Cleft lip and palate

Cleft lip and Palate

In memory of Tommaso Franchella (1980-2015)

Cleft lip and Palate

Section 1
Team approach
and surgical management

Section 2
Surgical partnership.
Cleft care in Low and Middle Income Countries

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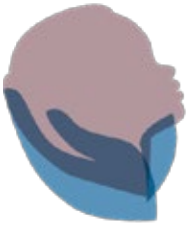

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Cleft lip and Palate

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Knowledge and best pratice in this field are constantly changing.
Pratictioners must always rely on their own experience and knoledge in evaluating and using any information described herein.

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Preface

Every year around five million babies are born with birth defects (also known as congenital disorders), most of them in developing countries. Birth defects are rapidly becoming the leading cause of childhood mortality and morbidity, and survivors face a lifetime of ill-health and the attendant problems of poverty, stigma and discrimination.

This book represents a huge effort to condense the experience of the Pediatric Surgery Department of the University Hospital of Ferrara, both in Italy and in humanitarian missions carried out in Low and Middle Income Countries. The content is based on material regarding pediatric plastic surgery volume, edited by me and my staff in 2007 and two doctoral thesis one on primary surgery and one on secondary surgery.

The first section intends to provide a selection of the procedures and resources that have shown the best results over the years. Milestones and difficulties are analyzed. Meticulous documentation and follow-up are actions that have allowed us to choose and recommend some techniques over other less-effective procedures. The complexity of the problems to be treated in an oral cleft patient require a team approach with the organization of a group of specialists dedicated to facilitate the interdisciplinary treatment of cleft patients, improve results, and prevent sequelae.

Today, the accepted goal is to obtain excellent aesthetic and functional results during primary reconstruction of the lip, nose and palate, in 85-90% of cases. Using less aggressive techniques, conserving and respecting growth, innervations and blood supply, will allow us to prevent the growth and development of facial alterations.

The second section examines the problems of the management of cleft lip and palate in Low and Middle Income Countries, and aims to raise the profile of a

condition commonly considered to be of only cosmetic importance in countries where infant and child mortality rates are high. Management of Cleft Lip and Palate in the Developing World is an important resource for anyone having - or planning to have - a commitment to develop services in that environment and to clinicians in the Developing World faced with large numbers of cleft lip and palate patients and limited resources of materials and manpower.

Thus, the book provides a contemporary overview of the epidemiology, aetiology and treatment of cleft lip and palate, and will be of use to a wide range of individuals, including students, biologists and clinicians, who have an interest in this subject.

The Editor in Chief

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Acknowledgements

We would like to thank all the parents who trusted us with the treatment of their children. An oral cleft delivery creates an emotional burden on the family, compromises the physical and psychological development of the patient as well as a disadvantage in being socially accepted.
This book represents a huge effort to condense experiences and suggestions. Our intention is to facilitate the multidisciplinary treatment of cleft patients, improve results, and prevent sequelae.
Many thanks to our cleft lip and palate team for their encouragement and friendship, especially to those who collaborated in producing this book and to the humanitarian organizations with whom we have worked with.
We would like to thank Paola Gonzalez Ruiz and Sara Marchetto for their skills and contribution to illustrating the book, Daniele Borsetto for the competent translation and Sara Storari, head of *Studio Fuoriregistro,* for helping us in the process of selection of the chapters and editing.
Thanks to *Chirurgo e Bambino Onlus* - through which this book will find its way to the Web and to so many people who can not read the printed book.
Finally last and not least we would like to express our gratitude to our families who have supported our efforts while writing the book.

The Editors

Contents

Section 1

Team approach and surgical management

1.
The basics of cleft lip and palate

1.1 Definition

Cleft lip and palate, also known as **orofacial cleft** and **cheiloschisis and palatoschisis**, is a group of congenital malformations that includes cleft lip (CL), cleft palate (CP), and both together (CLP) *(Stanier P, Moore GE, 2004)*. A cleft lip contains a defect in the upper lip that may extend into the nose. The defect may be on one side (unilateral), both sides (bilateral), or in the middle. A cleft palate is when the roof of the mouth contains an opening into the nose. It can affect both the hard and soft palate.

A wide variety of cleft forms must be considered: the most simple form is commonly known as "hare lip".

1.2 Epidemiology

Cleft of the lip and palate are the most common of the craniofacial malformation.

The true prevalence rate of cleft lip and palate worldwide is a question for debate and the prevalence of syndromic and non-syndromic cases should be treated separately.

The birth frequency of cleft lip, cleft lip and palate and cleft palate alone is not known in many regions of the world *(Mossey PA, Little J, 2009)*. General incidence is around 1: 700 live births. The prevalence varies widely depending on the records referred to whether the collection of data was hospital based or population based *(Watkins SE; Meyer RE)*. Figures range from 3.4 to 22.9 per 10,000 live births for cleft lip with or without palate (CL/P) and from 1.3 to 25.3 per 10,000 live births for isolated cleft palate (CP) *(Global Health Estimates 2014)*. CL/P appears more frequent in some areas of Latin America, China and Japan and less

frequent in Israel. Isolated CP is more common in Canada and Northern Europe and infrequent in Latin America and South Africa.

Published data shows that about 50% of orofacial clefts are cleft lip and palate, while isolated cleft palate or cleft lip evenly split the remaining 50%.

Isolated cleft lip and cleft lip and palate are more frequent in males with a sex ration of 2:1 whilst this ratio is reversed for cleft palate.

Two-third of the unilateral cleft lip, with or without cleft palate, are observed on the left side. Cleft palate is associated with bilateral cleft lip in 86% of cases and unilateral cleft lip in 68%.

The frequency at birth varies between different geographic regions and different racial groups.

Twice as many males as females are affected by this condition. In contrast, nonsyndromic cleft palate occurs in an equal male-to-female distribution.

The possible association with other congenital defects has been shown to be significant (13-50% in cleft palate, 7-15% in cleft lip, 2-11% in cleft lip and palate *(Calzolari E, Pierini A, et al 2007).*

New clinical syndromes in which a variant of this malformation are present continue to be reported and this aspect should be taken into account before the diagnosis of isolated anomaly and counselling of parents regarding future pregnancies. A lack of awareness of the categories and prevalence rates can lead to poor genetic counselling which leads to stronger negative effects on families. Population-based studies will be more appropriate in investigating a worldwide epidemiology. Spontaneous abortions, elective terminations, stillborn foetuses, and babies that died shortly after birth should be considered to get the true numbers of associated anomalies. It is also essential that every child should be thoroughly examined immediately after birth for the associated anomalies, because children with severe malformation may not survive long.

Research has identified that the distribution of clefting is influenced by the sex of the individual. Nonsyndromic cleft lip with or without cleft palate has higher prevalence among males while cleft palate is more prevalent among females. Interestingly the rate of occurrence in one of two monozygotic twins is 50%, highest rate of occurrence, African Americans show the lowest.

1.3 The embryological development of the face

Facial development

The development of the face occurs through a series of complex morphogenetic

events and rapid proliferative expansion. The embryological process is highly sensitive to environmental and genetic factors which explains the high prevalence of oro-facial malformations at birth. Very early in pregnancy (6-8 weeks) the shape of the embryo's head is formed. Five primitive tissue prominences grow:

- One from the top of the head down towards the future upper lip (Frontonasal Prominence);
- Two from the cheeks, which meet the first lobe to form the upper lip (Maxillary Prominence);
- And just below, two additional lobes grow from each side, which form the chin and lower lip (Mandibular Prominence).

If these tissues fail to meet, a gap appears where the tissues should have joined (fused). This may happen in any single joining site, or simultaneously in several or all of them. The resulting birth defect reflects the locations and severity of individual fusion failures (e.g. from a small lip or palate fissure up to a completely malformed face).

The upper lip is formed earlier than the palate from Frontonasal Prominence and Maxillary Prominences. Formation of the palate is the last step in joining the five embryonic facial lobes, and involves the back portions of the primitive prominences, called palatal shelves, which grow towards each other until they fuse in the middle.

The biologic mechanisms of mutual recognition of the two cabinets, and the way they are fixed together, are quite complex and obscure despite intensive scientific research.

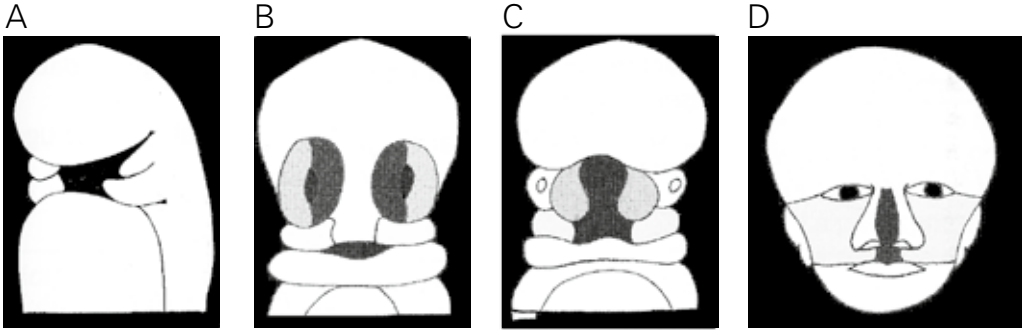


Figure 1.1 Development of the face from the mesoderm of the embryo and the progressive fusion of facial lobes.

The five most important fusion steps are:

1. At the end of the 4th week the mandibular prominences meet on the middleline and form the primitive lower lip and chin.

2. At the end of the 6th week medial nasal processes migrate towards the centre giving rise to the medial part of the maxilla. The upper portion forms the base from which the nasal septum develops, while in the lower part, the intermaxillary process grows between the two maxillary processes. The intermaxillary process is made from an external region (from which the philtrum of the upper lip is formed), an intermediate one (from which the region comprising the four maxillary incisors will develop) and one internal (protruding in the buccal cavity, which will form the primary palate).

3. The two maxillary processes are combined with the Intermaxillary process forming the arch of the jaw and upper lip.

4. The lower edges of the maxillary processes, merging with the underlying mandibular processes reduce the primitive mouth opening.

5. The top edges of the maxillary processes merge with the nasal processes forming the lateral portion of the maxilla; it is an incomplete process as it remains the continuity represented by nasolacrimal canal.

During the confluence and fusion of the facial lobes between the 4th and the 7th week, the embryonic eyes move from the side of the head towards the face and there is an upward migration of the primitive ear.

The formation of the palate

The palate originates from two structures: the primary palate and the secondary palate.

The primary palate, which separates the nasal cavity from the stomodeum (the primitive mouth), consists of the lower portion of the medial frontonasal prominence with contribution from the maxillary processes. These structures form together a horizontal triangular lamina with posterior vertex, called anterior palate. This part of the maxilla is fully formed by the 45th day. At this stage, in the stomodeal cavity three structures are present: the nasal septum (medial, vertical, resulting from the frontal process) and the palatine processes (two, lateral, horizontal, coming from the maxillary processes). These three plates come together in the midline, separating the definitive nasal cavity from the oral cavity.

The fusion of the palatine processes and the nasal septum begins around the 9th-10th week and will give rise to the secondary palate. A critical moment still under study in the development of the palate is the raising of the palatal shelves that from the sixth week of embryonic life grow on the vertical plane laterally to the tongue and then, following a proliferation of mesenchymal cells, on the horizontal plane above the tongue until meet and fuse on the midline. The palate can be considered formed by the tenth week. In murine models, an anomaly of this

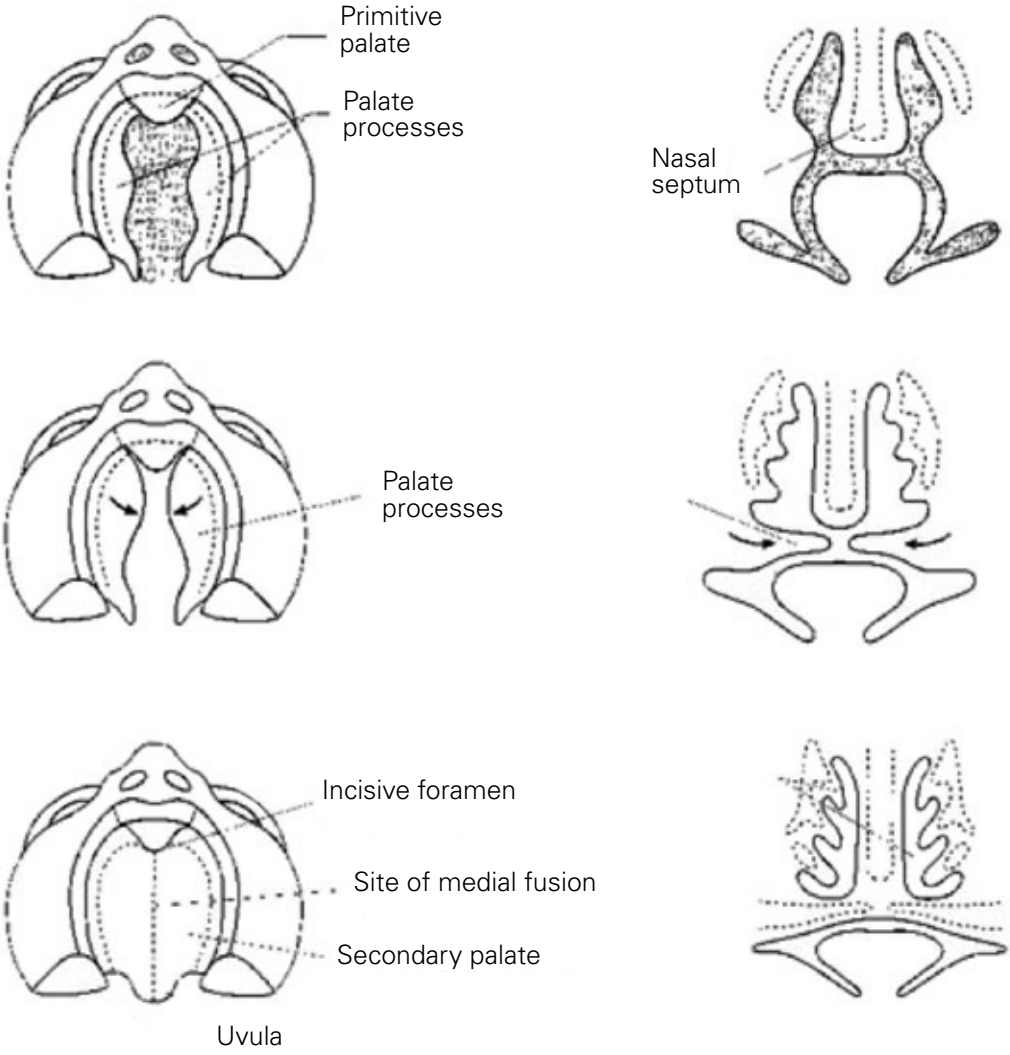


Figure 1.2 Development of the palate.

process is the most common cause of the development of a cleft palate *(Stanier P, Moore GE, 2004)*.

The tongue, which initially filled all of the stomodeum, is thus confined to the oral cavity.

The primitive palate and the anterior portion of the secondary palate undergo membranous ossification processes giving rise to the bony or hard palate. The remaining portion to the rear does not ossify and becomes the soft palate and uvula.

The fusion of the palate proceeds in the anterior to posterior direction, from the hard palate to the uvula and this explains why a soft palate cleft can occur not associated with cleft of the hard palate, but not vice versa.

The facial fusion process occurs between the 5th and the 8th week and a defect of this process determines an abnormality which appears as the persistence of a gap or cleft.

Cleft lip results from a defect in the confluence of the lateral and medial parts of the frontonasal prominence. Depending on the extent of the defect they take different forms, on one or both sides, simple or complete according to the degree of involvement of alveolus and nose.

Pathogenesis is due to an insufficient migration of neural crest cells and / or the lack of proliferation of these cells.

Cleft palate originates from a non-fusion of the palatine processes, which can be caused by:

- Non-proliferation;
- Excessive enlargement of the head;
- Failure to change the direction of development of the palatine processes in the formation of the palate;
- Inability of the processes to merge after coming into contact.

Genetic studies have shed light on the molecular mechanisms that determine the majority of these embryological steps. Epidemiology indicates that the cleft palate associated with cleft lip is secondary to a defect of fusion of the mid-face processes preceding the formation of the palate and then the isolated cleft palate has a distinct aetiological origin.

1.4 Classification of orofacial clefts

Cleft lip is classified according to its location and severity:

- **Unilateral** (on one side of the lip). Cleft lip, even in its mild expressions is associated with nasal deformity so that the nostril on the affected side is tilded and lower than the other nostril.
- **Bilateral** (on two sides of the lip). The bilateral cleft may cause the nose to be broader and shorter than normal.
- **Complete** (extends to the nose).
- **Incomplete** (does not extend to the nose).

If cleft lip occurs with cleft palate, the upper alveolar ridges, are also involved.

The **hard palate** is the anterior part of the roof of the mouth.

The **soft palate** is the posterior part of the roof of the mouth. A cleft palate may involve the soft palate with part of the hard palate or only the soft palate.

The large number of various anomalies found in the field of facial clefts has led to difficulty in creating a comprehensive system of classification.

An initial classification system was proposed by Davis, Ritchie and co-workers, who created a system of purely anatomical cataloguing. This system remained in use until the end of the 1950's and brings together the various deformities in cleft lip and palate, unilateral or bilateral, complete or incomplete and isolated cleft palate but little consideration is given to the alveolar process.

In 1958 Kernahan and Stark proposed a new classification based on the need to distinguish between primary palate and secondary palate. In this method a variant for which the incisive foramen is considered the dividing point between the two structures is included. This allows the division of the facial deformities into three large groups: cleft of primary palate, cleft of the secondary palate and cleft of primary and secondary palate.

Diagram of classification

In 1971 was introduced the striped "Y" logo to provide simple and easily recognizable visual classification suitable for the front of a clinical chart and for computerization.

Each of the arms of the "Y", starting from the outside, are the lip, the alveolus, and the hard palate as far back as the incisive foramen.

The remaining portion, the stem of the logo expresses the division into thirds of the hard and soft palate.

The important point of division between the two groups is the incisive foramen.

Coding of orofacial clefts

Numeric codes have been developed to enable international comparable studies on orofacial clefts together with other malformations, syndromes, associations and sequences.

These codes include the International Classification of Disease (ICD) codes. ICD 9 with the British Pediatric Association extension - ICD9 BPA – and the newer ICD 10 are widely used for coding and classifying oro-facial clefts and their related sequences, syndromes and associations. The EUROCAT Coding guidelines for registration of congenital anomalies Guide 1.4 (http://www.eurocat-network.eu/content/EUROCAT-Guide-1.4-Section-3) are freely available to all who wish to classify their cases and compare their data to the literature. This classification is essential in order to collect comparable epidemiological data.

