
- restoration of the nasal pyramid.

At the level of the soft parts:

- elongation and expansion of the orifices of the nasal cavities;
- reconstructions of the nasal spine.



(a) (b) (c)

Figure 3. (**a**)—Patient 6 months old before treatment. (**b**)—Surgical plan: facial bipartition (van der Meulen technique). (**c**)—4 years post-op (single procedure). Permission obtained.

2. Nasal dysplasia

This malformation is very rare. Embryologically, the nose is made up of two distinct and separate halves, and most of the nasal malformations are limited to one of the two halves. However, it is possible to make a sub-classification into four groups:

a. nasal aplasia;
b. nasal aplasia with the presence of a proboscis;
c. nasal cleft;
d. nasal duplication.

Treatment at the level of the facial skeleton:

- resection of abnormal or excessive tissues;
- correction of hypertelorism by medial rotation of the orbits;
- restoration of the nasal pyramid.

At the level of the soft parts:

- closure of the surrounding fragments of nasal mucosa by mobilizing and bringing the flaps closer;
- reconstruction of the nasal integuments [17].

3. Naso-maxillary dysplasia

This malformation is also known as cleft nose or ocular cleft No. 3 according to Tessier. This form of facial cleft can be complete or incomplete. In the incomplete form, the cleft is located at the base of the nasal wing and extends upwards in the direction of the frequently dystopian medial canthus; in the complete form, also known as oro-nose-ocular cleft, the cleft begins at the level of the upper lip as a simple cleft lip and then extends through the nasal cavities, bordering the foot of the nasal wing, which is distorted and displaced superiorly. The cleft therefore extends to the level of the medial canthus, which often presents dystopian inferiorly. A retraction of the maxilla and a deficiency or aplasia of the frontal process are often associated.

Treatment at the level of the facial skeleton:

- resection of abnormal or excessive tissues;
- correction of hypertelorism by medial rotation of the orbits;
- increase in maxillary volume.

At the level of the soft parts:

- dissection of the medial and lateral flaps of the cleft;
- approach and fixation of the muscular and mucous structures to the periosteum;
- elongation and interdigitation of the skin flaps of the cleft.

4. Maxillary dysplasia (medial/lateral/combined)—(Figures 4 and 5)

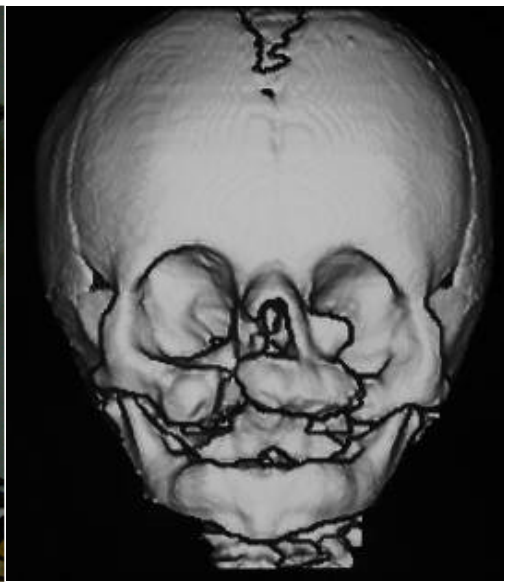
The principes of correction are similar to that of the naso-maxillary dysplasia. However, in such a condition, we are often faced with a combination of malformations affecting both sides at a different level (see Figure 4, where the maxillary dysplasia assume the median variant at the right side and the lateral variant at the left side. At the level of the facial skeleton:

- resection of abnormal or excessive tissues;
- correction of hypertelorism, when present, by medial rotation of the orbits;
- increase in maxillary volume if there is a deficit.

