Rare Cranio-Facial clefts
1. **Introduction.**

Cranio-facial clefts are a wide spectrum of malformations affecting the face and cranium in a great variety of forms. The low frequency of most of them has made its study, statistics and classification complex and uncertain for many, many years.

Clefts in the cranio-facial region range from the most commonly known cleft lip and palate to extensive cranio-facial clefts that can be dramatically disfiguring.

Facial clefts constitute the most challenging malformation as they are never the same. The surgeon must be skilful in cranio-facial surgery, maxillofacial techniques, soft tissue procedures for soft tissue reconstruction, and no less important, have a solid background in management of cranio-facial anomalies. Despite all this, restoring, functional and aesthetically, a clefted face is one of the most rewarding surgeries.

Brief history of classification efforts to bring “order out of chaos” (in words of Henry Kawamoto), description of the clefts and principles for surgery will be discussed in this chapter.

2. **Classification of cranio-facial clefts.**

Cranio-facial malformations, and between them, facial clefts, were first describe by doctors, anathomist, genetist, etc, etc. The general rule was to nominate each malformation with its own name, been the case of the same or similar malformation known with various names at the same time (Sdme of Franceschetti, Goldenhar, 1st and 2nd Brachyal sdmes., Treacher-Collins, Apert, Crouzon, etc, etc). After interest for this pathology has grown, several attempts to classified them had been carried out.

**Morian Classification**

Morian, in 1887, was the first to be credited for presenting a classification for Facial clefts. He described two types, in which the reference was the infraorbital foramen.
• Type I, the oculonasal clefts, in which clefts occupied the region between the infraorbital foramen and the middle line of the face, and,
• Type II, from the infraorbital foramen to the outer aspect of the face.

**AACPR Classification**

In 1962, the American Association of Cleft Palate Rehabilitation (AACPR), described a new classification for facial clefts and syndromes and divided them in four major groups:

- **1st Group**: Mandibular process clefts.
- **2nd Group**: Naso-ocular clefts.
- **3rd Group**: Oro-ocular clefts.
- **4th Group**: Oro-auricular clefts.

This classification does not include major midline facial clefts, Treacher-Collins Syndrome and does not integrate the underlying bone defects. So it was an incomplete classification.

**Boo-Chai Classification**

The deficiencies of previous classifications were clear for Boo-Chai, and based on the classification already described by Morian, amplified the description of oro-ocular clefts, by subdividing them into types I or II.

**Karfik Classification**

In 1966 Karfik was to consider for the first time embryological and morphological aspects of the cranio-facial deformities. He described five groups:

- **A group**: Rhinencephalic disorders
- **B group**: 1st and 2nd arch brachial disorders
- **C group**: Ophthalmo-orbital disorders
- **D group**: Cranio-cephalic disorders
- **E group**: Atypical Facial disorders

Other classifications were made by various authors such as Lund, DeMeyer, etc., but the leading of all was made by Paul Tessier, he presented it in 1973 during the 1st international congress of Cleft Palate, and was later popularized by Kawamoto in 1976.

**Tessier Classification**

This classification is the generally accepted one for the description of all the cranio-facial clefts. It represents an orderly anatomic classification system in which all the clefts, major and minor and despite its position are considered and numbered from 0 to 14 with a number 30 for a medial symphysis in the mandible. This system significantly simplifies the nomenclature of clefts. It is a purely descriptive system not related to embryological or pathological factors.
The system followed by Tessier is centred on the orbit, with facial clefts numbered from 0 to 7 and the cranial clefts from 8 to 14 in a counter clockwise rotation, being the 30 in the midline of the mandibular symphysis. The orbit is the central point of the schema and separates the facial structure from the cranium.

Other attempts had been made posteriorly by others groups as that of Van de Meulen in which correlation between the clefts and embryological events are the basis. However Tessier classification is easiest for its way of description and the simplicity of its nomenclature, and remains the more practical classification until now.