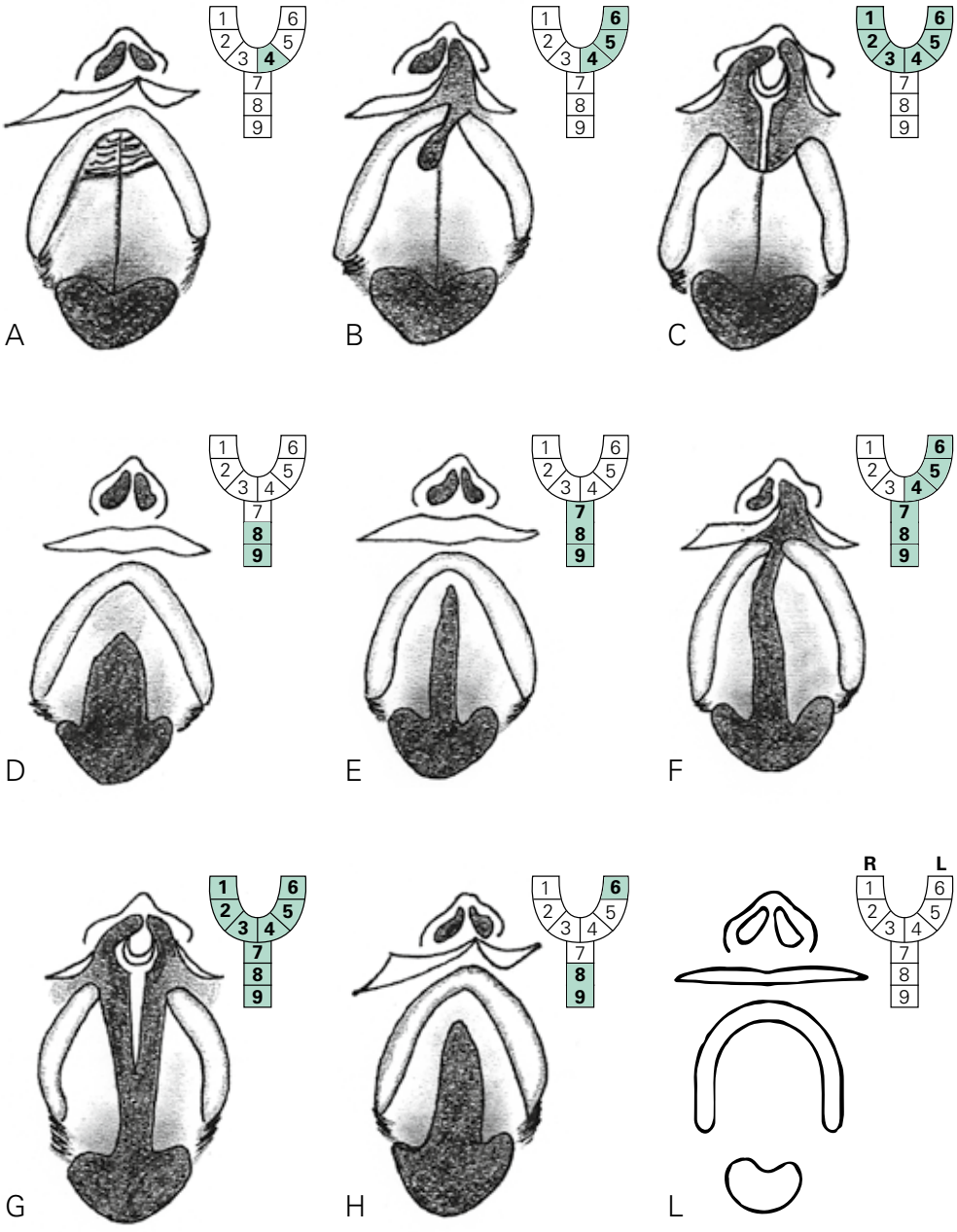


Figure 1.3 Normal conditions where 1 and 6 represent the lip, 2 and 5 the alveolus, 3 and 4 the primary palate separated from the secondary palate (7, 8 and 9) from the incisive foramen. (L) **+** Complete cleft of secondary palate. (E)
+ Complete unilateral cleft lip and palate (left). (F)

Morphological characteristics of cleft lip

Cleft lip can be complete or incomplete. In complete cleft lip there is involvement of the lip, the nasal floor, the alveolus and the primary palate. In 30% of complete cleft lip cases the so-called Simonart's band is present, a bridge that crosses the skin and ipsilateral nostril below the cleft.

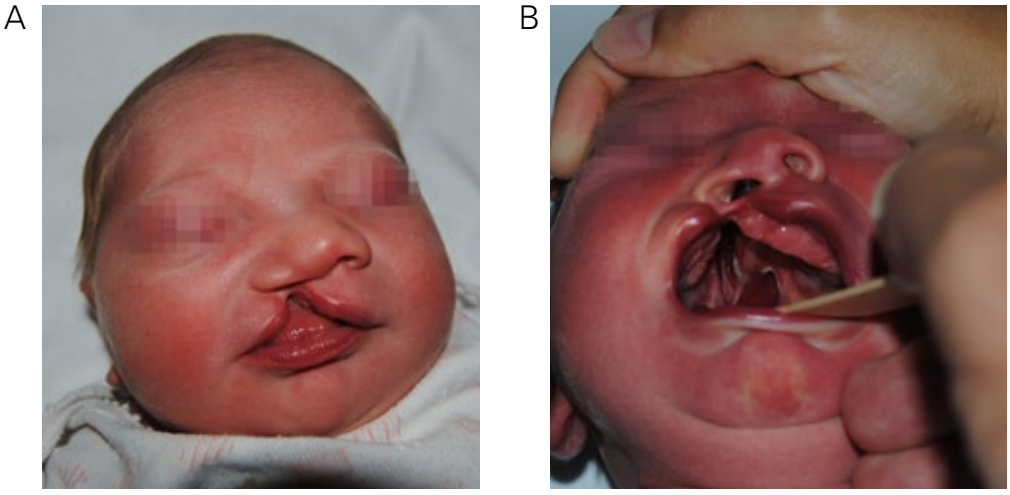


Figure 1.4 Complete right cleft lip and palate with Simonart skin band below the right nostril. (A) Frontal view. (B) Comprehensive view of cleft lip and palate.

There is a spectrum of incomplete forms of cleft lip. Yuzuriha Mulliken attempted to outline and simplify these by defining minor forms characterized by a segment depression of 3 mm or more at the mucocutaneous junction above the Cupid's bow. Those with a segment elevation of less than 3 mm are classed as microforms and mini-microforms those with disruption of the mucocutaneous junction without distorting Cupid's bow *(Sitzman TJ, Marcus JR, 2014)*.

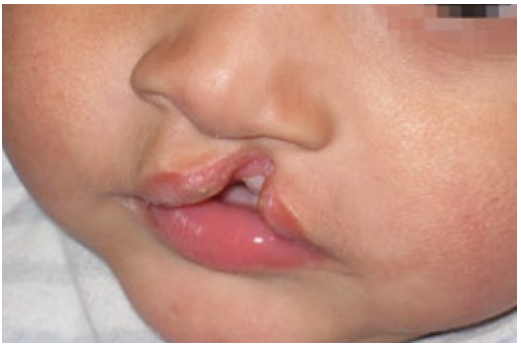


Figure 1.5 Incomplete left cleft lip.

In unilateral forms the nasal ala cartilage on the side of the cleft is displaced and flared (the degree depends on the severity of the cleft) and the tip of the nose is diverted to the healthy side. The middle portion of the lip is shortened with the vermilion thinned. The lining of the lateral portion is also lacking. The orbicularis oris muscle is deficient and has listings aberrant underlying nasal ala, the columella and the dermis level of the defect.

In the bilateral forms the middle portion of the lip is isolated and attached to the pre-maxilla that protrudes considerably with respect to the profile of the face and is attached to the vomer and the nasal septum. The nasal cartilages are flared and flattened on both sides. As with the unilateral deformity, the bilateral cleft can vary in degree of completness on one or both sides.

There is a direct relationship between the severity of the cleft and size of the alveolar defect and of the teething defect: the teeth can be altered both in number, shape and position *(Berkowitz S, 2013)*.

The incisive foramen is the point of reference of separation between the primary and secondary palate and then between cleft lip and complete cleft palate.

Morphological characteristics of cleft lip and palate

In complete unilateral cleft lip and palate there is communication between the nasal cavity and oral side of the cleft; on the healthy side the nasal septum is instead attached to the palatine process. This category actually includes a broad spectrum of anomalies: the palatine segments may be more or less separated, the affected side is usually mediated and lifted up, the nasal septum is deflected toward the side not affected, so much so that sometimes it can assume a completely horizontal inclination.

In bilateral forms, the oral and nasal cavities communicate from both sides, the nasal septum forms a midline structure firmly attached to the base of the skull, but relatively movable inferolateral which acts as a support to the pre-maxilla and columella. The number of teeth that can be contained in this central segment is related to its amplitude and its form. When the defect is complete in each of the two sides, it can not contain all of the permanent teeth.

Morphological characteristics of the isolated cleft palate

The defect may affect only the soft palate or both the soft and hard palate. It may extend from the uvula backwards for varying widths. In some cases it may be limited simply to the uvula, known as uvula bifida but does not affect the lip or the alveolus. Precisely because of this phenotypic variety it is important to manually review the posterior margin of the cleft seeking an incision that can reveal the presence of a submucosal cleft otherwise difficult to highlight.

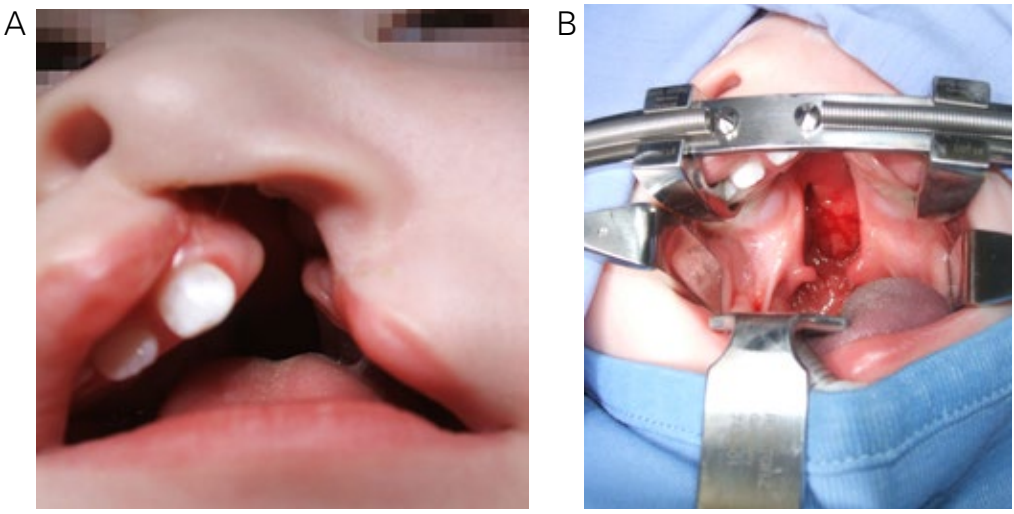


Figure 1.6 Left complete cleft lip and palate. (A) Front view. (B) Detail of cleft palate.

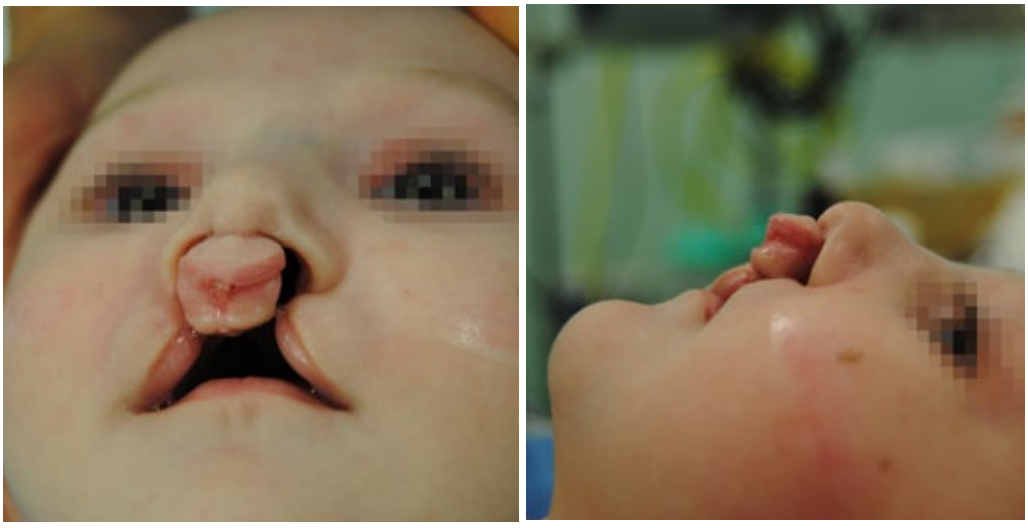


Figure 1.7 Infant suffering from bilateral cleft lip and palate with important protrusion of the pre-maxilla.

The palatal defect can be wide or narrow, U-shaped or V-shaped, the vomer is usually thin and sharp and frequent association with micrognathia gives rise to the hypothesis that one of the causative factors is the position of the tongue which occupies the space between the two palatine processes, hindering the merger due to micrognathia.

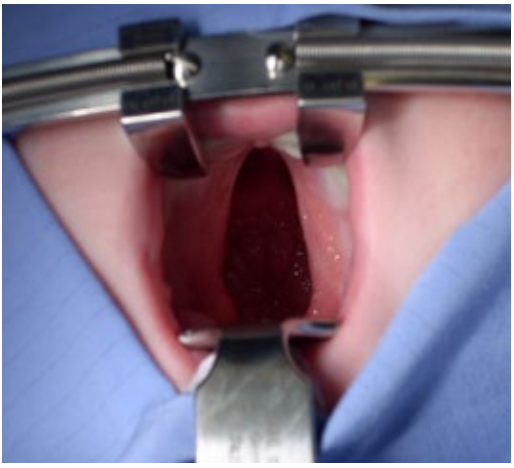


Figure 1.8 Complete Cleft Palate.

Submucous cleft palate

The submucosal cleft is diagnosed in the presence of the classic triad: bifid uvula, separation of the muscles of the palate in the midline in front of a continuity of the mucosal surface and palpable midline incision on the posterior margin of the hard palate.

From the functional point of view there is generally a deficit of thickness and length in the soft palate which makes difficult to produce a functional velopharyngeal valve, so that also for this defect a surgical correction is necessary, as well as a speech therapy.

Velo-pharyngeal insufficiency

Even the failure of the velum of the palate must be considered a congenital defect. Although less easily determined compared to cleft, it usually becomes evident when the patient acquires speech and with it the hypernasality typical of a cleft.

From the point of view of the anatomic cause, this can be a palate that is too short or a deficit in the hard palate.

1.5 Aetiopathogenesis

In the analysis of the factors involved in the aetiology of the cleft lip it is important to consider the concept of aetiological heterogeneity, or the existence of multiple causes for a single phenomenon.

The craniofacial formation is an extremely complex process, in which different factors both environmental and genetic are involved and interact. These factors can therefore interfere with the normal development of this area, resulting in the wide variety of defects described above.

This complexity explains the still incomplete knowledge, especially as regards the etiopathogenesis of forms not associated with syndromes (about 70%). The concordance of these defects is about 25-45% in identical twins and this proves the existence of an important genetic background, at the same time the fact that this correlation is not complete highlights the influence of certain environmental factors in the development of a defect in the embryo.

At least 600 syndromes include cleft lip and / or palate, of the currently known 6,000-7,000 syndromes. They fall into forms characterized by Mendelian disorders (defect in a single gene), forms resulting from chromosomal defects, forms associated with known teratogens and forms whose origin is still obscure. Despite this schematization, the advancement of molecular techniques has now enabled us to identify abnormalities involving small chromosomal regions, sometimes even specific genes, for which the distinction between the first 2 categories are now useless. The risk that there are associated malformations is higher in the forms of bilateral cleft lip and palate, the lowest risk is, instead, in isolated unilateral cleft lip.

Syndromes associated with cleft lip and palate

The syndromes associated with cleft lip and palate already characterized by a genetic perspective include:

- Van der Woude syndrome;
- The CLP Ectodermal dysplasia syndrome;
- The ectrodactyly, ectodermal dysplasia;
- The Ankyloblepharon, ectodermal dysplasia, clefting syndrome or Hay-Wells syndrome;
- The X-linked Opitz syndrome.

Among the syndromes associated with isolated cleft palate:

- The X-linked cleft palate syndrome that associates cleft palate and ankyloglossia;
- Treacher Collins syndrome: malformations of the middle and outer fold and eyelids that slope downwards, coloboma of the lower lid, and zygomatic mandibular hypoplasia and cleft palate;
- The Holoprosencephaly syndrome: spectrum ranging from abnormal midli-

ne to a complete failure of the forebrain division with cyclopia and often cleft palate;
- Stickler syndrome (hereditary arthro-ophthalmopathy);
- DiGeorge Syndrome or Velocardiofacial or Catch 22, characterized by heart defects, facial abnormalities, thymic hypoplasia, cleft palate, hypocalcemia.

There are then cleft associated with syndromes of unknown aetiology / not defined, such as:
- Amniotic band (can give typical forms or details of cleft lip);
- Pierre-Robin sequence: a condition characterized by micrognathia, glossoptosis and cleft of the soft palate.

Non-syndromic orofacial cleft

Given the multifactorial nature that characterizes the isolated malformations, the identification of genes responsible is still an obstacle. There have already been many association studies in the last decade to test the involvement of different chromosomal loci, but all have yielded contradictory results.

Among others, the genes encoding endothelin-1 (ET1), were analyzed whose mutation in the mouse produces phenotypic manifestations similar to those of the condition CATCH-22 in humans; the gene of the retinoic acid receptor alpha (RARA); the gene that encodes the TGFalfa and beta3.

Many genes have been identified as involved in embryonic development of lip and palate: the alteration of the expression of each of these factors can make a potential contribution to the onset of cleft.

What now seems encouraging and very interesting is that in-depth study of the syndromic forms are aiding identification of gene mutations that represent a risk factor for isolated forms.

Teratogenic factors

The following environmental factors have been indicated in the published literature in the development of cleft lip and palate:
- Maternal smoking;
- Hypoxia of altitude;
- Alcohol abuse during pregnancy;
- Maternal nutritional status with special attention to the role of folic acid.

Whilst as there is evidence of a role of dietary supplementation of folic acid in the prevention of neural tube defects, less is known on the prevention of cleft

lip and palate although epidemiological studies seem to confirm that its deficit during pregnancy predisposes to the development of clefts. A randomized study that compared women who took high doses of folic acid during pregnancy (2-5 mg/day) and mothers who had used lower doses (1 mg/day) a higher prevalence of malformations with intake of high doses was seen so the issue remains controversial *(Cobourne MT, 2004)*.

The importance of continuing to improve knowledge on the etiopathogenesis of the disease and therefore the risk factors (genetic and environmental) with their interaction is the best strategy for the prevention.