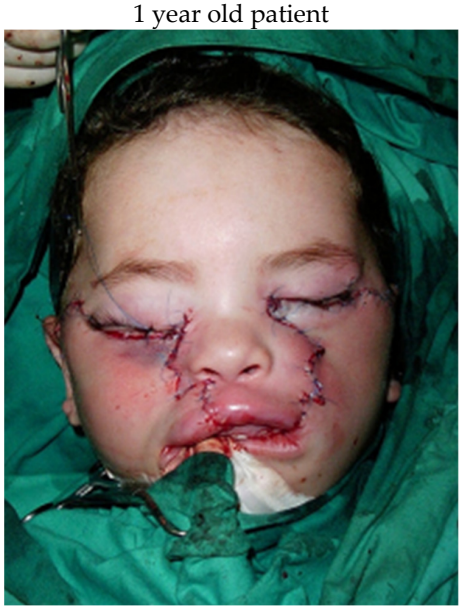
At the level of the soft parts:

- dissection of the medial and lateral flaps of the cleft; correction of the macrostomia with interdigitation of the orbicularis oris and its fixation;
- fixation of the muscular and mucous structures and to the periosteum;
- elongation and interdigitation of the skin flaps of the cleft.

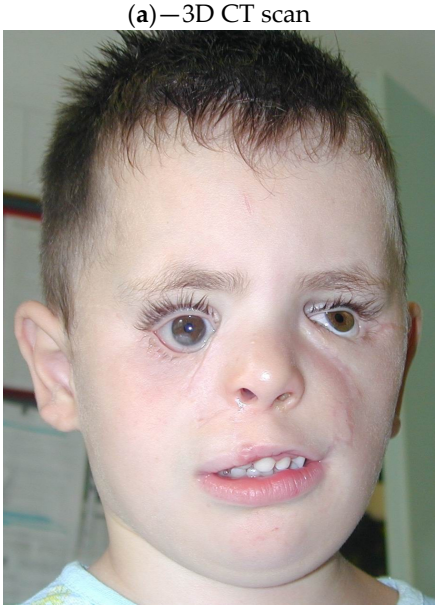
1 year old patient



(**a**)—3D CT scan

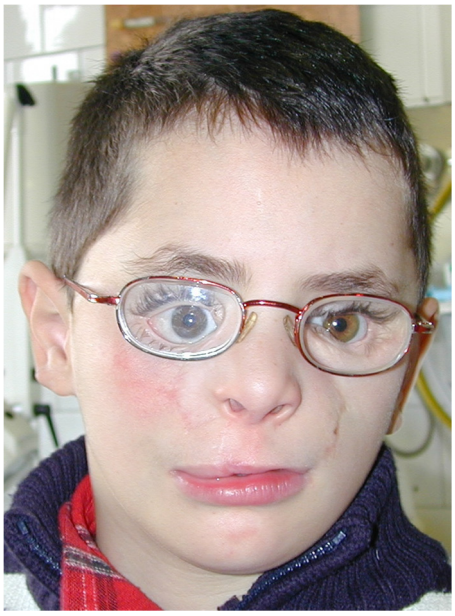


(**b**)—immediately after repair

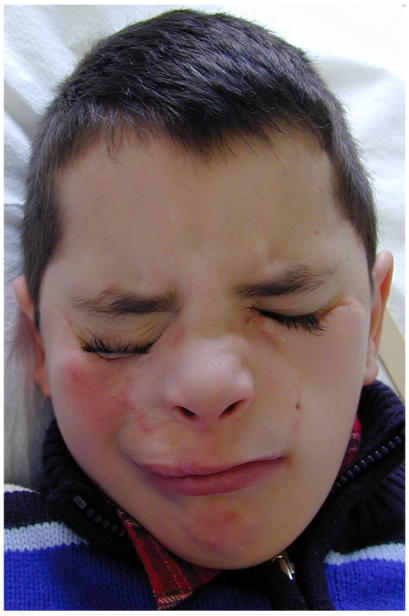


(**c**)—3 years after repair (single procedure)

Figure 4. *Cont.*

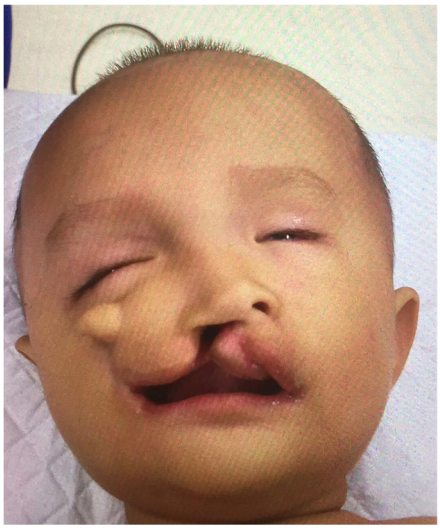


(**d**)—6 years after one single procedure



(**e**)—6 years after one single procedure

Figure 4. Maxillary dysplasia.