The clefts may involve soft tissues and bony structures in a different degree at different levels and can occur independently. Clefts could be presented unilaterally or bilaterally, though unilateral forms are the most common.

Clefts tend to follow the same axis, so is important to explore them very carefully in order to diagnose other features, though it does not mean that a cleft affecting soft tissues in the surface will affect in the same manner the underlying structures. Even more, defects on the bony structures are greater in clefts affecting the face than those in the cranium.

The extent and disfigurement caused by a cleft varies considerably, so a cleft could be as complete as to affect all the soft tissues with all the underlying bony structures or just be manifested as simple skin “notch” or small bridle in the oral mucosa.

3. Etiology
Formation of clefts takes place while the embryo is growing, and there is a pattern for the basic types of different clefts, so is important to know something about the embryogenesis of the face as a principle to understand these complex malformation.

**Fig. 2.-**

- Frontal process
- Lateral nasal part of frontal process
- Medial nasal part of frontal process
- Maxillary process
- Mandibular process

The different processes will joint together around the mouth, so anything that would interfere this “meeting place” or would cause the union to be ruptured will provoke a cleft. Correlation between the processes in the face of an embryo and the face of an adult will help to understand the morphology and distribution of clefts regarding the mechanics and chronology of cranio-facial morphogenesis.
Etiology of cranio-facial clefts is based on the same theories and principles as those described for cleft lip and palate. In fact, is the research on these more common clefts what has open the ways for the two main theories, that of “failure in fusion” or that of “lack of mesodermal migration”.

Latter Van der Meulen had proposed a more complex theory based on embryological events.

- **Theory of the Failure of fusion**

  Proposed by Dursy and His in the XIX century, is considered the Classic theory. It proposed that is a failure in fusion of the different processes what at ultimate instance causes the apparition of clefts. One or various of the different processes, or in some particular points of these processes, would be retarded or restrained in its growth, and so no contact and no fusion occurred with other processes and subsequently a cleft appears.

  ![Fig. 3](image)

**Fig. 3.** Correlation between the processes of the face of an embryo and an adult

  ![Fig. 4](image)

**Fig. 4.** Fusion between two processes fails because alterations at the ectodermic layer or a fail in that layer at the moment in which it must disappeared (to permit the mesoderm join the other part).

  Warbrick suggested that epithelial cells must disappear in the contact surface of the processes at the very moment of fusion between the processes. If these cellular layer persist, mesoderm could not join the opposite mesoderm, and fusion is not possible despite its union, then a cleft is produce at that point.

- **Theory of the mesodermic migration**

  This theory was presented by Pohlmann and Veau in the early years of the XX century, and they proposed that the lack of mesodermal migration and penetration resulted in a collapse of the ectoderm because a lack of support. This collapse is what finally ends in a cleft.
Fig. 5: Somewhere in one of the processes the mesoderm fails to follow under the ectoderm, and this, lacking support, collapses.

- **Van der Meulen theory**

By the latter part of XX century, Van der Meulen and its colleagues had suggested a more complex theory in which embryological concepts are better related with the concrete anomalies in a cleft. They proposed that clefting malformations are not really clefts but dysplasias. These dysplasias are the result of developmental arrests during the fusion of the facial processes. The differential defects are caused by absence or insufficient outgrowth of the ossification centres.

Finally it must be noticed that some clefs do not follow the path of a determined point or line of union between the facial processes and are perhaps due to a failure at the neural crest at the very moment of cellular migration or due to vascular failures that induced atrophy of the tissues as McKenzie and Craig postulated in 1955. In this direction, Poswillo, in 1975, demonstrated that the syndrome of the 1st. and 2nd. Brachial arches could be reproduced in laboratory, when a bleeding of the stapedic artery is produced.

4. **Cranio-facial clefts descriptions**

As has been said before, we will follow the description made by Tessier in its classification to expose and discussed each one of the current clefts, because is the generally accepted one.