2.
Multidisciplinary management of children with cleft lip and palate

The treatment of cleft lip and palate requires a team approach involving several specialists in order to obtain the best results both functionally and esthetically.

The concept of a multidisciplinary approach represents a fundamental evolution and is a key point in dealing with this type of disease and starts from birth or from prenatal diagnosis. It is now frequently used for defects that include cleft lip but less common in the treatment of isolated cleft palate.

The first advantage of the team approach is for the parents. The task of the team of specialists in charge of care is to reassure the family by explaining that the malformation is correctable and to overcome guilt feelings that the parents may have that they are responsible for this defect. The team can help the parents to overcome the period of initial confusion and focus their energy on creating an environment of positive support withing they could work.

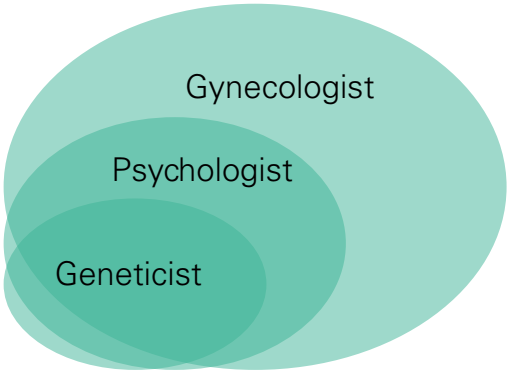
Forming a team of specialists also favors coordination between them which translates, from the practical point of view, in gaining time, cost reduction and saving of energy.

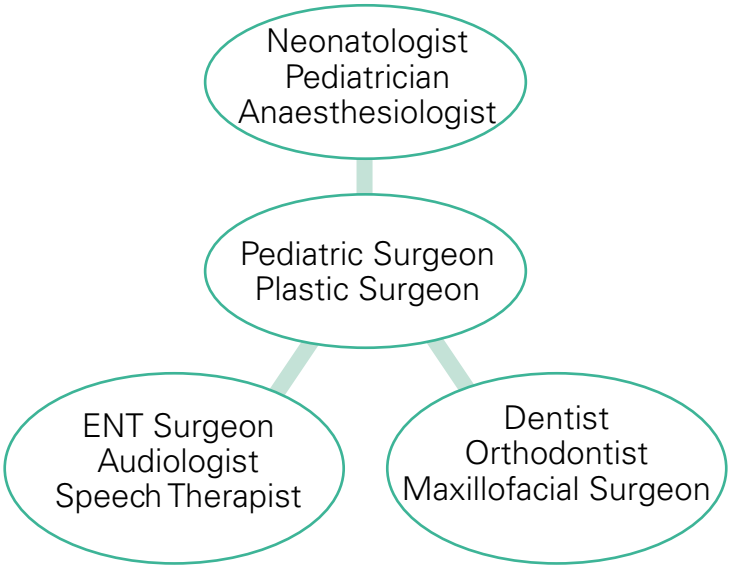
The task of these professionals will not be only to confirm the diagnosis, they will also investigate the presence of other congenital anomalies as part of a syndrome, in order to look after the clinical problems of both patient and family.

The main specialists within the team are the following:

- Geneticist, Gynecologist and Psychologist, involved in prenatal diagnosis;
- Neonatologist and Pediatrician;
- Pediatric Surgeon;
- Ear, Nose and Throat (ENT) Specialist;
- Audiologist;
- Speech therapist;

- Maxillofacial surgeon;
- Plastic surgeon;
- Dentist;
- Orthodontist;
- Anaesthesiologist.





Currently there are no clear guidelines as to which type of surgeon is best suited in terms of experience with the anatomical structures to reconstruct and carry out the closure of the defect. The official website of the Italian Society for the Study and Treatment of Cleft Lip and Cleft Palate and Cranio-Maxillofacial Malformations (www.silps.it) shows that in Italy the treatment of the disease is distributed almost equally between Maxillofacial and Plastic Surgeons. Few pediatric surgeons now deal with it. In the rest of the Western world the tendency is

toward the centralization of clefts patients to assure high level of surgical competence and performance. What seems to emerge from recent literature is that the initial source of training to treat cleft lip and palate is less important than experience. Specialists with extensive experience in treating a high-volume of cases, defined as surgeons operating at least 30 new cases per year seem best placed to treat these patients given their complexity *(Berkowitz S, 2013)*.

The **geneticist** is probably the figure most involved at an early stage, when parents are asking many questions about the causes, their possible responsibility and the risk of recurrence in future children. The geneticist has specific expertise regarding the pathogenetic mechanism of the malformation that can be decisive as a guide to therapy. The identification of the causal mechanisms of malformations, with their genetic and environmental aspects, can provide information not only for prevention but also in guiding treatment.

Knowledge of the pathogenesis, permits the most suitable timing of multidisciplinary therapy based on biological principles coordinating the various stages of therapeutic surgery with the moments of development of the structures affected.

The **ENT specialist** also plays a vital role in the early stages of evaluation of a cleft lip and palate. In the first months of life any pathology regarding hearing must be investigated. The diagnosis of hearing loss in children with cleft lip and palate is often difficult and delayed compared to the time when the malformation is recognized. The importance of hearing problems is often underestimated in early childhood when all the attention and concern of the family are for the more obvious anatomical defect.

The involvement of the middle ear is a very common condition in these children and the assesment of the aspect of the tympanic membranes is mandatory. In rare syndromic forms the cleft palate often has associated malformations of the ossicles or abnormalities of the inner ear with sensorineural hearing loss. If we exclude these cases the audiological disease is due to conductive hearing loss resulting from otitis media with effusion (OME).

The pathogenesis of the OME in children with cleft palate, appears to be related to a Eustachian tube dysfunction that causes insufficient ventilation of the tympanic cavity. The chronic hypoventilation and the recurrence of secretion determine a change in hypersecretive direction in the middle ear mucosa, setting the stage for chronic otitis media.

Otological treatment in children with cleft palate has the basic purpose of restoring ventilation of the middle ear, both to eliminate the secretion in place and to prevent exacerbations of the disease and possible chronic problems. The audi-

ologist may decide to augment the hearing range through less invasive and conservative interventions such as nasal washes, gymnastic tubal exercises (entrusted to the speech therapist) and speech treatments or more invasive procedures such as myringocentesis with or without placement of a drainage grommets. The hearing aid has showed to be a potentially good conservative treatment in the management of patients younger than 7-8 years old, whene there is still the possibility of a spontaneous improvment of the Eustachian tube function.

The essential initial task of the **neonatologist** and neonatal nurses is to identify and deal with any breathing and feeding difficulty and analyze the oral cavity as well as all the facial structures. This allows to recognize any further malformations, particularly micrognathia and macroglossia that can indicate Pierre Robin sequence.

Difficulties in feeding in a child with cleft lip and palate are typically directly related to the severity of the malformation. The staff of the Neonatology and the Pediatrician department should seek to maximize nutritional intake, promote the normal development of motor mechanisms of swallowing and a good parent-child interaction. The main objective is that of achieving an adequate weight to allow the surgical correction of the defect without excessive anaesthetic risk.

A patient with a cleft limited to the palate may suffer loss of the food bolus through the lips and excessive intake of air during sucking. In most cases breast feeding is possible instructing the mother to press the upper lip of the baby against her breast during lactation or against the teat of the feeding bottle.

The patient suffering from cleft palate poses a different and generally more complex problem being unable to create intra-buccal pressure to extract milk although good motor coordination is present. What usually happens is the input of milk in the nose and excessive fatigue of the baby during feeding with each feed being very lengthy. Aids created specifically to facilitate sucking are useful in this situation. The bottle is designed to increase the intake of milk and decrease fatigue and length of feeds. It is usually equipped with bulb teats with Y or X shaped holes and valves. The body of the bottle can be compressible to allow the parent to adjust the flow through pressure (links: www.medelabreastfeedingus.com, http://store.enfamil.com, www.bottlesandfeeders.respironics.com).

The breast feeding of patients with cleft lip and palate or isolated cleft palate, except minor forms affecting the soft palate, often present many difficulties.

It is advisable to breastfeed keeping them semi-sitting with the head in a neutral position so that gravity facilitates swallowing and reduces the nasal reflux. Equally useful are nasal aspiration and / or washing the nasal cavities with a syringe

after the meal. The use of a glass can be introduced as soon as the baby can sit up unaided, and it is often useful to let the child get used to a glass before surgery so that they can feed in the post-operative period without applying suction. Weaning and the introduction of the spoon may follow the normal developmental milestones. In some cases, especially in premature infants or those suffering from other diseases, it may be necessary to provide nutrition through a feeding tube.

Children suffering from cleft lip and palate are at high risk of developing speech disorders. This is due to both the anatomical and structural characteristics that distinguish them and the high incidence of recurrent otitis media and hearing loss that impact on the production and the understanding of language. The development of language in these patients may be further hampered by the orthodontic problems that are often associated with, other craniofacial malformations and by emotional and psychological aspects.

The malformation affects the articulation of sounds through the elements of the mouth (tongue, lips, soft palate and teeth), modifing the phonation that is the perfect balance between the vibration of the vocal folds and the sounds resonance throught the pharynx and oral/nasal cavity.

The major errors in articulation are related to a decrease in intra-buccal pressure resulting in distortion of the consonant sounds and an increase of the flow of air through the nose to compensate for the shortage. These compensatory mechanisms may worsen the intelligibility of speech output, so patients benefit greatly from phoniatric treatment and intensive speech therapy.

Another mechanism of compensation concerns phonation where patients reduce the volume of their voice to reduce its nasality. This can lead in the long run to the formation of vocal nodules that further modify the sound.

Instruments have been designed capable of assessing the extent of the voice disorder but the gold standard remains the human ear of the examiner; speech assessment guides have been created to facilitate the comparison of the analysis of language perception between different centres (eg. GOS.SP.ASS of Great Ormond Street Hospital).

The main aspects to be assessed are:

- Resonance according to which sounds such as B and D can be replaced respectively with M and N;
- The nasal emission of plosive sounds /p/ and /t/, of fricative /f/ and /s/ replaced with pharyngeal sounds;
- The nasal turbulence;
- The grimaces or unconscious movements with which the child tries to block the escape of nasal air;

• The acceptability and intelligibility of speech.

The most common disorder of the tone of voice is the hypernasality, given mainly by the inability of the velum to implement a suitable valve mechanism with the rear and lateral pharyngeal walls resulting in air leakage and distortion of many sounds such as: b-p-t-d-g-k-s-ò-tò-z-f (velo-pharyngeal insufficiency).

In general re-education should begin as early as possible after reaching the potential conditions for velo-pharyngeal occlusion.

The recommendations are to start treatment at around 36 months: before, the speech therapist's role is limited to observation of the child and counseling of parents. The speech therapy aims to correct and create the articulation, correct the resonance not caused by velo-pharyngeal insufficiency and eliminate compensatory articulation. Equally important is the recognition of patients with velo-pharyngeal insufficiency and to select those eligible for surgery. Finally, the speech therapist deals with the rehabilitation after surgical correction of this disorder.

Whilst the pediatric surgeon, plastic surgeon and maxillofacial surgeon may all be involved in the primary correction of a cleft, the latter is undoubtedly the most specific professional to operate on those patients in whom multiple surgical procedures and the malformation cause an abnormality of growth of the skeleton that often results in malalignment and / or maxillary hypoplasia. These abnormalities are increasingly treated by means of osteogenetic distraction. The maxillofacial surgeon is also involved in the treatment of palatine fistulas that can be a complication of early closure of the defect of the palate, but also a late complication that develops as a consequence of abnormal craniofacial growth. This specialist is involved in the evaluation and treatment of alveolar defects and / or velo-pharyngeal insufficiency.

Universal Parameters for Reporting Speech Outcomes in Individuals With Cleft Palate
hypernasality
hyponasality
audible nasal air emission and/or nasal turbulence
consonant production errors
voice disorders

(Henningsson G, Kuehn DP, 2008)

Dentists and Orthodontists are involved in the management of patients with cleft lip and palate from birth starting with the possible need for a device for suction. These are now used less and less with the advent of specially created baby bottles: they may realign alveolar segments prior to surgery and remain involved until adulthood. Their role increases with the eruption of the teeth, both the deciduous and then the permanent dentation. The dentist will have to take care to replace missing or malformed teeth and to remove supernumerary teeth and will also work with the maxillofacial surgeon in coping with frequent deficits of jawbone and the consequent malocclusion *(Kasten EF, Schmidt SP, 2008).*

3.
Primary surgical treatment

3.1 Surgical timing

At the age of three months, depending on the complexity of the defect, the surgical therapeutic programme starts usually with the reconstruction of the lip. In the past the first operation was generally scheduled following the rule of 10: baby weight greater than 10 pounds, blood haemoglobin greater than 10 g/dl, age 10 weeks and above. The current preferred strategy is to adjust the treatment and the timing to the individual patient which translates practically in a trend to early treatment in the majority of centres.

The closure of the palate is performed between the 8th and the 18th month, in two different moments for the soft and hard palate after a gap of a few months or in a single intervention. The functional role of the repair is attributed to the closure of the soft palate.

After the first surgical phase (lip repair, palate repair - one or two stages) several types of cosmetic or functional defect may persist. Dysmorphism is generally corrected after adolescence by secondary surgery, although some cases may require early intervention to reduce the psychological impact on the child and family. Following the latest protocols nasal deformities are corrected at the time of primary surgery, with possible adjustments at a subsequent time in order to achieve more aesthetically valid results.

3.2 The unilateral cleft lip repair

In cleft lip, there is a constant tissue deficiency that varies in size. To make sure that the reconstruction can be effective and as anatomic as possible it is essential that all the tissue available is both preserved and used.

The objectives of repair are:

1. Reconstruction of a normal anatomy and function of the lip. This includes the elongation of the lip, the construction of the Cupid's arch through the conservation of a segment of the cutaneous-mucous membrane line of the inner lip, the maintenance of a regular vermilion, the realignment of the orbicular muscle.
2. Reconstruction of the nasal floor using nasal mucus periosteal flaps from the side wall of the nose and the mucoperichondrial septum.
3. Repair of the nasolabial fistula.
4. Correction of the nasal deformity by bringing the base of the ala closer to the middle portion of the lip.
5. Narrowing of the columella.
6. Realignment of the maxillary segments.

The extent and severity of the defect are essential in determining which of these objectives will be met within the primary surgery and which require secondary surgery procedures.

Restoring a normal function of the orbicularis muscle is probably the most important aspect of proper reconstruction. The failure to achieve this goal with primary surgery as a rule requires a revision at a later time.

In the case of severe malformations it is more important than ever to have a thorough preoperative assessment. If there is doubt at this early stage that a goal can be achieved immediately this should be discussed with the family explaining that later surgical repair of residual deformity and secondary surgery may be necessary. Elements such as the degree of the cleft, the associated nasal deformity and the presence or absence of maxillary hypoplasia, allows an experienced surgeon to predict the outcome of the surgery. Other factors such as wound healing and scarring cannot be predicted and parents need to be informed of this.

The time of correction of the lip, according to international protocols, is 3-6 months old.

The development of surgical techniques has always been based on anthropometric measures that allow the comparison of the anatomical elements in physiological and pathological conditions. In the case of children a fourth dimension has to be applied, that of growth, in order to use retrospective anthropometric data and improve the initial three-dimensional design of nasolabial repair.

The measures most relevant related to unilateral cleft deformities concern the heminasal width (from the subnasal point, angle between the columella and the upper lip, the alar point, lateral end of the nasal ala), nasal width (from alar point to

alar point), the projection of the tip of the nose with respect to the subnasal point, the length of the columella on each side (from the subnasal point to the highest one of the columella), the amplitude of the columella (between the highest points of the columella of each side), the height of the lip (from the subnasal point to the peak of the Cupid's bow crest or bottom of the column of the philtrum on each side) and the length of the lip (from the peak of the Cupid's bow arc to the ipsilateral lip corner).

What makes the design of the surgical repair difficult is that all these structures will grow and that the anthropometric measures should be proportionate not so much in the immediate post-intervention but between 5 and 10 years of age of the child. Changes in these measures can occur in different ways on the side of the cleft and healthy side even after the cleft was correct. For example the heminasal amplitude tends to grow more on the affected side in the first 6 years of life (Mulliken-La Brie). Structures with slow growth such as the projection of the nasal tip and columella will be kept intentionally longer than in healthy children of the same sex and age, contrary to structures that grow faster such as the nasal width *(Sitzman TJ, Marcus JR, 2014)*.

Primary rhinoplasty

The purpose of primary rhinoplasty is to close the anterior nasal floor, to relocate the alar base and to give symmetry to the nasal base and tip. This approach allows for both a functional and aesthetic improvement without compromising nasal and facial growth.

After the cleft lip dissection is completed the muscles and soft tissues of alar base are separated from maxillary attachments. The lower lateral cartilage is separated from its cutaneous attachments by creating a medial and lateral superficial tunnel and the alar base can be repositioned.

Surgical techniques

In some cases the lip adhesion technique is still used. Its aims are similar to those of the pre-surgical orthodontic treatment and it is based on the closing by means suture threads of the tissue present on the two margins of the cleft and that is then discarded during the real surgical correction. Proponents support this procedure for the positive effect on alveolar defects and because it would seem to reduce the tension at the time of repair and ultimately increase the thickness of the orbicularis muscle. Unfortunately it requires additional anaesthesia for the patient as well as a further formation of scar tissue. The use or not of lip adhesion should be based not only on the preference and experience of the surgeon, but also on the preliminary evaluation of the amplitude of the defect.

The literature describes several methods of surgical repair of cleft lip, some have a purely historical interest: Le Mesurier in the 1950s and 60s was the first to describe an operation in which the analysis of the anatomy of the defect pre-surgery and planning of the flaps had an essential role. This opened the way for modern techniques which are still used, principally the one designed by Millard.

The Millard Rotation and Advancement Technique for unilateral labioplasty

Millard introduced this repair technique in 1955 with the main aim of moving the line of closure of the defect from the groove to the column of the philtrum itself where the scar is better camouflaged.

Over the years the technique of Millard has undergone several changes from both the author himself, and also of other cleft surgeons allowing the emergence of more and more variants corresponding to the individual needs of the case.

Among the advantages that have made it so popular are:

- The minimal tissue waste;
- The ability to hide the suture and re-create a Cupid's bow of normal appearance;
- Good access and good amount of tissue that the technique provides for the reconstruction of the alar base and the nasal floor;
- The versatility that allows modification of the planned incisions even after they have already been drawn if the result does not satisfy the surgeon.

Some of the disadvantages are:

- The potential retraction of the vertical scar;
- In case of severe tissue deficits excessive tension on the rotation flap that worsens scarring;
- The difficulty sometimes of rebalancing the vermilion that is thinner at the level of the previous cleft than vermilion of the opposite side.

The correction starts with the location of the essential points of reference (Figure 3.1).