(**a**)

(**b**)

Figure 5. (**a**)—6-month-old patient before treatment. (**b**)—1 year after one single procedure. (Note proboscis at the right side into the cleft). Permission obtained.

5. Malar or zygomatic dysplasia

This malformation is also known as Treacher Collins syndrome or Cleft No. 7 according to Tessier classification.

Treatment at the level of the facial skeleton and of the soft parts:

Through a coronal approach, the malar defect is closed with a composite temporal bone graft inserted into the infraorbital region and fixed to the maxillary periosteum through a sub-ciliary giving the malar region its prominence. A bone graft is used for reconstruction of the lateral orbital floor and wall, lifting the eyeball, and correcting the anti-mongoloid slant. The superolateral orbital rim may be transposed medially to improve the shape of the upper orbit, creating a superior orbital ridge. Correction of the lower eyelid deficiency, and of the lateral canthal dystopia, is performed by transposition of a musculocutaneous superior V-flap and by an external canthopexy [18].

2. Surgical Strategy

The determination of the times (timing) relating to the treatment of these complex malformations is of great importance in order to obtain satisfactory results and to reduce the iatrogenic morbidity related to iterative surgical treatments, especially in those carried out in pediatric age. The ultimate goal of reconstructive surgery is to obtain an optimal result in a short period of time.

The development and experience in the field of pediatric craniofacial surgery, the best knowledge of the embryogenetic processes of the growth of the craniofacial region, and the retrospective analysis of the results related to the early treatment of these complex craniofacial malformations have established that the achievement of optimal results can only be obtained when the results are also stable over time [19–24].

Different aspects related to the treatment of facial clefts can be articulated at three levels:

- skeletal;
- muscular;
- cutaneous.

In addition, some aspects relate to:

- the determination of surgical times (timing) relating to corrections;

and finally:

- the programming of skin incisions.

2.1. Skeletal Level

The reconstruction of the facial skeleton begins by removing the abnormal structures, then carrying out the transposition of parts of the facial skeleton and carrying out bone grafting when necessary. Failures during this reconstruction phase may be due either to inadequate mobilization or fixation of facial bone segments, or to bone and/or cartilage growth deficiency or to loss of self-grafted bone tissue following resorption.

2.2. Muscular Level

The reinsertion of muscular structures must be carried out by transposing and fixing the dystopian muscular elements, which thus serve to build and maintain shape, to revive facial expressions and finally to stimulate growth. Failures in this reconstruction phase, such as those represented by canthal dystopia, are attributable to the loss of tension of the tissues resulting from their fixation and mobilization. This gradual and often incomplete loss of tension occurs during the period of tissue fragility during the healing phases; in addition, the alterations in the tissue structure are also prone to occur under the influence of external factors such as:

- contraction of subcutaneous scar tissue in the first two post-operative months;
- traction of the orbicular muscle through the canthal ligaments;
- traction generated by inadequate dissection of the periosteal tissues of the orbital structures;
- inadequate fixation of bone structures in the midline;
- inadequate exemption of the tissues contained within the orbital cavity.

2.3. Skin Level

Skin reconstruction through the transposition of flaps or autografts protects the underlying structures and preserves the facial morphology by fixing the skin to the craniofacial bone structures in the most strategic points. Failures in this reconstruction phase may be related to the eternal conflict between surgery and scar contraction. The benefits of the initial intervention can be compromised and affected by growth.

2.4. Programming of Skin Incisions

The skin incisions must be made in such a way as to respect the integrity of the facial aesthetic units such as the frontal region, the bridge of the nose or the filter, following the skin folds or furrows, and following the lines of least tension.