- **Clefts nº 0**

Is a median facial dysraphia located in the central line of the nose, between its tip and the upper lip. Clinically it could be seen from a single width and broad nose with a central diastema to a complete defect of the lip and palate with absence, broad or duplicated nasal septum.

It is usually accompanied by nº 14 cleft, and result in a variety degree of hypertelorism.
• **Cleft n° 1**

Is a paramedian cleft that affects the soft tissues of the lip at the level of the cupid bow’s area to the dome of the alar cartilage, resulting in a notch in the dome of the nostril with an extension to the medial aspect of the eye brow. The cribiform plate and ipsilateral nasal bones could be widened and thus, certain degree of hypertelorism may be found.

• **Cleft n° 2**
This is a very infrequent cleft, though anatomically could correspond to a typical unilateral cleft lip. When the nose is involved, it is characterised by affecting the dome of the alar cartilage, the triangular cartilage of the nose and the nasal bone.

When affecting the maxilla, a cleft over the lateral incisive, hypoplasia of the piriform aperture, wide or absent frontal apophisis of the maxilla, and widened ethmoid cells are present. However, when the ethmoid is involved, certain degree of hypertelorism may be evident.

Cranial extension of this cleft constitutes cleft nº 12.

Fig. 8: a) Cleft nº 2 affecting the maxilla  

b) Cleft nº 2 affecting the lip

• **Cleft nº 3**

  This cleft is known as a naso-ocular cleft, or a medial orbitomaxillary cleft. It corresponds to the naso-lacrimal line in the embryo where the medial nasal, lateral nasal and maxillary processes join. When affecting the soft tissues, nº 3 cleft appears in the form of an unilateral cleft lip with a defect in the nostril. The lacrimal system is obliterated and a coloboma in the inferior eyelid is usually present, the medial canthus is in a lower position. Distortion of skeletal structures includes absence of the frontal process of the maxilla and the medial wall of the maxillary sinus. In this situation, the orbit, sinus, nasal cavity and oral cavity are interconnected.

  Usually is presented as unilateral form.

  The cranial extension is nº 11 cleft.

Fig. 9: a) Schema of facial cleft nº 3 affecting soft tissues and bony structures

b) Cleft nº 3 fully involving lips, maxilla, nose and orbits with anophthalmia.
• **Cleft nº 4**

Also known as median orbitomaxillary or oculo-facial cleft. When affecting soft tissues this is almost a straight cleft from the lip to the lower eyelid. At lip level, this cleft is situated just out of the cupid’s bow and represents also a unilateral cleft lip that uses to continue upwards through the cheek to involve the lower eyelid and, if prolonged superiorly, a coloboma of the medial eyelid and distortion of the eyebrow could be found. In severe cases, as the cleft passes through the orbit, an anophthalmia may exist. As this is a more lateral cleft that nº 3, the lacrimal system, though could be distort, is intact, as usually is the medial canthal ligament. On the skeletal structures, when complete, it gives continuity and communicate the mouth with the maxillary sinus and the orbit. When the bilateral form is present it distortion of the nose and premaxila is found in a similar way to the bilateral cleft lip and palate.

![Fig. 10: a) Schema of bone defect in Cleft nº 4 b) Left Cleft nº 4 combined with a right cleft nº 3](image)

This cleft when continues up to the cranium corresponds to nº 10 cleft of Tessier classification.