Point 2 as the peak of the Cupid's bow on the healthy side and the point of maximum depression of the arc as point 1: the distance 1-2 is shown along the mucocutaneus line to the side of the cleft until point 3 is identified.

The point 4 is marked on the base of the healthy nasal ala side for determining the height of the lip (distance 2-4).

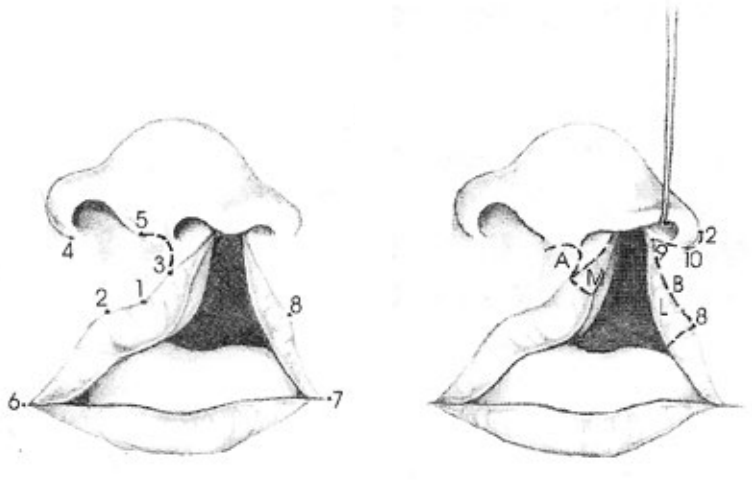


Figure 3.1 Reference points for the creation of the flaps in the Millard technique.

At this point the incision line is drawn for a rotation flap at C, starting from point 3, vertically towards the columella from the side of the cleft and then describing a curve that keeps very close to the base of the columella without reaching the crest of the philtrum on the healthy side (point 5).

Once the distance between the commissure 6-2 and end of the Cupid's bow has been determined this is marked on the side of the cleft from the commissure (7) until point 8.

This measurement can be performed with two compasses or better with a metal wire of exact length that can be shaped according to the profile of vermilion. The same technique is used to transfer the distance 3-5 on the side of the cleft to locate the point 9 that can be inside the nasal vestibul. The distance 8-9 represents the height of the lip on the side of the cleft (Figure 3.1).

The midpoint of the alar base position is point 10 which determines the arrival of the incision line starting from point 9. Often, in front of total cleft and an extent greater or less than the length of the nostril, the length of the incision can be prolonged beyond the point 10 towards the lateral end of the alar base (point 11), or even beyond, with circumalar trend, to reach a hypothetical point 12.

The incision of the line 8-9-10 and possibly 11 and 12 thus leads to the formation of the flap of advancement on the side of the cleft.

It is important in the Millard procedure to save the mucous tissue margins of the cleft. This is achieved by sculpting two edges L and M based on the mucosa of the upper vestibulum; the M flap is used for the closure of the cleft at the level of the nasal floor and sutured with the mucosal side facing the oral cavity.

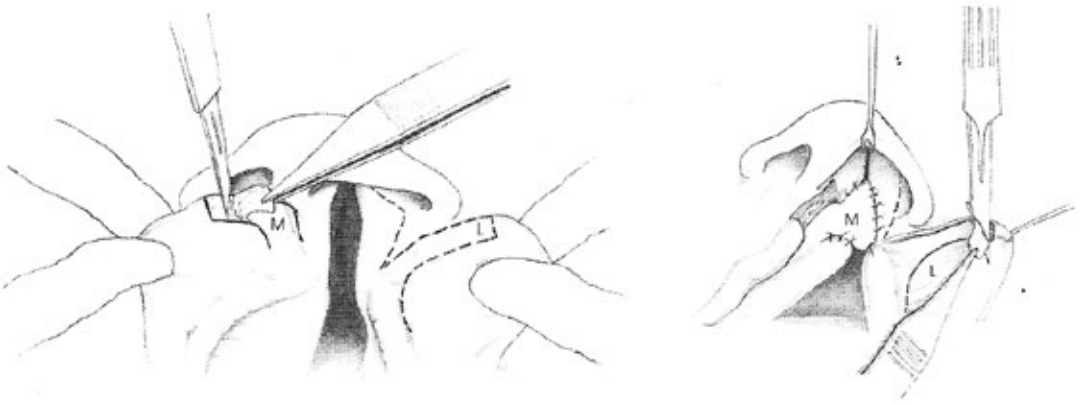


Figure 3.2 Dissection of the flaps for closing the nasal floor.

The L flap instead is rotated and sutured in the nasal vestibule at the level of the incision practised posterior to the upper edge of the alar cartilage.

The first incision is that for the inferior rotation along the line 3-5 which serves to establish the appropriate location of the Cupid's bow.

The amount of rotation required depends on the difference between the height of the lip on the two sides. The incision 3-5 is often not sufficient to align the top of the Cupid's bow on both sides for which it is possible to advance towards a hypothetical point X. This demonstrates the versatility of the technique of Millard as previously mentioned. Through this incision the alar cartilage is detached from the skin upwards and towards the controlateral alar cartilage allowing it to be able to recreate, after suitable repositioning, a good symmetry of the component parts of the cartilaginous framework of the nose.

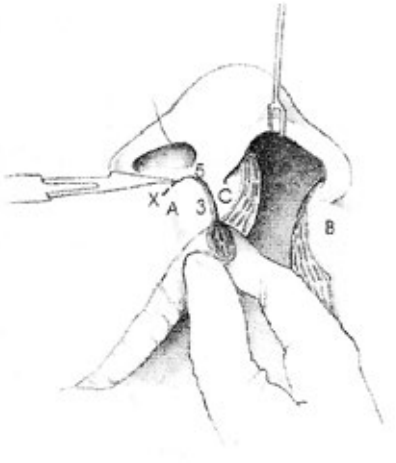


Figure 3.3 Creation of the first flap for the inferior rotation.

The incisions in the nasal mucosa can be placed according to a line, finishing just behind the top edge of the alar cartilage, bringing together the upper ends of the incisions practised in the modelling of the edges L and M.

The last step is the creation of the advancement flap on the lateral side of the cleft using an incision of the line 8-9-10 and possibly 11 and 12 together with an adequate incision of the nasal mucosa (Figure 3.4 A) *(Franchella A, 2007).* The latter has the purpose of separating the soft tissue at the level of their anchorage to the maxillary process and the nasal bone and to allow the proper sliding of the flap on the lateral side of the cleft. The dissection must be blunt and must proceed on a pre-periosteal plane, preserving the infra-orbital nerve and freeing the orbicularis muscle that can be repositioned and sutured to the medial lip element fibres.

Some authors (eg. Salyer) propose that the muscle must be repaired as an indipendent layer after being separated from its abnormal anchorages and from the skin, without a dissection which can promote excessive harmful scarring.

It is advisable to completely separate the inferior cartilage, both laterally and medially, from the mucosa and from the overlying skin, keeping the anchoring of these tissues only at the level of the nasal dome. These manoeuvres will allow the repositioning of the lateral superomedial alar cartilage prior to reaching a position symmetrical to the contralateral side. If asymmetry remains further dissection is required, and the L flap can be used.

At this point the three flaps ABC, if all the operations of disconnection have been correctly performed, are free to rotate to be repositioned as required for the reconstruction of the lip. The C flap is rotated and positioned in the small defect that is created on the shorter side of the columella and then sutured, in its front part, to the columella and posteriorly to the skin of the nasal vestibule (Figure 3.4 B).

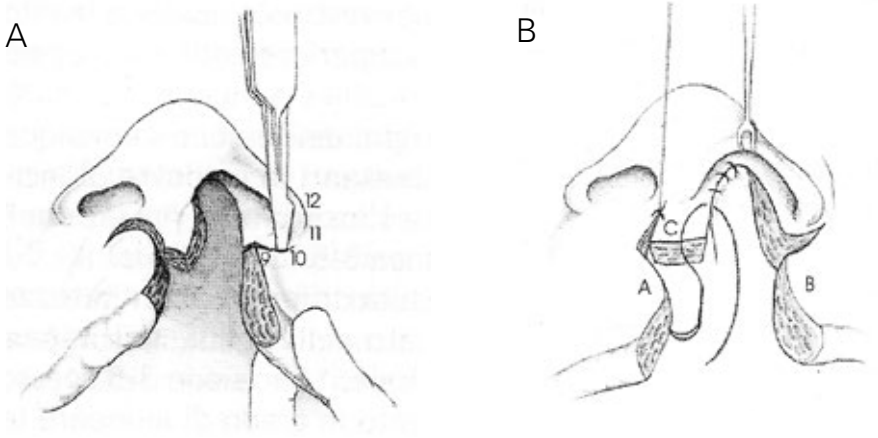


Figure 3.4 (A) Creation of the flap advancement. (B) Repositioning and suture the flaps.

First, instead of proceeding to the alignment of the advancement B flap with rotation A flap, it is necessary to model the two edges to make them compatible. The manoeuvre results in a regularization of the profiles that makes convex the edge of the A flap and concave the edge of B flap through modest excisions of the skin.

The sutures, with the correct pairing of the flaps, complete the procedure.

The main aim of this intervention is the creation of an elastic lip of symmetrical appearance with minimal scarring.

It is appropriate to use some technical devices in particular a non-absorbable monofilament suture (nylon 5 or 6.0) for the skin, which is then removed 5-6 days after surgery or absorbable monofilament of the same size.

After removal of the sutures the application of adhesive paper strips (eg. Steristrips) straddling the incision lines is useful. In ethnic groups at higher risk of hypertrophic scarring the patch can be maintained for a few months with good results. In alternative or in addition a daily massage of the scar with oil or a specific cream provides equally good results in preventing scar contracture.

The primary correction of nasal deformities, when these remain even after careful dissection of the tissue, is achieved by suspension techniques. Examples are tubular stents, fixed inside the nostril or suspended precisely with a stitch to the glabella to get the elevation of the nasal dome and a more physiological roundness of the arch (Mc Coomb, Salyer, Cutting, Stal). These manoeuvres can obviously create additional scars. It is therefore important to avoid an overcorrection of the nostril maintaining a nostril of a wider diameter on the side of the cleft because secondary correction of a too narrow nostril is more difficult *(Bardach J, Salyer KE, 1991).*

In an international comparison, held in 2005 between centres that dealt with cleft lip, it was found that 84% of surgeons used a variant of the Millard technique. Among the principal variants available, in addition to those introduced by Millard himself (for example for the primary correction of the nose). Mohler proposed in 1987 a variant of the rotation flap which is extended towards the junction between the lip and columella with the purpose of using the columella to stretch the medial portion of the lip. Stal began to use a notch at S to the rotation flap so as to increase the height of the medial lip. Mulliken, similarly to Mohler, introduced an incision to the rotation flap curved towards the margin of the cleft, extended towards the columella.

Unilateral cleft lip repair technique with triangular flaps according to Tennison and a variant according to Skoog

The principles on which this technique is based are:

- Creating triangular flaps for restoring a proper shape to the lip and the Cupid's bow;

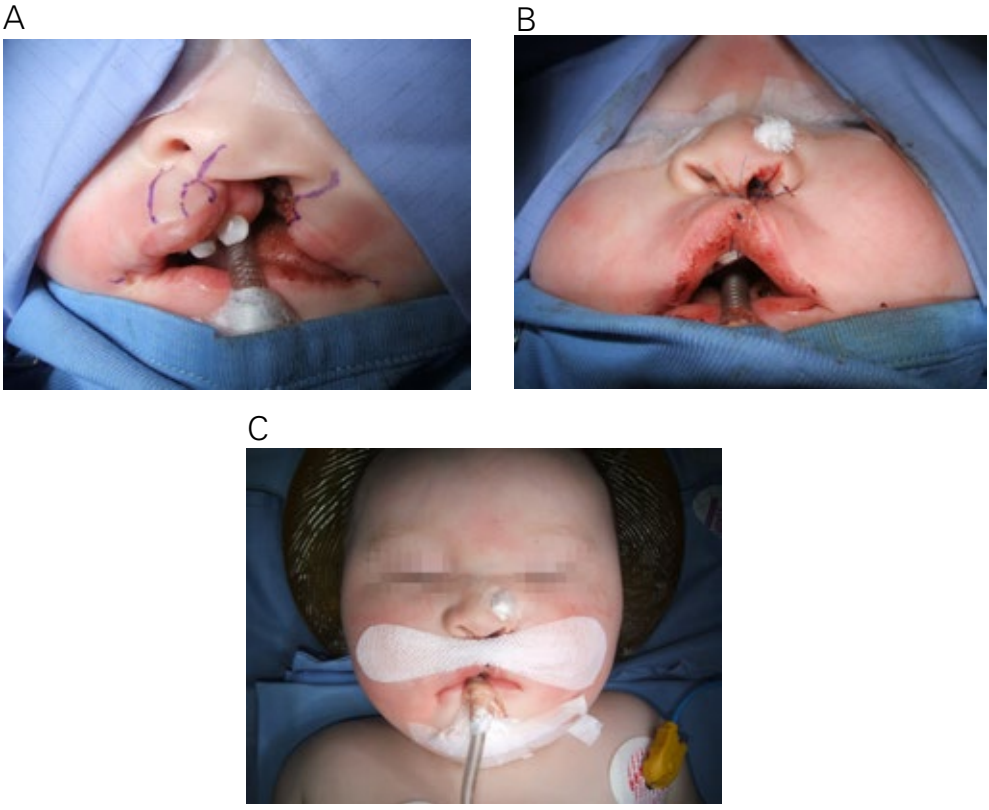


Figure 3.5 (A) Surgical incision lines for Millard labioplasty. (B) Post operative aspect with associated Mc Coomb rhinoplasty. (C) Dressing.

- Z-plasty suture to minimize the tendency to contraction of a vertical scar;
- Realignment of the orbicularis muscle on a transverse line;
- Reconstruction of the nasal floor;
- Simplification of the design and subsequent standardization of the procedure.

This last point makes easier this approach for those who for the first time have to repair a cleft lip with respect to the technique of Millard that gives the surgeon greater freedom of initiative.

The technique was later developed by Marcks, Hagerty and Randall, and finally by Skoog and Bardach.

This repair technique lost popularity in the 1970s and 80s with the advent of the Millard technique. A criticism often cited is that the scar alters the appearance of the column of the philtrum and often the philtrum itself if very wide flaps are necessary.

Key measures from which to create a symmetrical lip are the vertical size of the healthy lip and the distance between the labial commissure and the peak of the Cupid's bow from both sides.

As a first step, the point 1 and 2 are identified which respectively indicate the end of the base of the columella and the highest point of the Cupid's bow on the healthy side, by determining the height of the healthy lip.

Along the medial margin of the cleft the line 3-4 is drawn, making sure that 4 matches at the meeting point of the vermilion and the margin of philtrum in the cleft side. The incision line 4-5 perpendicular to the latter is then identified so that the sum 3-4 and 4-5 coincides with the measurement 1-2 in order to fill the height defect of the affected side (in 85-95% of defects this is about 4 mm). Typically these measurements are done with an excess of 0.5 mm to cope with the subsequent scar contracture *(Bardach J, Salyer KE, 1991)*.

On the lateral side the lines 6-7 of the same length of 3-4, 7-8 and 8-9 of the same length of 4-5 are drawn by placing 9 on the vermilion margin and 8 at suitable height to form an isosceles triangle (Figure 3.6 A).

For easy identification of point 9, which will become the highest point of the Cupid's bow from the affected side it is necessary to report the exact distance between the commissure and point 2 on the cleft side with an apposite compass (Figure 3.6 B).

It is important to ensure that the incisions on both sides have cut through the skin, muscle and mucosa to prevent scar retracting.

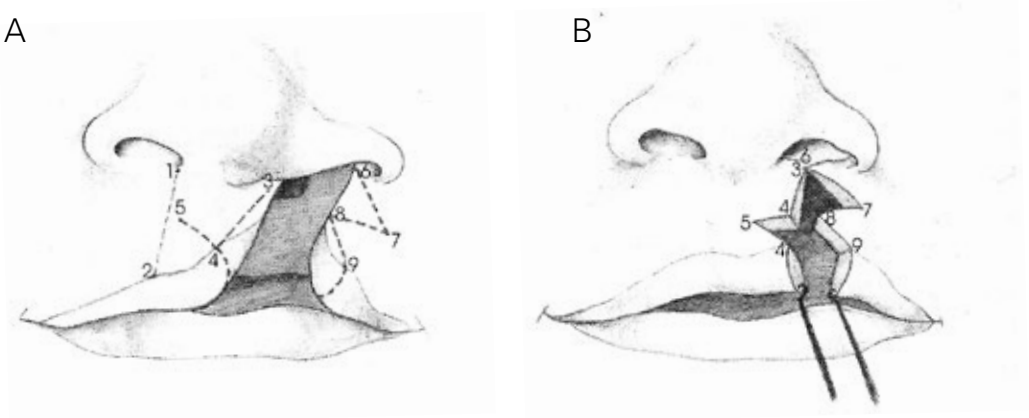


Figure 3.6 Reference points and incisions in the Tennison technique.

The variant proposed by Skoog identifies accessory landmarks that allow other incision lines for the creation of a small triangular flap at the base of the nostril from the side of the cleft (C1-C3) able to creep into the incision C2 (Figure 3.7).

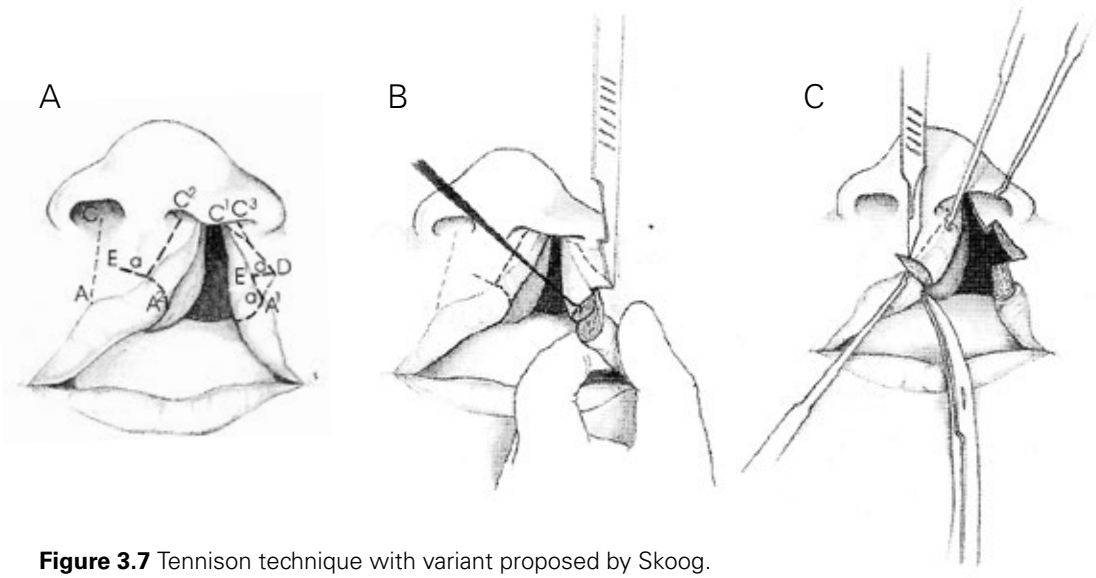


Figure 3.7 Tennison technique with variant proposed by Skoog.