3. Discussion

The application of these principles is generally rewarded with good results, but failures are observable at every level of reconstruction. Surgical experience requires the surgeon is first aware of the limits of surgery; secondly, it teaches that we can speak of good results only if they are foreseeable and reproducible. Unfortunately, the boundary between doing too much and doing too little is often very small. *"The surgeon who does too much can irremediably ruin the child's face; the surgeon who does too little can ruin the soul".*

Trying to define the limits of craniofacial surgery today seems to show that the surgeons have already reached their limits at the first and second levels of reconstruction. A growth that can reconstruct the nose has not yet been reported, and the canthus that remains fixed in all circumstances is also illusory. The only opening for obtaining an optimal result lies only in the third level; that is, skin reconstruction, where the surgeon makes incisions and consequently scars. It is at this level that the surgeon must exert the most effort and where they must pay the most attention. However, scarring sequelae can be reduced by avoiding the number of surgical interventions, in optimal programming, by meticulously predicting the direction and topography of the skin incisions. Therefore, surgeons must devote the most attention to these areas. The negative effects of scarring sequelae can be counteracted by the optimal planning of the interventions, the design of the skin incisions and the optimal anchoring of the mobilized tissues. The benefits of the initial intervention can be compromised and affected by growth. The desire to correct a malformation early is understandable from many points of view. However, the extensive mobilization of skin flaps must be avoided in a child during the growth stages. Local expansion of the skin surface resulting from local reconstruction techniques and stabilization of the results through bone autograft or alloplastic grafting can help to improve morphological appearance and avoid unnecessary scars and iatrogenic damage due to iterative interventions. The repetition of these interventions may, however, preclude the surgeon from achieving the desired results and reducing the number of scars. The skin incisions must be made in such a way as to respect the integrity of the facial aesthetic units, such as the frontal region, the bridge of the nose or the filter, following the skin folds or furrows, and following the lines of least tension. The application of these principles is generally rewarded with good results, but failures are observable at every level of reconstruction.

4. Conclusions

The management of craniofacial clefts is centred on a multi-disciplinary team. The role of the patient's counsellor is very important in the team; counsellors work with patients experiencing a wide range of emotional and psychological difficulties to help them bring about effective change and/or enhance their wellbeing. The goal of treatment is to restore function, cosmesis and normal anatomical alignment of structures and cavities. Critical aspects of the management focus on eyelid reconstruction to prevent globe exposure, functional correction of macrostoma and recreating separations between nose, mouth and orbits. The timing of reconstruction is crucial.

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Conflicts of Interest: The authors declare no conflict of interest.