• **Cleft nº 5**

This is the rarest of all the oblique facial clefts. There is a cleft lip with the fissure just medial to the angle of the mouth but not in the commisure (would be another type of cleft). It courses upward across the lateral cheek to reach the lower eyelid between the medial and lateral third. Microphthalmia or anophthalmia are infrequent is this type of cleft. When involving the underlying structures, the defect starst at the alveolar premolar area to continue upwards through the maxilla lateral to the infraorbital foramen entering the orbit at the infero-lateral angle. Dystopia is most frequently seen. Upward projection to the cranium corresponds to facial cleft nº 9.
• **Cleft nº 6**

This cleft has been widely accepted as a minor form of the Treacher-Collins syndrome. Deformity in these cases includes coloboma at the medial third of the lower eyelids, antimongoloid eye slant. The ears may be prominent or normal at the most of the cases but some hearing deficit is present. Defects at the bony structures are characterised by a cleft or bone deficit at the zygomatico-maxillary suture with an intact zygomatic arch. Occasionally hair may be found at the malar area.

• **Cleft nº 7**

This is the most common and well known cleft. Is also known as hemifacial microsomnia, microtia, otomandibular dysostosis, Syndrome of the 1st and 2nd brachial arches, etc. (Goldenhard syndrome may be considered as a cleft nº 7 but it included epibulbar cysts and vertebral anomalies). As this cleft includes several malformations involving different facial areas the best name for it would be Lateral Facial dysplasia.
Clinically the cleft runs from mild forms with slight asymmetry between the hemifaces with small differences at the ears, to severe forms of hemifacial microsomia will full microtia with complete malformation of the middle ear and auditory canal, total absence of the mandibular ramus with severe hypoplasia of the maxilla and zygoma that could cause lateral canthal dystopia, accompanied by facial palsy and absence of the parotid gland an duct.

![Fig. 13.- a) Minimal form of cleft 7. Notice de right eye slant and shortness of the right mandibular ramus.](image1)

![b) Full form of cleft nº 7. Microtia, facial palsy, maxillary and mandibulary hypoplasia,hypoplasia of parotid gland.](image2)

- **Cleft nº 8**

  This rare cleft corresponds to a defect in continuity with the lateral canthus of the eye that extends to the temporal region. The lateral coloboma may be occupied by a dermatocele. When the underlying bones are affected it takes the form of a cleft in the frontozygomatic suture.

  When number 8th cleft is combined with the 6th and 7th clefts, the whole deformity is called Treacher-Collins Syndrome.

![Fig. 14.- a) Bone defect in cleft nº 8](image3)

![b) Cleft nº 8 at the lateral canthus](image4)

- **Cleft nº 6,7 and 8**

  When all this clefts appears together is called the Treachers-Collins or the Francescheti Syndrome. In those cases zygomatic bone is absence as could be the
zygomatic arch, there is a coloboma at the lower eyelid. Ears may be in a lower position or constricted.

Fig. 16.- a) Bone defects in Treacher Collins Syndrome.

b) Typical face of 6, 7 and 8th clefts

- **Cleft n° 9**
  
  Tessier cleft n° 9 is a superolateral orbital cleft traversing the lateral third of the upper eyelid and superolateral angle of the orbit and it could be considered that corresponds in the cranium to the facial cleft n° 5, but this is a rare combination.

Fig. 17.- Cleft n° 9 defect in the skull

- **Cleft n° 10**
  
  This cleft is located at the upper eyelid forming a coloboma in the medial third. In severe cases a complete lack of the eyelid may be seen, the cleft continues upward to the eyebrow dividing it in two portions. In the skull, it affects the central superior orbital rim, lateral to the supraorbital foramen, and continue up to the frontal bone. The defect at the skin may takes the form of a bridle or a scar tissues running upwards. At bone level, the orbital roof and supraorbital rim may be depressed or cleaved, and in complete forms of the cleft a frontal encephalocele may be found. When this is the case, the orbit appears to be rotated laterally and inferiorly.

  It seems to correspond to facial cleft n° 4.
**Cleft nº 11**

This cleft is located at the superomedial orbital rim. There is a coloboma in the upper eyelid in its medial third that sometimes continues to the eyebrow and from here to frontal hairline. When affecting the skull may be a cleft in the upper orbital medial rim, with involvement of the ethmoid and in severe cases a paramedian encephalocele. When is affected the ethmoid, certain degree of hypertelorism may be present.