As in other labioplasty techniques in the variant of Skoog the operations of detachment of the structures play an extremely important role in allowing a better sliding of the planes as well as the mobilization of cartilaginous parts constituting the nose.

It affects primarily the upper labial frenulum, generally very short, to make it longer, using a Z plasty. A further incision is made on the mucosal side dissecting the soft tissue from the periostal plane to creep up over the cartilaginous nasal structures and detach from the skin and reposition them in a more symmetrical and harmonious manner.

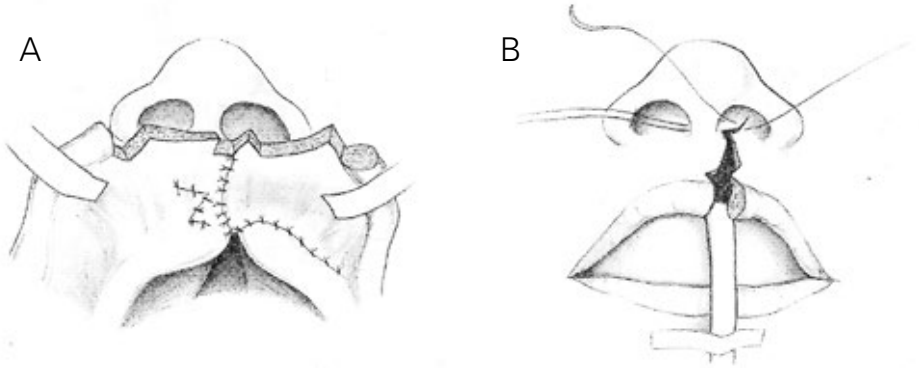


Figure 3.8 (A) Sutures on the mucous plane. (B) Approaching of the skin flaps for the suture.

It is evident in the ending appearance of suture the characteristic of the Skoog technique to recreate very specular planes of adhesion and to limit the tension.

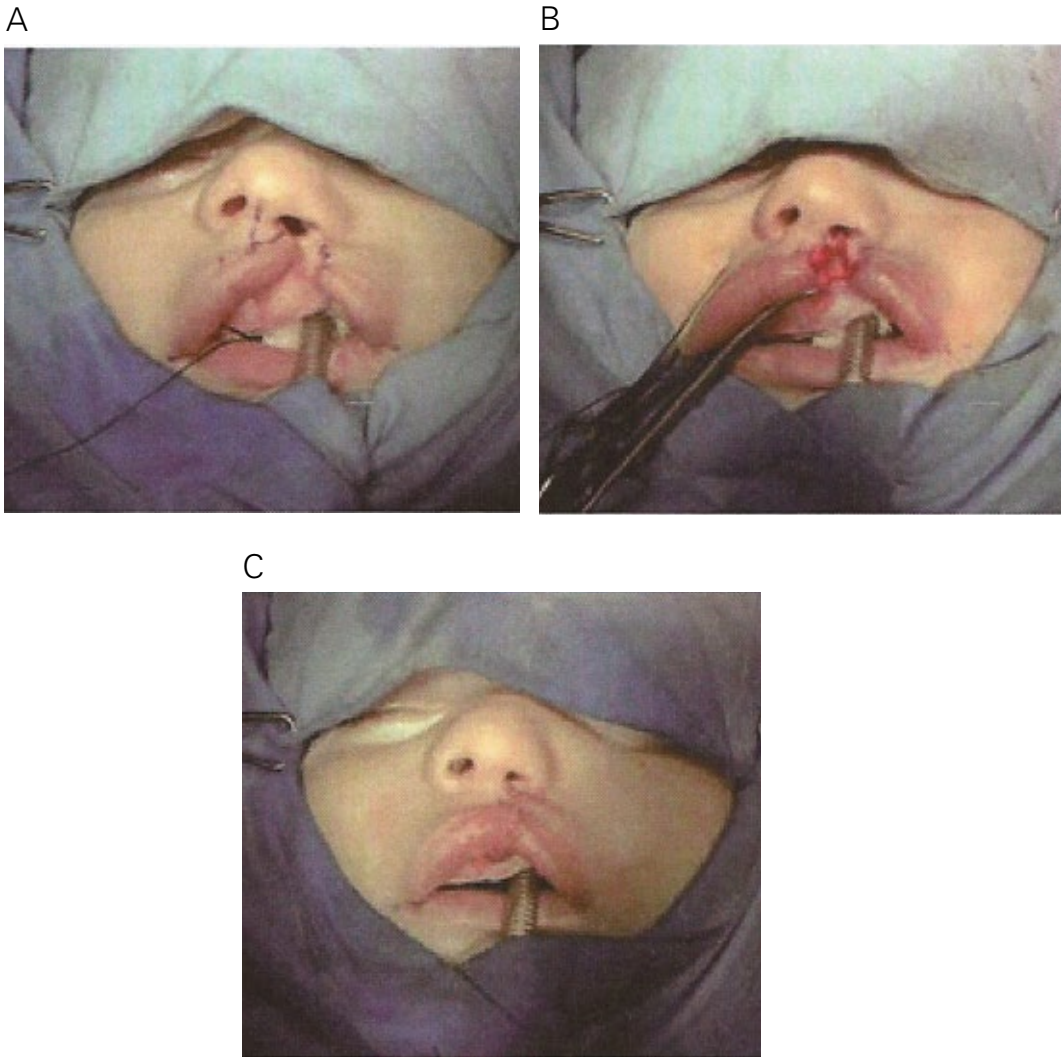


Figure 3.9 Labioplasty following the Skoog technique. (A) Design of the triangular flaps. (B) Creation of the flaps. (C) Appearance at the end of surgery.

The mucous plane is sutured first with separate stitches and knots facing toward the vestibule, following the same line in a Z-plasty pattern that becomes evident on the skin, then the skin surface and the muscle are sutured together starting from the vertices of the triangles *(Franchella A, 2007)*.

This technique is feasible in most unilateral cleft cases, however, when a wider triangular flap is required, the deformation of the philtrum columns will be

greater. One of the variants is to use two flaps instead of one when the sides of the triangular flap are longer than 5 mm.

3.3 Bilateral cleft lip repair

The incidence of all forms of bilateral cleft lip varies, according to case studies, from 9.6 to 16%. The bilateral forms are more common in males, with a sex ratio of 2:1 males to females. The bilateral cleft is more frequently associated with cleft palate.

Anatomy of bilateral defects

A bilateral cleft lip can be complete or incomplete on one or both sides. Any number of variations can exist.

The isolated bilateral cleft lip is the less severe form. It is generally characterized by the persistence of a connection between the lateral portions of the lip and philtrum (Simonart band) that is confined to the upper portion of the lip. It may comprise, in incomplete forms in addition to the skin, also all tissues that constitute the thickness of the lip including the muscle fibres, even if the latter do not penetrate the philtrum anchored to the skin at the level of the base of the nasal cartilages. The philtrum varies in size, typically has a semi-circular shape and can not present a clear boundary between mucosa and skin (mucocutaneous junction).

The more severe forms may be constituted by cleft lip and the alveolus only. More commonly, from complete cleft lip, the alveolus and palate. This form is by far the most frequent. In this case, the defect is most often asymmetrical, the premaxilla protrudes and can deflect and rotate toward one of the two sides, the philtrum is immobilized on the tip of the nose due to the protrusion of the segment below, the cleft palate is generally symmetrical. What determines the difficulty of correction of the deformity is the relationship between the pre-maxilla and maxillary processes, sometimes too close to allow the retraction of the isolated central segment without an orthodontic aid. The nasal deformity is more severe than in isolated labial forms and includes, in varying degree, the excessive brevity of the columella, the flare of the tip of the nose and space between the alar cartilages and, in asymmetrical shapes, septal deviation.

Surgical repair

The complexity of this malformation has made conventional wisdom, at least until the entire first half of the last century, that this surgical correction was much

more difficult than in the unilateral forms. Despite the expertise of surgeon, it was considered almost impossible to eradicate the "stigmata" of both intrinsic and iatrogenic origin , as the brevity of the columella, the convex profile of the upper lip, the irregularity of the philtrum where it is often difficult to recognize a mucocutaneous margin and the "festoon" appearance of the side of the lip which contrasts with a thin medial tubercle known as "whistling lip" deformity.

These traits have guided the search for principles that unite new surgical techniques, in particular:

- Symmetry that seems to be favored by a correction in several stages;
- Restoring a muscle continuity (the philtrum of patients is in fact without the orbicularis of the mouth);
- The construction of a philtrum of an adequate form and amplitude;
- The formation of the medial tubercle from labial side elements;
- Primary correction of position of the nasal cartilages to recreate the tip and the columella.

The debate is still underway regarding the possibility of closing in a single operation or at different times (first closing one side then the other or temporary closure of both sides followed by final closure at a later time) and using or not the philtrum. Recent decades have witnessed the emergence of two trends in particular:

1. The one-stage repair of the lip and nasal deformity;
2. Improved techniques for orthodontic alignment of the jaw.

Even with respect to the pre-surgical orthopedic treatment there is no agreement and some surgeons prefer to let the pre-maxilla grow to a protruded position up to 5-6 years, put it back together with bone grafting and then to correct the alveolar defect *(Mulliken JB, 2001).*

In addition to the protrusion of the pre-maxilla another obstacle can be hypoplasia of the philtrum. Several techniques are cited for growth promotion and to permit its use at the time of primary repair of the lip in the reconstruction of the central portion of the lip and the philtrum:

- Gentle massage 10 times a day for 1-2 minutes for a period of 6-8 weeks;
- Repair in 2 stages (ie. first the closure of one side of the cleft which increases the blood supply to the philtrum and at a subsequent time the correction of the contralateral defect);
- Positioning of a breathable patch that covers all the elements of the upper lip and the pressure of which would facilitate not only the retraction of the pre-maxilla, but also precisely the growth of the philtrum.

The lip adhesion has now been abandoned for this type of defect *(Bardach J, Salyer KE, 1991)*. Similarly techniques that use the philtrum in stretching of the columella or in the reconstruction only of the upper portion of the lip are now considered outdated.

In the literature there an almost total agreement for undertaking the primary repair at age 3-4 months to prevent growth and thus a excessive protrusion of the pre-maxilla. When a correction is made in several stages, the second operation is carried out 6-8 weeks after the first.

The decision on surgical strategy must be individualized according to the type of defect and experience of the surgeon. The elements that can hamper one-stage repair are the presence of a very small philtrum, an asymmetrical cleft and very protruding pre-maxilla.

Whatever the technique used it is mandatory that the primary restoration includes the reconstruction of the nasal floor (not always easy in the complete forms in which the pre-maxilla is completely separate from the alveolar arch) and the medial repositioning of the nasal ala. The correction of the other nasal deformities such as a lengthening of the columella, the creation of the nasal tip and the narrowing of the base of the ala can be carried out at different times, simultaneously with the primary repair or with the secondary repair, at 6-8 years of age or over (*Bardach*).

In bilateral cleft lip repair (single stage) it is useful to derive the reference points. One begins by marking the middle point of the mucocutaneous margin of the philtrum, the two peaks of the future Cupid's bow will lay laterally to this by 2.5-3.5 mm, at the base of the columella the philtrum will shrink 2-3 mm with respect to its lower edge. To place the 2 peaks of the arc it is important to take as a reference also the measure that goes from the lateral commissure of the lip at the point at which this begins to curve upward because of the cleft.

The Bardach Straight Line Technique

This technique, normally used for the closure in a single operation can be used in multiple stages, in cases of a very small philtrum or a columella excessively protruding: it exploits the rapid growth of the philtrum after the first operation. The skin of the central portion of the lip is specifically maintained wider at its upper part to allow the use of the excess in subsequent moments for minor corrections, such as for example the lengthening of the columella. The nasal floor is rebuilt as for unilateral cleft: the surface layer joining a muco-periosteal flap from the lateral wall of the nose and a muco-perichondral flap from the medial septum, the deeper layer by joining the lateral segments of the jaw to the pre-maxilla (not

always possible when the pre-maxilla is very protruding). In this operation it is important to reconstruct the nasal floor and places the alar bases symmetrically *(Bardach J, Salyer KE, 1991).*

Salyer modification of the Millard technique

The design of incision of the prolabium is performed so that the distance between the 2 points lateral to the mucocutaneous edge is about 6 mm and the width of the skin in the middle portion of the philtrum is higher than the latter by 2-3 mm. The two lateral flaps will be rotated upward to form part of the nasal floor; the mucocutaneous flaps from the lateral portion of the philtrum will inferiorly rotate to contribute to the vestibular lining, the remaining prolabial tissue is turned to reconstruct the vestibular sulcus. At the level of the lateral elements, the skin is minimally dissected leaving muscle in the vermilion flap to give fullness to the latter. This leaves the orbicularis oris muscle and lateral lip elements to be brought together from either side. The vestibular lining is closed by suturing the skin lining flap turned over from the prolabium with mucosal flaps from the lateral elements. The orbicularis oris muscle from both sides is joined in the midline, in front of the pre-maxilla when possible, or inserted in the prolabium so that the scar will fill in the muscle defect.

Lateral vermillion flaps are important in building up the central portion of the lip and for reconstruction of the tubercle. The skin flap from the central philtrum is sutured in its new location to form the central portion of the lip and to allow the permanent closure of the upper lip. The result foresees that the nose maintains a flattened and widened aspect that will be corrected only in a second stage towards 12 months old *(Bardach J, Salyer KE, 1991).*

MullikenTechnique for the simultaneous repair of the bilateral defect of lip and nose

The flap that will constitute the philtrum is designed with slightly biconcave side margins (for a 3-4 months caucasian baby to a length of about 6-8 mm, the width between the 2 peaks of the Cupid's bow of 3-4 mm and the width at the junction with the columella of about 2 mm). Laterally to the flap 2 skin strips are disepithelized to form the columns of the philtrum (the excess skin of the philtrum is then discarded). The two Cupid's bow peaks are established on the lateral elements of the lip to have enough skin to build the central depression of the philtrum and sufficient mucosa to constitute the medial tubercle. Then two triangular flaps are drawn with sides formed by the bases of the nasal cartilages and the mucocutaneous junction of the lateral elements. The incision lines are designed to create the

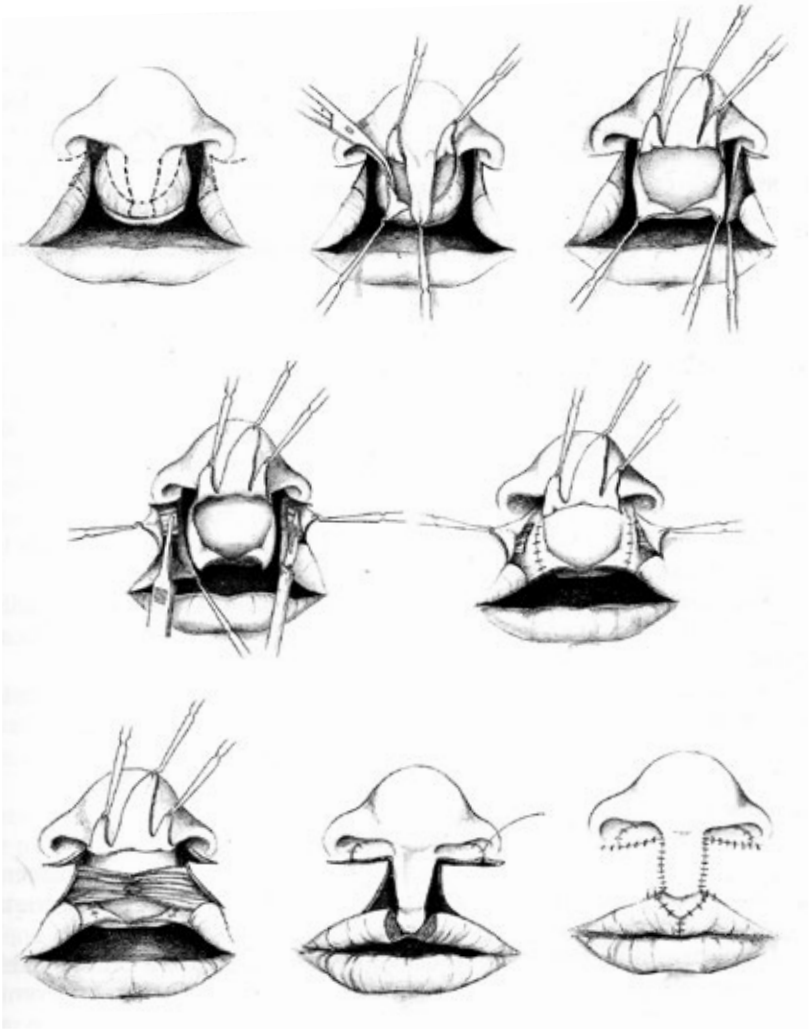


Figure 3.10 Salyer modification of the Millard technique.

flaps, the orbicularis is dissected from the lateral elements and the alar cartilages are exposed through the incision lines. The mucosal flaps of the lateral and medial portions are lifted so as to allow to juxtapose the mucoperiosteal flaps to close the alveolar defect. The mucosal flaps from the lateral portions will go instead to form the front wall of the vestibulum, on which is then juxtaposed the orbicularis muscle that will be sutured in the midline and anchored to the periosteum of the anterior nasal spine. The medial skin flap is secured to the muscle to create the typical philtrum depression. The redundant mucosal margins of the lateral elements will constitute the tubercle.

Before completing the closure suturing the skin flaps the correction of the nose is completed which foresees in this technique two incisions on the nostrils rims through which mattress stitches are placed between the two domes in order to approach the medial crus of the nose and to suspend the medial crus to the ipsilateral alar cartilage. These sutures help to bring the base of each cartilage up to a distance less than 25 mm. The vertex of each triangular flap to the alar base is secured to the underlying muscle medially so as to give a rounded shape to the nostril and prevent the elevation of the ala when the baby smiles.

At this point the excess skin can be excised and the skin flaps can be sutured completing the closure of the upper lip.