References

1. Leslie, E.J.; Marazita, M.L. Genetics of Cleft Lip and Cleft Palate. *Am. J. Med. Genet. C Semin. Med. Genet.* **2013**, *163*, 246–258. [CrossRef]
2. Kruszka, P.; Li, D.; Harr, M.H.; Wilson, N.R.; Swarr, D.; McCormick, E.M.; Chiavacci, R.M.; Li, M.; Martinez, A.F.; Hart, R.A.; et al. Mutations in SPECC1L, encoding sperm antigen with calponin homology and coiled-coil domains 1-like, are found in some cases of autosomal dominant Opitz G/BBB syndrome. *J. Med. Genet.* **2015**, *52*, 104–110. [CrossRef]
3. Yoon, A.J.; Pham, B.N.; Dipple, K.M. Genetic Screening in Patients with Craniofacial Malformations. *J. Pediatr. Genet.* **2016**, *5*, 220–224. [CrossRef]
4. Mayou, B.J.; Fenton, O.M. Oblique facial clefts caused by amniotic bands. *Plast. Reconstr. Surg.* **1981**, *68*, 675–681. [CrossRef] [PubMed]
5. Moore, M.H. Rare craniofacial clefts. *J. Craniofac. Surg.* **1996**, *7*, 408–411. [CrossRef]
6. Roosenboom, J.; Hens, G.; Mattern, B.C.; Shriver, M.D.; Claes, P. Exploring the Underlying Genetics of Craniofacial Morphology through Various Sources of Knowledge. *BioMed Res. Int.* **2016**, *2016*, 3054578. [CrossRef] [PubMed]
7. Weinberg, S.M.; Cornell, R.; Leslie, E.J. Craniofacial Genetics: Where Have We Been and Where Are We Going? *PLoS Genet.* **2018**, *14*, e1007438. [CrossRef]
8. Wilkie, A.O.; Morriss-Kay, G.M. Genetics of craniofacial development and malformation. *Nat. Rev. Genet.* **2001**, *2*, 458–468. [CrossRef] [PubMed]
9. Cohen, M.M. Malformations of the craniofacial region: Evolutionary, embryonic, genetic, and clinical perspectives. *Am. J. Med. Genet.* **2002**, *115*, 245–268. [CrossRef]
10. Dixon, M.J.; Marazita, M.L.; Beaty, T.H.; Murray, J.C. Cleft lip and palate: Synthesizing genetic and environmental influences. *Nat. Rev. Genet.* **2011**, *12*, 167–178. [CrossRef] [PubMed]
11. Fogh-Andersen, P. Genetic and Non-Genetic Factors in the Etiology of Facial Clefts. *Scand. J. Plast. Reconstruct. Surg. Hand Surg.* **1967**, *1*, 22–29. [CrossRef]
12. Hunt, J.A.; Hobar, P.C. Common Craniofacial Anomalies: Facial Clefts and Encephaloceles. *Plast. Reconstr. Surg.* **2003**, *112*, 606–616. [CrossRef] [PubMed]
13. Fearon, J.A. Rare Craniofacial Clefts: A surgical Classification. *J. Craniofac. Surg.* **2008**, *19*, 110–112. [CrossRef] [PubMed]
14. Tessier, P. Anatomical classification of facial, cranio-facial and latero-facial clefts. *J. Maxillofac. Surg.* **1976**, *4*, 69–92. [CrossRef]
15. Van der Meulen, J.C.H.; Vaandrager, J.M. Facial Clefts. *World J. Surg.* **1989**, *13*, 373–383. [CrossRef] [PubMed]
16. Van der Meulen Jacques, C.H.; Gilbert, P.; Roddi, R. Orbital surgery. In *Ocular Plastic Surgery*; Mosby-Wolfe: Barcelona, Spain, 1996.
17. Roddi, R.; van der Meulen Jacques, C.H. Encephalocele. In *Ocular Plastic Surgery*; Mosby-Wolfe: Barcelona, Spain, 1996.
18. Roddi, R.; van der Meulen Jacques, C.H. Treacher Collins. In *Ocular Plastic Surgery*; Mosby-Wolfe: Barcelona, Spain, 1996.
19. Friede, H.; Johanson, B. Adolescent Facial Morphology of Early Bone-Grafted Cleft Lip and Palate Patients. *Scand. J. Plast. Reconstruct. Surg. Hand Surg.* **1982**, *16*, 41–53. [CrossRef] [PubMed]
20. Kamerer, D.B.; Caparosa, R.J. Temporal Bone Encephalocele: Diagnosis and treatment. *Laryngoscope* **1982**, *92*, 878–882. [CrossRef]
21. Marchac, D.; Arnaud, E. Midface surgery from Tessier to distraction. *Childs Nerv. Syst.* **1999**, *15*, 681–694. [CrossRef]
22. Patipa, M.; Wilkins, R.B.; Guelzow, K.W. Surgical management of congenital eyelid coloboma. *Ophthalmic. Surg.* **1982**, *13*, 212–216. [CrossRef]
23. Tabrizi, R.; Ozkan, T.B.; Mohammadinejad, C.; Minaee, N. Orbital Floor Reconstruction. *J. Craniofac. Surg.* **2010**, *21*, 1142–1146. [CrossRef]
24. Tardy, M.E., Jr.; Denneny, J., III; Fritsch, M.H. The Versatile Cartilage Autograft in Reconstruction of the Nose and Face. *Laryngoscope* **1985**, *95*, 523–533. [CrossRef] [PubMed]