It is considered to be the extension of facial cleft nº 3.

**Cleft nº 12**

The cleft extends from the medial canthus to the eyebrows and the frontal hairline. This is a more medial cleft passing through the frontal process of the maxilla and nasal bones medially to the canthus. In its way up to the cranium it involves the ethmoid and the cribiform plate. The resulting deformity is flatness of those bones giving the clinical aspect of hypertelorism. It is considered to be the extension of facial cleft nº 2.
• **Cleft nº 13**
  The Tessier nº 13 cleft displaced laterally the eyelids and the eyebrows and continues to the frontal hairline. At the skull, the cleft produces a widen ethmoid with widen olfactory grooves and cribiform plate resulting in hypertelorism. The defect at the ethmoid and frontal bones may produced a paramedian meningo-encephalocele which displaced downward the cribiform plate enlarging the intercantal distance and thus the hypertelorism.
  Corresponds to facial cleft nº 1.

![Fig. 20. Patient with a left cleft nº 12 and concomitant clefts nº 0, 1 and 13.](image)

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**Fig. 21.**

- **a)** Right Cleft nº 13 with a cleft type nº 2 already repaired.
- **b)** Severe case of combination of bilateral cleft nº 13, with bilateral 11, 2 and 0.

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• **Cleft nº 14**
  This median cleft corresponds to the extension of cleft nº 0. It could be presented in two forms, those with an excess of tissue in the midline or those with an important defect. The fail in the forward movement of the eyes gives place to a big defect in the ethmoid area that may result in a wide meningoencephalocele occupying the defect or just a cleft. Normally the ethmoid bone is prolapsed with a wide or duplicated crista.
galli and the distance between the olfactory grooves increased. All this alterations result in hypertelorism.

**Fig. 22.- a)** Cleft n° 14, Hypertelorism plus enlargement of the face because the meningo encephalocele

**b)** Cleft 0-14 showing hypertelorism with a wide and broad face due to arrest in the migration of the eyes.

**Fig. 23.- a)** Cleft n° 14 accompanied by clefts 2-12 in the right and 2-10 in the left. Encephalo locale pushing downward the ethmoid.

**b)** Cleft n° 0-14 with a Meningocele occupying the ethmoid area and widening the face

**Fig. 24.-** Meningo-encephalocele protruding between the palatal halves
5. **Surgical treatments of Cranio-facial clefts.**

Treatment of this complex variety of deformities is a very demanding surgery as it demands from the surgeon skill and experience in a great variety of procedures from different specialties as Neurosurgery, Plastic Surgery and Maxillo-facial surgery.

Some of the clefts, like cleft lip and palate, through complex and difficult both in management and surgery, are well known entities with well established techniques at certain ages.

Other clefts, needs complex craniofacial techniques to be treated, techniques that requires experience and skill in certain neurosurgery procedures with dominium of maxillofacial techniques for the skeletal facial alterations.

Most of the clefts are highly demanding in plastic surgery procedures as a great variety of flaps and/or tissue expansion techniques would be needed for reconstruction of eyelids, eyebrows, lips, a functional nose and an aesthetic ear.

Even more, some of these clefts may require sooner or later from orthognatic surgery, so the Cranio-facial surgeon must also be skilful in orthognatic surgery with experience in many of the maxillo-mandibular osteotomies.

Finally, no cleft is similar to another, and the great variety of clinical expression between clefts similar or equal in type and origin makes this surgery the most challenging and demanding of all the Craniofacial surgery, sometimes disappointing but the most completely rewarding.

Whole complete volumes would be necessary for discussing and presenting all the surgical techniques and procedures for treating these clefting anomalies, so we will review and discussed some of them. It does not mean that they are the only ones, but the more frequently used by the author.