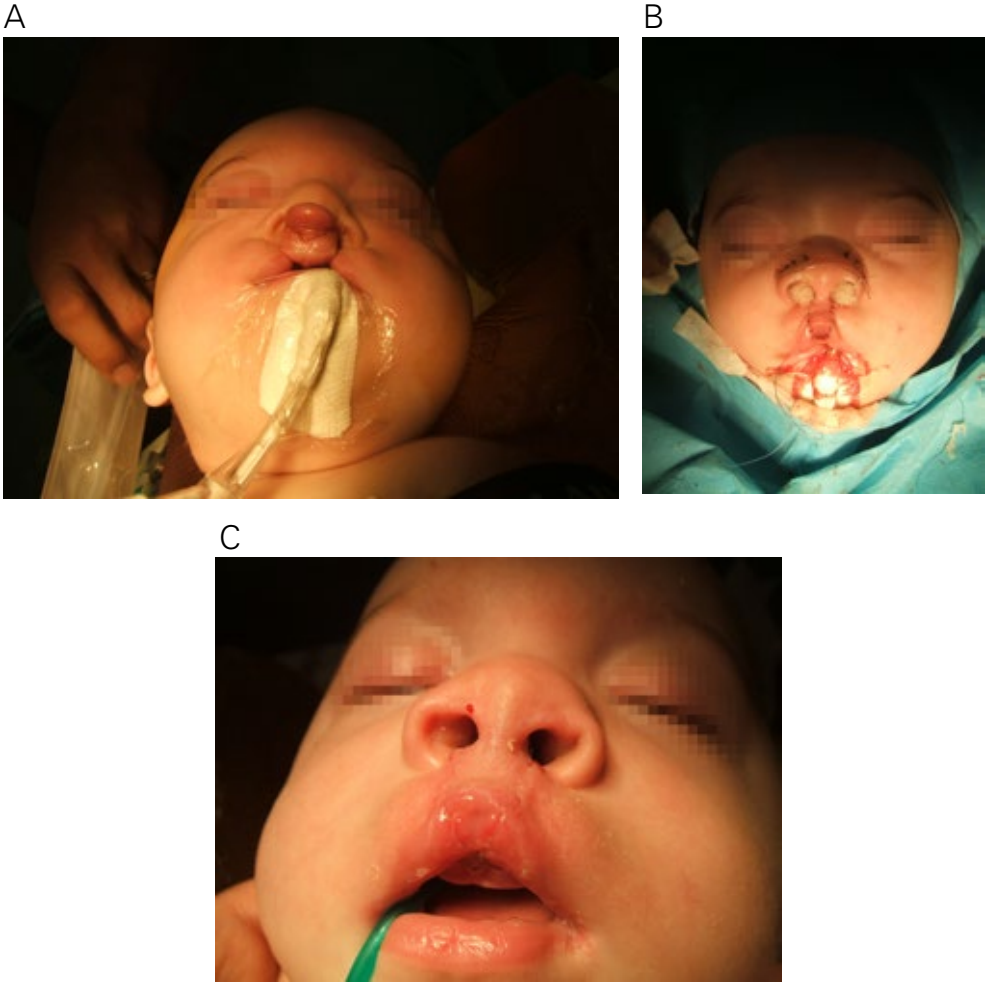


Figure 3.11 Bilateral cleft lip repair.

This technique reduces the need for secondary surgical revision procedures. The most common reasons that lead to further surgical intervention are the redundancy of the mucosal lateral elements ("whistling lip"). The correction of this nasal deformity must be planned after the eruption of permanent incisors and when the pre-maxilla have joined the final position.

3.4 Cleft palate repair

From birth the palate is involved in key functions such as swallowing, feeding, speech development, maintaining air flow and the control of the middle ear function.

From a functional point of view the palate can be divided into 3 areas: the anterior palate (25%), mid-palate (50%), posterior palate (25%). The elevator veli occupies the central portion originating from the eustachian tube, the right and left body of muscle on the midline of the velum acting as motors. The palatopharyngeus muscle is located in the same portion, it originates from the top of the buccal surface and from the nasal surface and runs inferiorly until the posterior tonsillar pillar. The palatoglossus originates from the posterolateral transverse fibres of the tongue forming the most superficial muscle of the mid-palate anchored to the anterior tonsillar pillar.

The anterior portion of the palate is more static and includes the tensor veli or of the palate muscle that originates from the eustachian tube and pterygoid hamulus and ends with an aponeurosis on the anterior part of the raphe. The palatopharyngeus muscles, palatoglossus and uvula are instead part of the remaining 25% of the posterior palate.

The elevator of the palate is a transverse and horizontal muscle curtain suspending the soft palate. When it contracts it pushes the palate postero-superiorly and laterally facilitating the closure of the velo-pharyngeal sphincter. In the case of clefts the elevator fibres run in antero-posterior direction anchored to the posterior margin of the bony palate and this prevents its main function. There are 2 other abnormal insertions of the muscle that must be interrupted during the correction of the defect, one with the aponeurosis of the tensor muscle and the other lateral one with the superior constrictor of the pharynx.

The most likely synergistic role of the elevator and tensor muscle of the velum in dilation of the eustachian tube also explains the functional deficits that patients with cleft palate often have *(Sitzman TJ, Marcus JR, 2014)*.

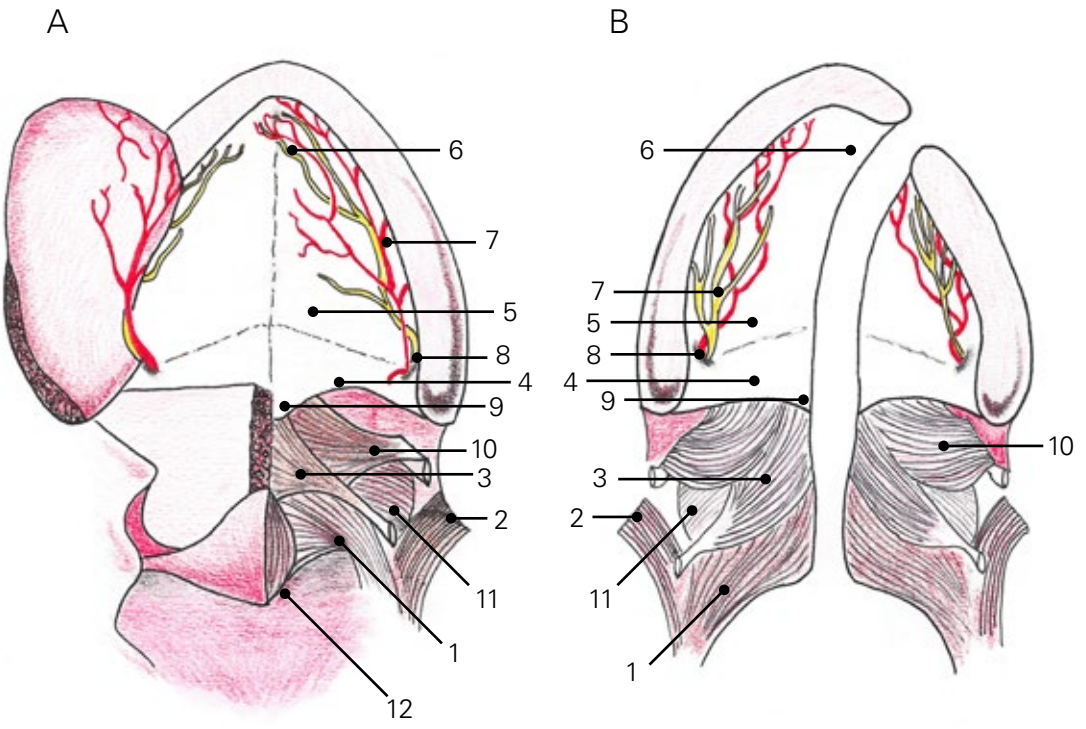


Figure 3.12 (A) Normal anatomy of palate and of velopharingeal sphincter. (B) Unilateral cleft palate.

(1) Palatopharyngeus muscle.
(2) Pterygomandibular raphe and superior constrictor muscle.
(3) Palatoglossus muscle.
(4) Horizontal Bone of Lamina Palatine.
(5) The process of the maxilla.
(6) Incisive foramen.
(7) Spheno-Palatine vessels and nerve.
(8) Greater palatine foramen.
(9) Posterior nasal Spina.
(10) Pterygoid hamulus and tensor veli palatini.
(11) Levator veli palatini.
(12) Musculus uvulae.

The primary objective is to achieve a functional closure of the palate so as to ensure proper development of the jaw, preventing recurrent ear infections (and any associated hearing loss), to allow a normal language development and to improve swallowing.

Generally the techniques for the reconstruction of the palate pose some basic purposes:

1. Restore the normal anatomy of the palate muscles;

2. Extend the palate thanks to the reconstruction of the anterior soft palate (palatine aponeurosis);
3. Separate the nasal cavity from the buccal cavity to stop the reflux of liquid and solid foods through the nose;
4. Perform a closure of the defect free from tension to prevent scars and fistulas.

The need to promote speech and not stifle the growth of the jaw are the elements that guide the choice of therapy and timing.

Traditionally, those in favor of an early correction of the defect (first year of life) use as an argument the need to create a fertile ground from a functional point of view to allow the development of language. Those against the problem that the early closure, especially of the hard palate, inhibits the growth of facial bones. This would seem to be true in particular in techniques that involve leaving exposed large areas of bone waiting to heal and where the sutures generate tension *(Bardach J, Salyer KE, 1991).*

Fears about the physiological growth of the maxilla have led over the years to several surgical approaches. Among these approaches is the concept of primary and early closure of the soft palate, followed after several years by the closure of the hard palate (9-14 years, Schweckendiek). Subsequent studies have revealed a high incidence of velo-pharyngeal incompetence thus closure in two stages has been modified especially by Perko both from a technical and timing point of view with closure of the hard palate between 18 months and 5 years of age.

The literature does not give clear directions with a extreme variability of timing and surgical techniques. Rohrich (2004) and colleagues have detected a significant language deficit in patients undergoing late closure of hard palate (48.6 months) than earlier (10.8 months) with a lack of improvement in the facial skeleton growth in the delayed intervention. A similar observation also emerged in another study of Robertson and Jolleys (1974).

The current trend, even with the changes provided by different protocols, is to correct the cleft palate at the age of 6-18 months. When the correction is at an early age two surgical operations are preferred, completing the correction within the first two years of the child's life.

Surgical techniques

Regardless of the technique used, the key elements to consider are the palatine artery respect and the liberation of all the aponeurotic muscle insertions of the posterior border of the hard palate and pterygoid hamulus.

The preoperative assessment to decide the surgical technique should consider:

- The width of the cleft and its form (V or U, the latter rarer but more difficult to correct);
- The position of the jaw segments and the degree of alveolar defect for complete cleft lip and palate;
- The possible extension of the cleft in the hard palate and its entity;
- The position of the lower edge of the vomer;
- The inclination of the palatine processes;
- The length, size and symmetry of the soft palate and its distance from the posterior pharyngeal wall;
- The degree of motility of the lateral pharyngeal walls;
- The size and condition of the tonsils *(Bardach J, Salyer KE, 1991)*.

As with cleft lip, cleft palate also has a huge range of corrective possibilities. Every author has proposed a new technique with several variants. The following are the most used today.

Whatever the technique it is advisable to place the patient in mild Trendelenburg with neck hyperextension, obtain a good exposure of the defect by means of a gag type Dingamm and, before starting with the incisions, infiltrate the area with a ready to use commercially available local anaesthetic and adrenaline solution in order to reduce bleeding.

Von Langenbeck palate repair

This technique is based on the preparation of 2 bipedicled mucoperiosteal flaps and on the principle, already introduced earlier by Dieffenbach to mobilize the tissues lateral to the cleft through releasing incisions, moving them and fixing them along the midline.

In detail, the purpose of the intervention is to unstick the tissues proceeding with an incision at the level of both sides of the cleft in the anterior-posterior direction. The lateral relaxing incisions start rostral to the base of the anterior palatine pillar and pass medially to the palate border near alveolar ridge.

Once the incisions have been created the tissues are moved using a periosteal elevator that can pass between the bone surface and the periostium, inserting them from lateral incisions, and always following a trend from the hard palate anteriorly back to the soft palate.

During this procedure the posterior palatine vessels should be respected to avoid the intense bleeding caused by its accidental interruption.

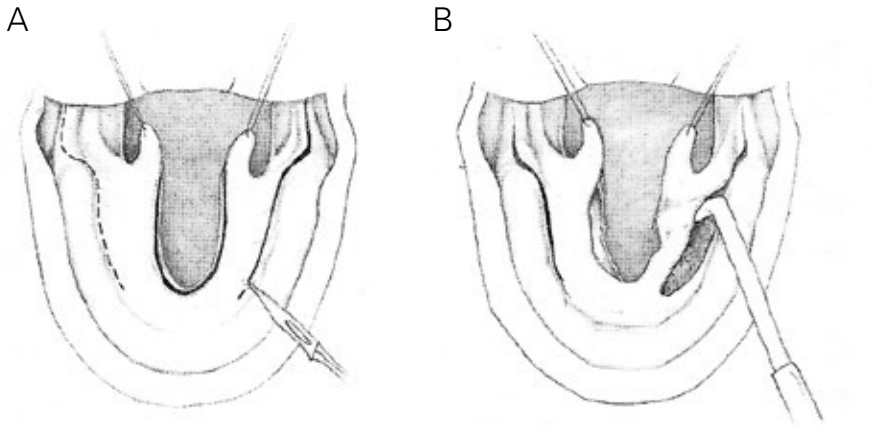


Figure 3.13 Von Langenbeck palate repair. (A) Lateral releasing incisions and then incisions of the edge of the cleft. (B) Detachment in the anterior to posterior direction.

Incisions are made on the medial margins of the cleft so to separate the nasal mucosa from the buccal mucosa.

If the detachment is performed correctly, the two side flaps thus created will be brought to meet in the midline without difficulty or stress.

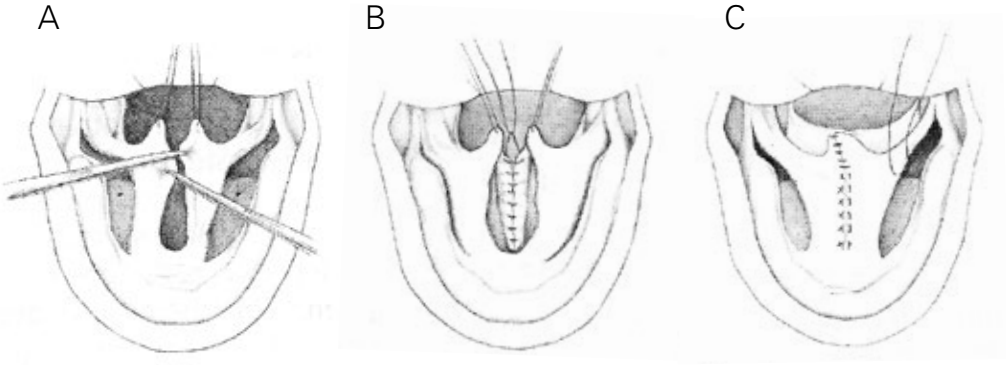


Figure 3.14 (A) Elevation of bipedicled flaps. (B) Suture of the nasal mucosa. (C) Suture of the muscle and oral mucosa.

The first layer of the mucosa that is sutured along the midline is the nasal that is then detached from the underlying bone surfaces and gently brought forwards to the contralateral portion to create the floor of the nasal palate sometimes incorporating a mucosal flap from the vomer.

After addressing the previously incised posterior pillars, the suturing continues with the oral floor moving up towards the uvula. After the reconstruction of the palate, its surface tends to remain raised from the bone plane. To ensure adherence, two stitches are placed between the lateral margin of the flaps and the soft tissue posterior to the palatine lamina. This allow to bring together the nasal floor and the oral floor of the palate and reduces the tension on the medial sutures.

One Stage palate repair

With the closure of the palate in a single operation it is possible to treat different forms of cleft:

- Complete with medial cleft;
- Complete unilateral cleft with insertion of the vomer on the healthy side;
- Bilateral cleft palate without protrusion of the premaxilla;
- Bilateral cleft palate with protrusion of the premaxilla.

One of the advantages that must be taken into consideration in the therapeutic decision is the fact that the correction of cleft palate with a single step inevitably reduces the rate of morbidity and mortality for the patient.

The Von Langenbeck technique gives closure in only one intervention but not for large defects in order to avoid excessive exposure of bone with lateral incisions. Most often it is used for the correction of isolated cleft of the soft palate or for the completion of the anterior palate in modest defect cases where closure of the posterior palate has already occurred.

The techniques to be considered are mainly two: the closure of the palate with two flaps or four flaps. Both use the fundamental principles of palatoplasty:

- Mobilization of the mucoperiosteal floor of the nasal side;
- Mobilization of the mucoperiosteal floor of the oral side;
- Identification and preservation of the neurovascular bundle;
- Synthesis of the mucoperiosteal plans to fill the defect;
- Elevator muscle mobilization from each side of the cleft palate and restoration of its morphological and functional integrity;
- Spontaneous healing of any areas of bare bone exposed by the trasposition of flaps.

Four Flap palate repair

The four flap technique is used in cases of complete cleft palate (posterior and anterior palate) with medial vomer.

The incision lines are designed to delineate the four corners of the oral side of the cleft, or two flaps in the front top and two at the rear top.

The mucoperiosteal flaps will be mobilized and pulled to fill the defect.

In general the technique using four flaps is not very versatile and not devoid of complications such as:

- Important scar retractions;
- Growth alterations of the anatomical structures involved in the cleft;
- High incidence of fistulas and orthodontic problems.

Two Flap palate repair

The two flap technique is the most versatile surgical technique to treat cleft palate in a single operation.

The technique exploits the principle that the detachment of mucoperiosteal flaps cause their rotation by an inclined plane to a horizontal plane and this allows the surgeon to exploit the full breadth and, in most cases according Bardach and Salyer who prefer this approach, this means he can close the defect on the midline without the need to drain and then without incisions that expose the bone.

Two incisions are drawn along the medial border of the cleft between the oral and the nasal mucosa (distinguishable because slightly darker), exposing the muscles. From the apex two divergent incisions are started towards the alveolar processes, at the canine level, the incision line continues from here with the lateral incisions that are extended up to about 1.5 cm posteriorly to the maxillary tuberosity. This creates 2 V-shaped mucoperiosteal flaps. The dissection is carried out in the latero-medial direction with the help of a periosteal elevator preserving the neurovascular bundle which exits the great palatine foramen (in case of large defects the bundle can be stretched by means of blunt dissection to separate it partially from the flaps and increase the mobility of the soft palate).

Even in soft palate limited clefts, the detachment of the nasal mucosa from the posterior margin of the hard palate and the dissection of the soft palate muscles are manoeuvres necessary to promote the reconstruction of the palate in a more posterior position.

The closure begins with the combination of the nasal layer in the anterior-posterior direction up to the uvula so that the nodes remain on the nasal side, then pass to the combination of oral mucosa and muscular layers with stitches in a mattress fashion. At the end of this operation generally no exposed bone remains at the lateral incisions.

In the case of a defect which includes the hard palate the procedure is similar: the anterior divergent incisions are conducted until the alveolar process at the level of the lateral incisive. To decrease the tension, especially in the case of large defects it is necessary to peel off the oral mucoperiosteal flap back to the

maxillary tuberosity. Equally important to the elongation of the soft palate and a correct closure in layers is the detachment of the muscles and the nasal mucosa layer from the posterior medial margin of the hard palate.