- **Hypertelorism correction**

Hypertelorism is the result of various clefts and may be presented in two forms. One of them represents the true orbital hypertelorism, in which the orbits are just separated with no other implications, in the other one, an augmented interorbital distance is accompanied by a an antimongoloid slang of the orbits and upper external rotation of the whole hemifaces. May be corrected in two ways:

  o **Orbital medialization.** (Fig 25.)Through a coronal approach, hole dissection of the frontal bone, temporal fossa, infraorbital region and the four orbital walls is done. An “all around” orbital osteotomies with ostectomy of a central segment of nasal and ethmoid bone (according to the desired interorbital distance). This operation may be done intracranially (frontal bone craniotomy is done as access for full orbital mobilization leaving a frontal bar to give stability to the orbits) or subcranially (only the lateral walls and orbital floor are moved). Once the orbital osteotomies are done the orbits may be displaced to the midline leaving an intercanthal distance of 15 mms and fixed between them and to the frontal bar. Bilateral canthopexie and some nasal contouring may be necessary. (see below from a to b)
o Hemifacial rotation. (Fig. 26) This is done when the need is to mobilise to the midline the orbits as well as the whole hemifaces because the face is short and wide and the oclusal planes are slant. The procedure is similar to that described above but with some differences. The piece of nasal and ethmoid bone to be resected will have a trapezoidal shape (rather than rectangular) and an additional triangular osteotomy is done at the junction of the two palatal halves until reaching the interincisival diastema. The osteotomy is performed in a Lefort III fashion and both hemifaces must be liberated and loose to be brought to midline and fixed superiorly with wire or miniplates, and inferiorly with wire and with a previously done dental splint. Bone grafts are used to fulfil the area of the lower forehead. When this medial rotation is accomplished, simultaneously, the hypertelorism is corrected, the orbital and oclusal slant are in a more horizontal plane, the wideness of the face is decreased, the same face lengthened and the nose narrowed. This technique is very predictable by previous planning on cephalometrics. Technical details as canthopexie, etc. are done in a similar way that to medial orbital mobilization.


**Canthopexie**

Canthopexie is one of the most important steps in cranio-facial surgery. Everything could be properly done, but if the medial canthopexie fails, a telecanthus appear and the final look would be disappointing. The keypoint for a successful medialization of a relapsed tendon is its adequate repositioning at its anatomical position. The technique use is that of transnasal canthopexie.

Unilateral or bilateral, the successful canthopexie would follow some basic principles:

- Precise localization of the medial canthal tendon.
- Solid suture of the tendon with wire or any non absorbable suture.
- Accurate trephine at the posterio-superior aspect of the lacrimal groove on each side to permit the suture pass through.
- Helped by a Reverdin needle, the suture is passed from the initial sutured tendon through the nose cavity to the contralateral orbit, sutured to that tendon and passed again to its original position. Then the suture is tightened until the desired intercanthal distance has been obtained. Wire suture allows more precision while tightening. Careful overcorrection is desired.
Fig. 27. From a to d, procedure of transnasal canthopexy with wire using small trephines on the medial orbital walls by small medial canthal incision.

Telecanthus, sagging and poor results are observed if the canthopexie has not been properly done. The most common technical errors are malposition of the trephine were the suture must past through, use of absorbable sutures and sutures done over soft tissues, not over the medial canthal tendon.

• Bone grafts and bone healing

Treatment of clefts at the craniofacial skeleton means osteotomies but also the employment of bone grafts to stabilise osteotomies and defects, bone segments or just performing reconstructions as those used for the nose.

The most important factor for a bone to heal is stability, of course bone to bone contact and good local conditions are important, but without stability, the bone undergo a process of resorption, that finish with loss of mass, strength and volume.

The skull is composed by membranous bone and the process of healing occurred by fibrous union alone. The osteotomy line of contact, form a callus consisting partly of cartilage, and four weeks later some stability appeared. Despite there is evidence of bony union, the line of contact remains radiographically translucent. It does not mean that consolidation has not been obtained, it only means that deposits of minerals in the fractured site of a membranous bone are such that radiolucency persists.