Whatever the defect, after closing the flaps on the midline, this should allow the positioning of the closing stitches on other margins without difficulty. If this creates a tension the source should be investigated and eliminated by expanding the detachment.

In the area of the hard palate it is important to fix with "mattress" stitches the oral and nasal layer to prevent the creation of dead space.

If despite the efforts the combination of the mucosa is not possible along the lateral incisions and bone remains exposed, it is advisable to protect these areas with cellulose polymer gel or fibrillar to facilitate healing even if it is not shown that this prevents abnormal scarring.

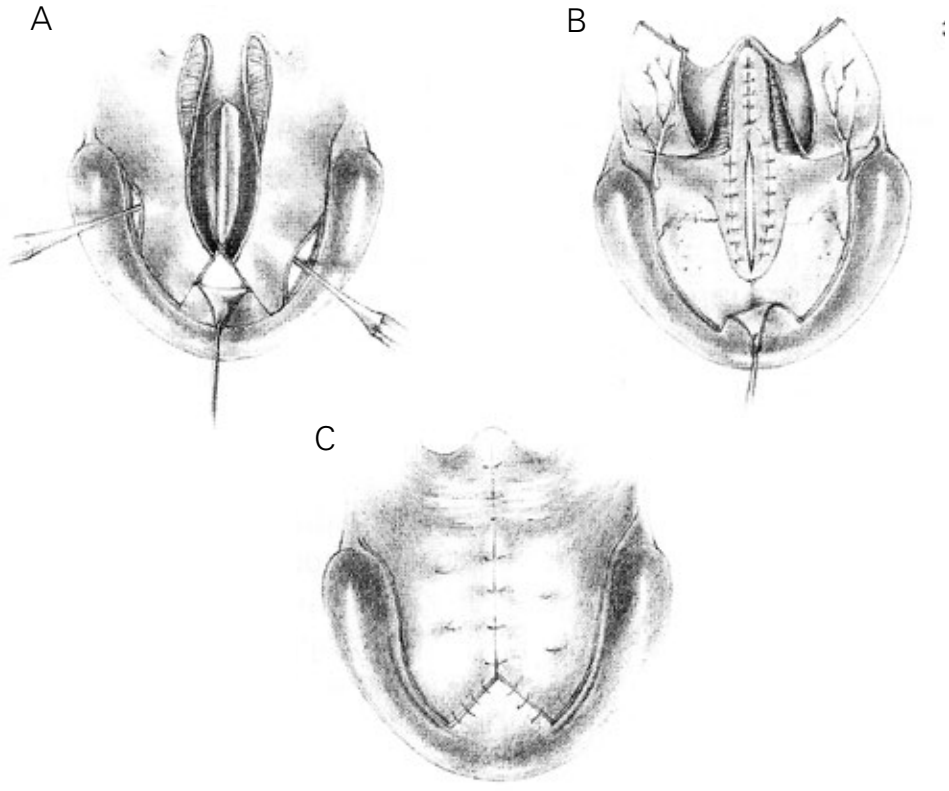


Figure 3.15 Two Flap palate repair. (A) Elevation of mucoperiosteal flaps. (B) Closure of nasal mucosa. (C) The muscle and the oral mucosa are sutured in the midline.

For the treatment of bilateral forms the surgical steps remain the same except that for the synthesis of the nasal floor in which it is necessary to use two mucoperiosteal flaps from vomer (variant introduced for the first time by Campbell and Pichler, then from Widmayer). When the vomer is located on the midline and its posterior edge reaches beyond the soft palate it is possible through a midline incision to raise two mucoperiosteal flaps that are sutured to the nasal mucous periosteum of each side creating a nasal layer with low tension *(Bardach J, Salyer KE, 1991)*.

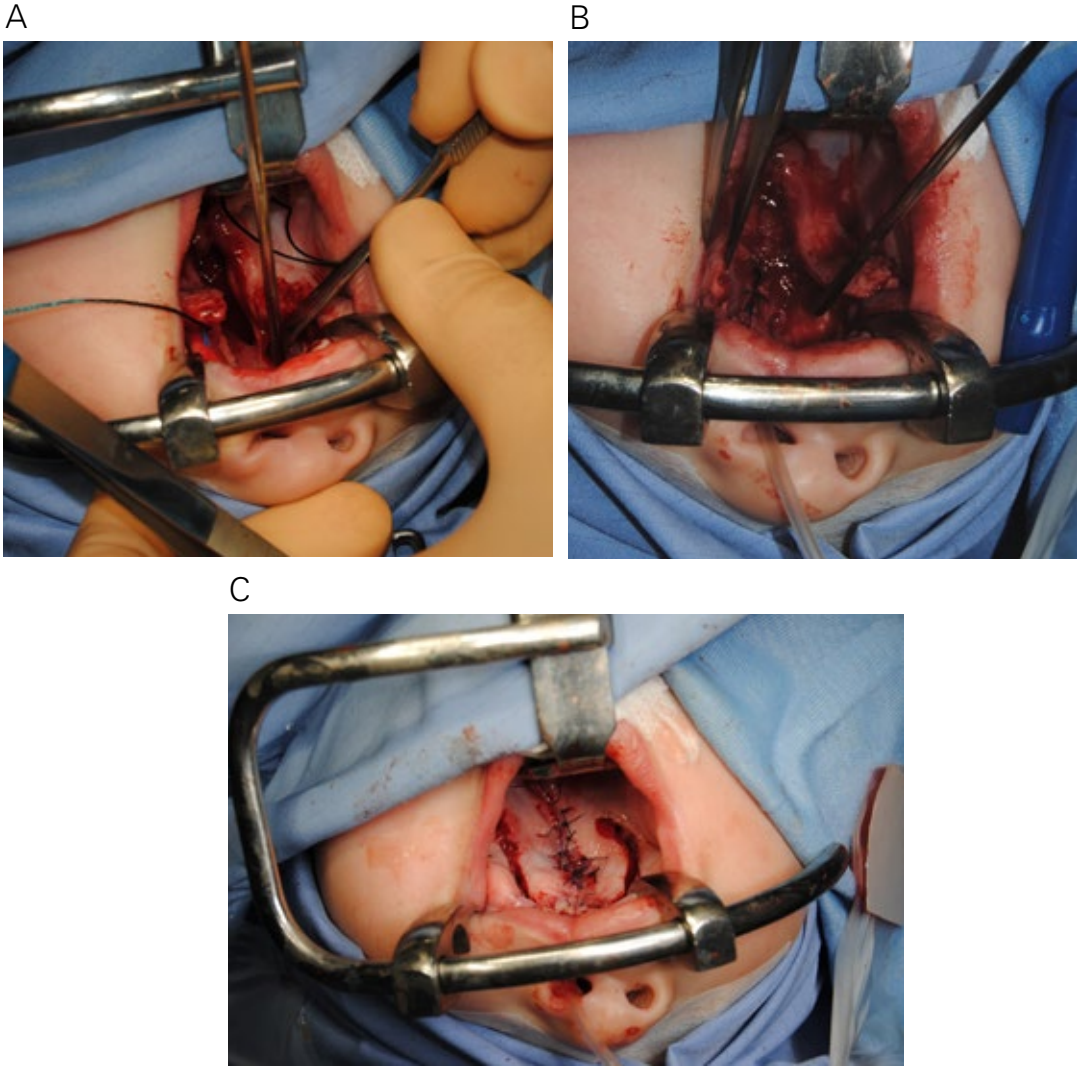


Figure 3.16 Two Flap palate repair. (A) Detachment of the flaps. (B) Suture of the nasal mucosa. (C) Appearance of the palate at the end of the surgery.

Palatoplasty techniques in a single intervention are not always advisable in cases of large cleft and also in cases of smaller defects it is still necessary to perform an extensive mobilization of the mucoperiosteal flaps.

Two Stage palate surgery, according to Widmayer Perko

The technique is quite versatile and has the peculiar characteristic of being able to be used in complete unilateral cleft palate and precisely in the defects involving both the primary and secondary palate.

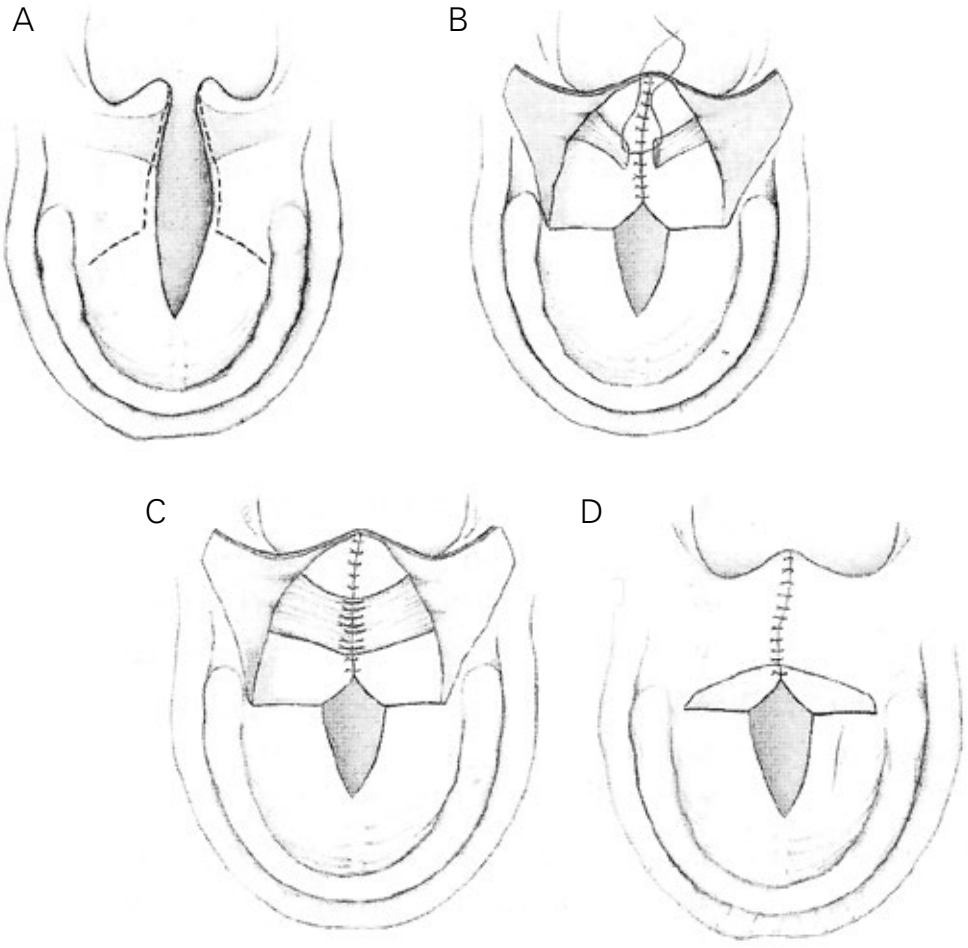


Figure 3.17 Closing the soft palate according to Widmayer Perko. (A) Incision lines. (B) and (C) Closure of the nasal floor and reunification on the midline of the muscles. (D) Suture of the oral floor.

The Widmayer-Perko two stage palatoplasty is mainly based on the principle, already established in 1955 by Slaughter and Schweckendiek, that the closure of the posterior palate facilitate a spontaneous approach of the anterior defect margins (mechanical action assisting the recovery of the functionality of the elevator muscle of the palate).

The medial edge of the cleft is incised starting from the posterior third of the hard palate to encourage better detachment. The oral plane is detached from the nasal plane and the muscles are released from the posterior edge of the hard palate. The lateral incisions begin about 1.5 cm posterior to the maxillary tuberosity and proceed forward for 1-1,5 cm remaining along the medial border of the alveolar process. This creates two bipeduncolated flaps.

The dissection proceeds as in the previously described techniques to allow to bring together on the midline prior of the nasal mucosa and then of muscle and oral mucosa together.

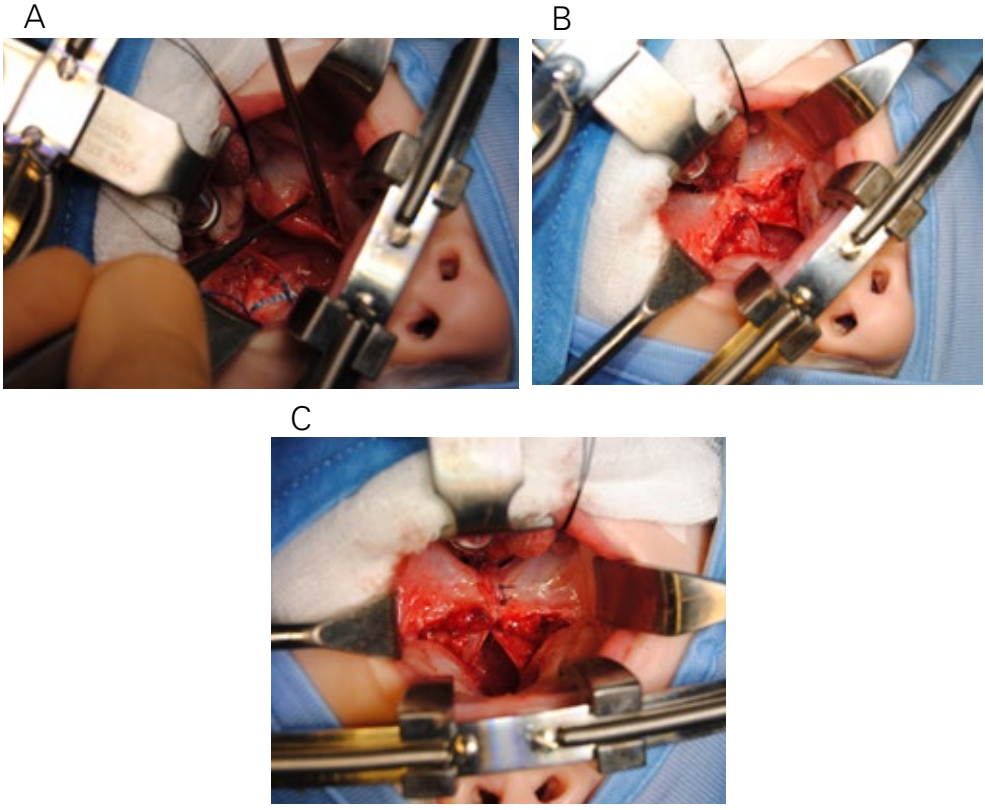


Figure 3.18 The first stage of palate repair according to Widmayer Perko. (A) Preparation of the flaps. (B) Closing in a double layer. (C) Appearance of the palate at the end of surgery.

After this first intervention the defect of the hard palate may shrink to the point of being reduced to an oro-nasal fistula which can be closed with a minimum detachment of the edges or even if there are no functional implications, treated conservatively. In other cases the anterior defect may remain unchanged or even worse because the palatine processes take on a more vertical path, and then a true second operation is required.

In this case the medial edges of the defect are incised and then mucoperiosteal flaps created similar to those illustrated for the two flaps technique making sure to properly detach the scar area located in the front portion of the previous palatoplasty so as to allow a medial suture of flaps in a double layer *(Bardach J, Salyer KE, 1991)*.

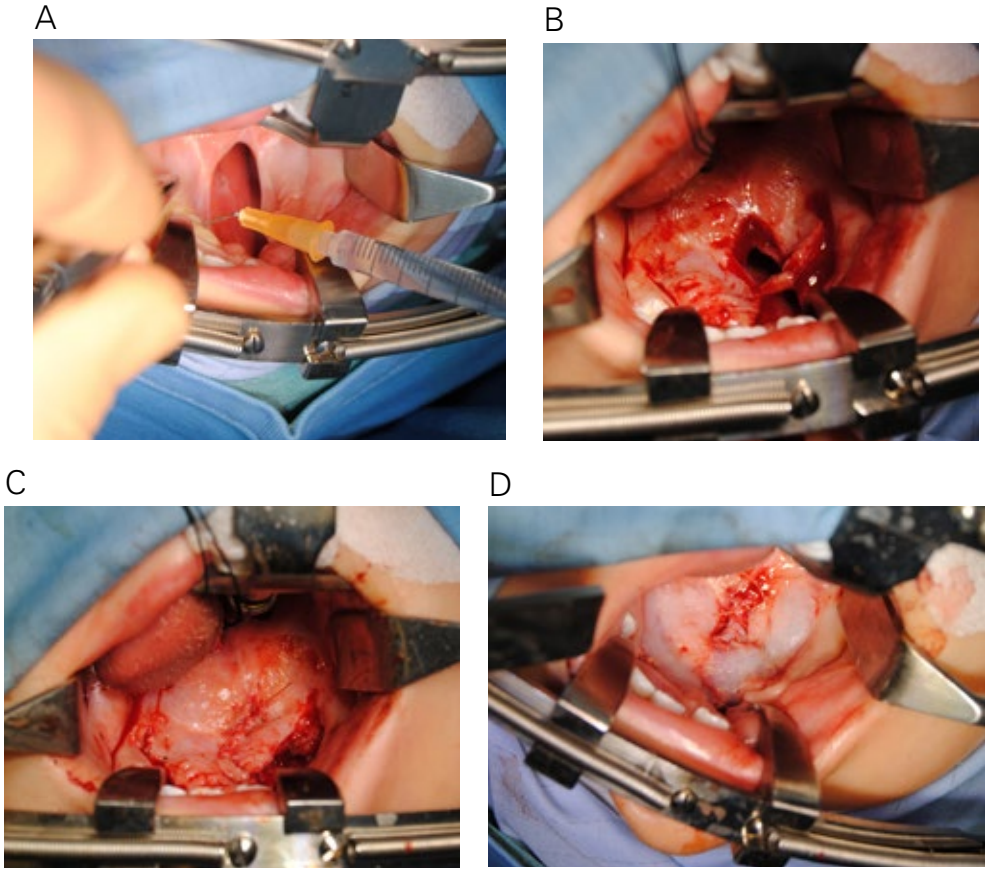


Figure 3.19 The second stage palate repair according to Widmayer Perko.
(A) Preparation with infiltration of Mepivacaine adrelinate. (B) Creation of the Two Flaps. (C) Completion of the closure. (D) Final appearance after application of fibrin glue in the areas that remained bloody.

The Furlow Z-Plasty palate repair

The Furlow technique is widely used to repair cleft of the soft palate, both in cases where there is an isolated cleft of the secondary palate and in the eventuality of a unilateral total cleft palate in which the above technique is used together with techniques for the reconstruction of the mucosa of the hard palate.

The Furlow technique is based on opposing Z-plasty (2 flaps on the oral mucosa and 2 on the nasal mucosa) with the peculiarity that the Z-plasty has the property of stretching in one direction and shorten in the other so lengthening the soft palate and narrowing the velo-pharyngeal gap, thus functioning as both palatoplasty and pharyngoplasty *(Sitzman TJ, Marcus JR, 2014)*.