However, if there is not good contact between the bones segments the healing would fail. For that reason bone grafts must be employed, when necessary to lead to a better bone healing. Loose segments would tend to resorption altering the final result, both functional and aesthetically.
The calvaria is the best donor site area for grafts because its same membranous origin and also, there is a considerable amount of bone at disposal, and no less important because is in a neighbour area. Vascularized bone grafts as those with its pedicle in the galea (partial or full thickness) are better than non vascularized, but the later if properly fixed under good soft tissue conditions, could maintain their volume and strength quite well.

In summary, good bone healing requires:
- Perfect reduction of bone segments to stable position.
- Rigid fixation of the fragments
- Get good soft tissue cover
- Allow enough time for the bone to heal.

Fig. 28. - Harvesting a) monocortical grafts and b) full thickness calvarian bone grafts

Fig. 29. - a) Schema of monocortical vasculariced bone graft and B) Design for a full thickness bone graft
**Local flaps in cleft surgery**

Cleft surgery has always been demanding for soft tissue coverage. From the beginning local flaps has been the only choice for surgeons. After, the advent of myocutaneous, osteomyocutaneous and free flaps were a good option when anything was at hand, but if they provide full coverage the aesthetic result was far from be acceptable.

As we have told before tissue expansion has represent a great advance in great clefts surgery, but we must consider that in undeveloped areas (and this manual is addressed to them), some times access to expanders is just impossible or, simply, it would be impossible for a child to spend one or two months visiting the specialist for the weekly instillation. So local flaps, specially in undeveloped areas, are gaining acceptance again as the procedure of choice for repair facial clefts.

With the exception of unilateral or bilateral cleft lip, where there are specific technical designs for each of them, for the rest of the clefts there are no special design of flaps, and this is the main challenge when operating clefts. It is all up to the surgeon experience and skill for designing and performing the flaps. These flaps usually takes the form of multiple, symmetrical or asymmetrical Z-plasties.

Fig. 31.- Single Z-Plasty and canthopexie for clefts affecting the medial part of the orbit

The small z-plasty flaps could vary from simple skin flaps to the more complex ones involving oral mucosa and muscles. Pre-operative planning must be excellent, if not the flaps will not correspond each other and distortion will appear invariably. Canthopexie must be done when the medial canthus is affected. When needed bone grafts to restore the orbital rim and the maxilla must be done simultaneously.
Fig. 32.- Various asymmetrical Z-Plasties for correction of oro-naso-ocular clefts plus a medial canthopexie.

- **Tissue expansion in craniofacial surgery**

  Cleft defects are characterised by the loss of shape and mass, affecting soft tissues as well as bony structures. Despite excellent management of bony defects, if soft tissues are not properly reconstructed the final result will be, aesthetically and functionally, less than acceptable.

  This lack of tissue has been a great problem since the initial times of surgery. Local and distant flaps as well as full thickness grafts had been used, but one of the sacred rules in reconstructive surgery is to use the more similar skin for that of the defect we want to repair, and this rule was rarely followed because the lack of appropriate skin and subdermal tissue. All this produced very limited aesthetic results.

  Tissue expansion has come to solve many of these problems, because it permits the surgeon to increase the amount of skin for reconstruction. It is based on the skin property of elasticity, that permits elongation under internal or external stretching forces (as happens in pregnancy).

  Tissue expansion permits controlled increase in the amount of skin, by the election of the appropriate expander for the shape and size of the defect, and the volume and frequency of instillation.

  The procedure consists in the subcutaneous implant of a silicone balloon with a valve attached to it by a thin tube. Some principles must be followed to ensure viability and success:

  - Careful planning is the key for success. The skin that would be transposed must include whenever is possible, an axial pattern of vascularization.
  - Incisions for implanting the expander must be as much parallel to the smallest axis of the expansor as possible, if not your incision will be expanded at the same time with a high risk of dehiscence.
  - Expanders must be placed in a neighbour area to the defect, thus permitting reconstruction by rotational or advancement flaps.
  - The expander must be under skin in good conditions no full thickness scars, sores or radiated skin.
  - The valve must be positioned far from the tissue expander (internal or external) in order to avoid the risk of punction over the expander.
  - The face has its own aesthetic units (Fig. 29), units that when fully repaired do not give the aspect of “patches”. If flaps crosses the lines between this aesthetics areas, the aesthetic result will be seriously compromised.

  Immediately after its placement, the expander may be inflate, wait for 2 weeks, and then start weekly instillations with sterile saline (twice a week p.e.). As a general rule, the amount of tissue gain is roughly 1.5 times the width of the implant.
Fig. 33.- a) Multiple shapes and volumes may be found for specific donor areas

b) Improvised tissue expander with a foley catheter

Tissue expanders are expensive devices rarely found in undeveloped areas, so oneself must make his way, and foley catheters may be a limited alternative, but an alternative at least.

When planning soft tissues reconstruction at the face we must think about the only single and first opportunity, it means that if we need skin for prolabium, nose, eyelids, cheek, etc. we must have in mind from the beginning how we will use the available skin for each of these defects. In that way, only one tissue expander could provide you with enough skin for reconstructing several aesthetic areas of the face in more or less stages. If the Tissue expansion is poorly planned or the flaps badly executed the result will be poor and the best skin and opportunity to the child gone.

Fig. 34.- Aesthetic units of the face

- Forehead
- Superior eyelids
- Inferior eyelid
- Nose
- Cheeks
- Upper skin lip
- Chin area
Fig. 35.- Two examples of “what not to do”

**a)** Poor planning with no consideration about the length of flaps and the necessary frame reconstruction for the nose resulting in a complete collapse and the worst of all, waste of first and better choice options (forehead and nasolabial flaps)

**b)** Failure in considering the aesthetic facial units and a myocutaneous flap (far from the cheek) instead of using a cheek flap, makes this cheek “heavy”, pulling downward the lower eyelid and giving the aspect of a patch in the face.

Fig. 36.- Perfect planning and execution: 1 expander gives tissue enough for reconstruct in 3 stages the nose, the lower and upper eyelid, and the left forehead.

Fig. 37.- An extremely challenge and difficult case: 1 tissue expander allows reconstruction of the bilateral cleft lip, lowering the prolabium, tissue for closing the clefts at cheeks and tissue for a nose reconstruction. Of course complete treatment will go on in further stages.

However matters could go wrong and the best way to avoid complications is a close control of the expander. In the evidence of an excessive thinness of the skin, signs of tissue suffering or inflammatory changes, the first that we must do is retire some saline from the expander and avoid any tension on the skin, wait to see the evolution and then in a week or two restart with fluid instillation or just admit failure an retire the expander waiting for the skin to heal.
6. Summary

Facial clefts are among the most difficult deformities between Cranio-facial anomalies. They involve a great variety of organs and structures in many different ways and despite there is a pattern for all the clefts, its clinical presentation is never the same between two patients.

The surgeon involved in the treatment of Cranio-facial alterations must show great dominium of surgical procedures from different specialties, if not incomplete treatment will be done and disappointing results will be obtained.

Many procedures are needed for an integral treatment of a severe clefted patient, some has been discussed here, because its prominent role as part for the most of the rare cranio-facial clefts.

Other techniques for the most common clefts, those of cleft lip and palate, has been omitted here, because they need by themselves a whole book or chapters.

Procedures for microtia, facial palsy and orthognatic surgery has been also excluded because lack of space in that chapter and they will be discussed forward.

Dr. Fernando Ortiz-Monasterio, one of the leading Plastic Surgeons of these times, teaching young surgeons in a workshop, 1988.