As a starting point in the application of the Furlow technique it is necessary to identify the incision lines.

Two flaps with anterior base and 2 with posterior base are prepared. By convention, the oral muscle-mucous flap posterior based is drawn on the left side of the cleft. It is necessary also creates lateral releasing incisions which extend from the maxillary tuberosity posteriorly until the retromolar trigone region. In the case of complete cleft palate the incision lines also include the hamulus, the junction between the hard palate and gingiva, the medial edge of the cleft, and the uvula.

The incision to prepare the left oral triangular flap posterior based starts at a 60° angle to the medial margin of the omolateral cleft, from the hamulus laterally to the junction between soft and hard palate medially. The incision for the right oral triangular flap anterior based begins from the uvula medially, laterally to the hamulus with a 60°-90° angle respect to the medial border of the homolateral cleft. Between these lines and the lateral release incisions a mucosal bridge must be left.

The uvula medial edges are demucosated.

After the incision of the oral flaps the abnormal attachment of elevator of the left palate at the distal edge of the hard palate, at tensor palate aponeurosis and at the pharyngeal constrictor fibres is interupted. The apex of the homolateral flap is lifted with a traction suture to proceed posteriorly with dissection and separation of the nasal plane up to the uvula. Similarly the right corner is raised and dissected while leaving in the layer below the palatopharyngeus muscle and palatoglossus occupied by the posterior part of the velum, from the rear vertex to the anterior junction between the soft and hard palate.

In the case of cleft which includes the hard palate, through the lateral releasing incisions and using a periosteal elevator you can proceed medially with the detachment of mucoperiosteal flaps from the hard palate. The same is done to detach

the nasal mucosa and the periosteum from the nasal surface of the hard palate. The dissection is extended around the hamulus and the vascular pedicle to allow a circumferential mobilization of the flaps, and then a medial suture without tension.

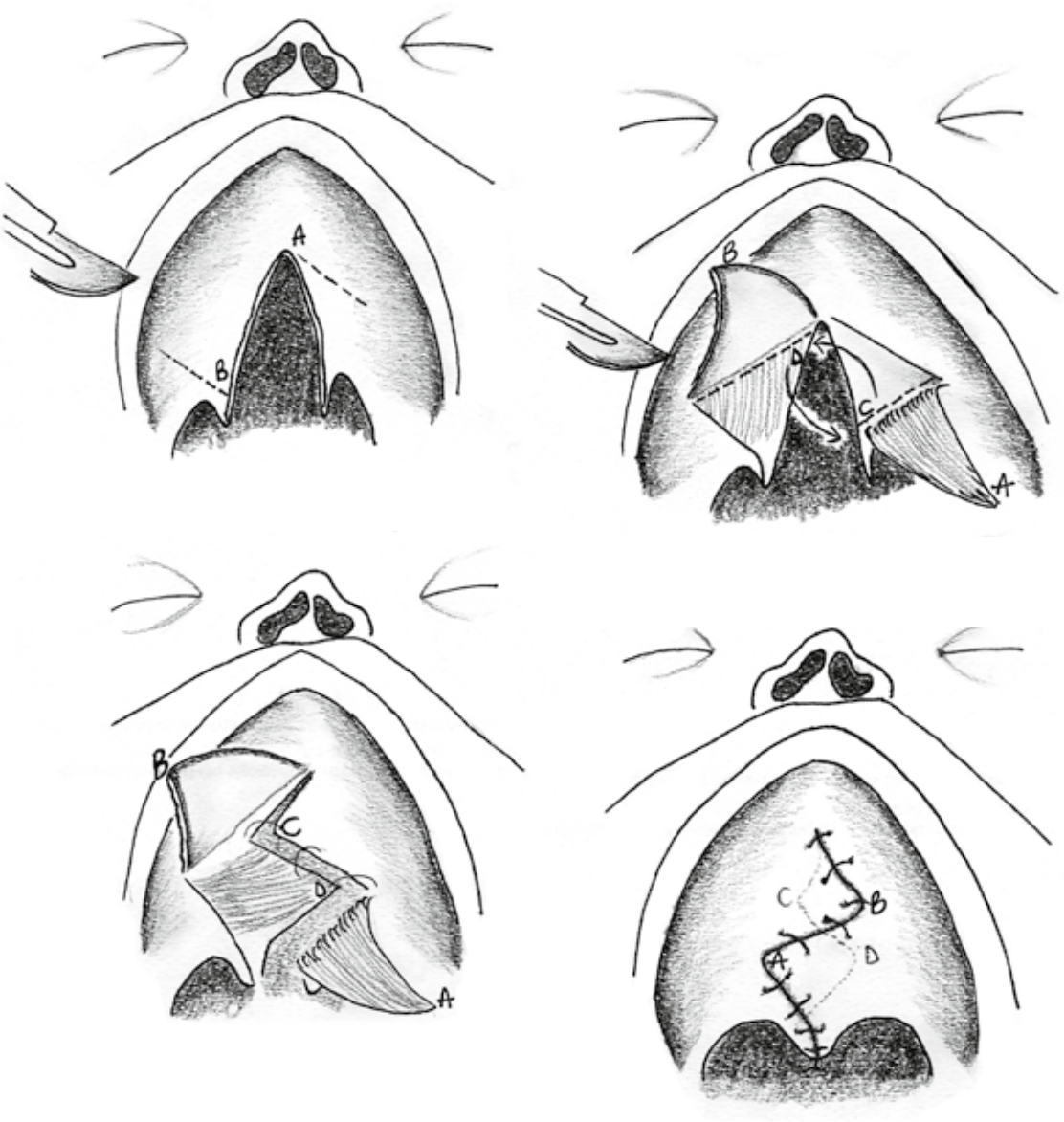


Figure 3.20 Furlow palate repair.

The nasal triangular flaps are then prepared: the incision of the left, with anterior base, starts medially from the base of the uvula and continues laterally up to where the elevator of the velum protrudes from the skull base in a specular manner to the homolateral oral flap. The creation of mucosa-muscle right flap will require the release of the attachement of the muscle at the posterior border of the hard palate, to the tensor palate aponeurosis and the superior constrictor of the pharynx laterally. The incision of the flap will start medially to the detached summit of the muscle and will continue posterolaterally up to the hamulus.

Sutures begin with the uvula combining both the nasal reconstruction plane and the oral one; right nasal posterior base flap is juxtaposed on the midline and placed to occupy the area vacated by the lifting of controlateral flap whose apex is instead set at the corner of the right nasal flap and posterior edge of the hard palate. In this way the fibres acquire horizontal and transverse movement. The sutures are placed so as to cover only the mucosal layer. In case of complete cleft the suture continues forward to close the nasal defect of the hard palate possibly with the help of flaps of the vomer.

Regarding the oral layer, the posterior based mucosa-muscle left flap is fixed contralaterally to the corner created by the lifting of the flap that is fixed to the left anteriorly to the latter completing the closure.

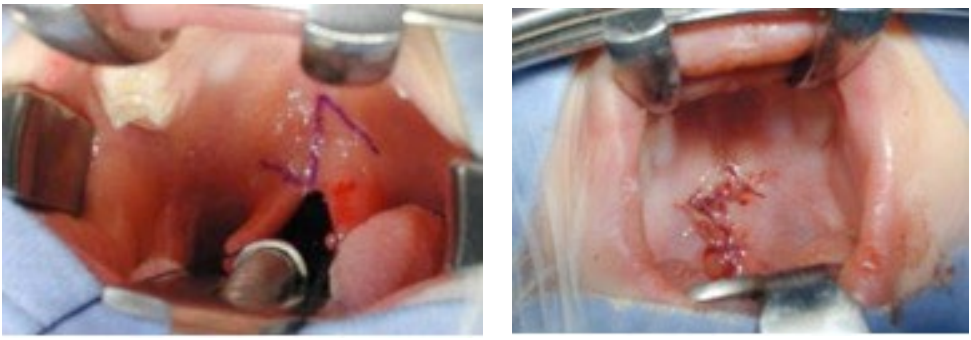


Figure 3.21 Cleft palate repair according to the Furlow technique.

3.5 Bone grafting of alveolar cleft

Alveolar cleft is defined as the maxillary bone discontinuity that occurs anteriorly the incisive foramen, it requires specific strategies that are to be included in the primary treatment of the malformation. Leaving a bone defect at this level would

force the patient to limited possibilities of orthodontic treatment and to live with instability of the maxillary segments.

Surgical options are:

- Early bone graft, during the first year of life: many doubts have been cast upon the advisability of performing this operation;
- The free grafting of tibial periosteum (following Stricker), carried out simultaneously with the Labioplasty. This technique is now universally abandoned as this type of approach leads to a negative interference with the subsequent growth of the facial mass;
- Gengivoperiostioplasty at the same time as labioplasty, that was introduced for the first time by Skoog (1965). It consists in the closure in a double layer of periosteal flaps between the alveolar defect and is based on the principle that this operation induces bone growth without the need of a bone graft. This technique is facilitated by association with nasoalveolar moulding or systems such as the Latham device as Millard himself has proposed, which bring together the alveolar segments. The choice of this approach remains controversial since so far, clinical studies show gengivoperiostioplasty has an early negative effect on the subsequent development of the maxilla *(Berkowitz S, 2013);*
- Late bone graft, in the phase of the mixed dentition, accepted as the most effective method to avoid the problems mentioned above and interferes less with the growth of the maxilla; It was introduced by the group in Oslo (Boyne and Sands) in 1977.

The latter technique provides a wide exposure of the defect by cutting along the edges of the cleft and the gum line that is extended posteriorly to the first permanent molar and anteriorly to the central incisor of the affected side of the cleft to allow the cover of the graft. On the palatine side 2 mucoperiosteal flaps are lifted from the margins of the defect.

Several donor sites have been proposed such as the ribs, tibia and scalp. However, the most widely used is the anterior iliac crest, from which fragments of cancellous bone are taken, leaving the cortex intact. The cavity left in the extraction location is then filled with hemostatic felt (collagen). The cancellous bone choice is dictated by the fact that, inside the latter, the osteogenic cells survive and, if placed in good blood supply conditions, begin to produce new bone tissue much faster than they would if they were of cortical bone (in a few days).

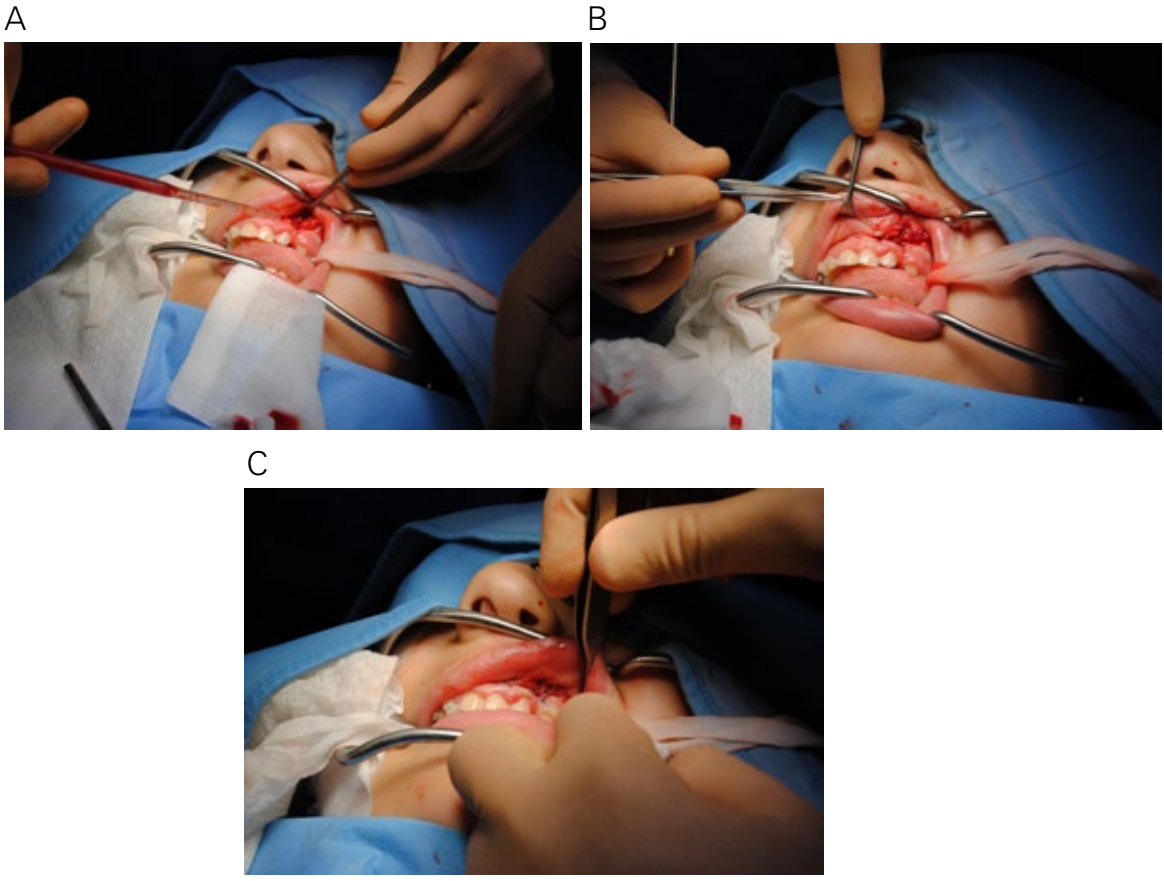


Figure 3.22 Bone graft. (A) Filling of the defect with cancellous bone fragments. (B) and (C) Closing the flaps.

The entire defect is filled with the grafted fragments especially at the base of the alar cartilage to improve the symmetry, the previously prepared flaps are then advanced and sutured over the graft. In case of bilateral cleft both sides are inserted simultaneously.

The ideal bone graft timing is calculated considering two factors in particular: the growth curve of the various segments of the maxilla and the clinical objective of the graft itself. The sagittal and transverse jaw growth ends around 8-9 years, while the growth of the vertical dimension essentially depends on the deposition of alveolar bone stimulated by the continued eruption of teeth. The graft is transformed into functional alveolar bone, if this occurs before the eruption of the canine ipsilateral to the defect (8-11 years). The eruption of the canine, sponta-

neous or guided by orthodontic devices, can be used as an incentive to maintain adequate bone height. Performing the surgery when the cervical portion of the canine is still covered by bone protects it from possible iatrogenic injuries that appear to be one of the contributory causes of the phenomenon of dental root resorption.

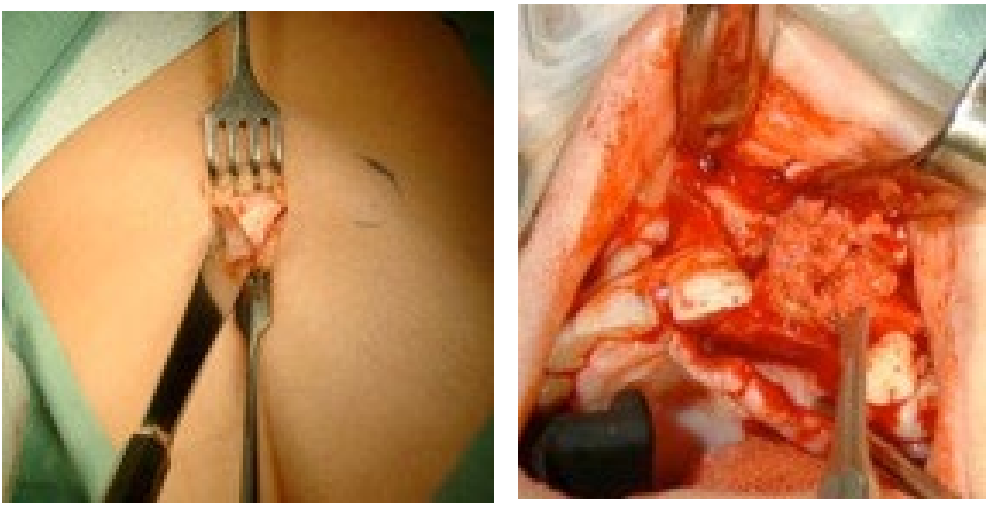


Figure 3.23 Bone graft from iliac crest.

Finally, a primary goal of the bone graft is to promote the orthodontic closure of the cleft and this fact seems to be facilitated by the practice of grafting before the eruption of the canine.

Some authors have proposed an earlier intervention, between 5 and 6 years of age to facilitate migration within the graft and also the eruption of lateral incisor that is often lacking in these patients or are malrotated and need to be removed. Whilst a very interesting subject more clinical trials are needed to prove that a graft at a younger age does not interfere with jaw growth *(Berkowitz S, 2013)*.

4.
The orthodontic treatment and maxillary surgery

Regardless of the techniques used for the primary correction of the cleft and the experience of the surgeon, there remains a certain proportion of patients who develop an abnormal pattern of maxillary growth, even if all the patients are operated by the same surgeon and with the same protocol, reflecting individual and inherent differences in the growth of the facial mass.

Maxillary growth failure is generally three-dimensional (length, width and height) and retrusion is more noticeable in patients with a unilateral defect as those with bilateral cleft are benefited from the initial protrusion of the premaxilla that decreases gradually over time by acquiring a position nearly perfect in their late teens.

Patients with maxillary hypoplasia usually have some signs represented by anterior and posterior crossbites and a generally concave profile.

A suitable relationship between the incisors in the deciduous dentition phase is decisive for the development to adjust the front portion of the jaw or that form and function are dependent on each other. For this reason orthodontic treatment should accompany the patient with cleft lip and palate from an early age as part of a multidisciplinary protocol, typically alternating phases of active correction with maintenance phases and stabilization.

The approaches can be different in different centres, but typically include:

- Presurgical orthopaedics;
- Treatment in deciduous dentition;
- Treatment in mixed dentition;
- Treatment in permanent dentition.

1. Pre-surgical orthopedics (0-3 months)

Historically, attempts to correct cleft using non-surgical techniques began in 1500. The beginning of the era of modern orthodontic appliances can be considered 1954 with McNeal, who published a work on neonatal manipulation of

maxillary segments in complete unilateral and bilateral cleft lip and palate. Initially these attempts were not very successful due to the belief that they could not help much in avoiding surgical correction. Today it is common opinion that this aim is impossible. There is agreement that this type of device in some way hampers growth of the jaw. The debate is still open between those who support the surgical correction and those who support a combination with a pre-surgical treatment. The arguments justifying this opinion are the lower costs, the greater precision, the additional action on the nasal cartilages and the absence of scarring compared to traditional lip adhesion surgery *(Grayson BH, Santiago PE, 1999).*

The rationale behind the use of devices in the first months of life (generally restricted to selected cases) is to restore the correct relation between the soft tissues, cartilage and bone as during the first months of life the tissues are highly plastic. This should facilitate the subsequent surgical operation and reduce the need for further surgery after the primary correction.

The therapy, also known as early orthopaedic treatment, may be divided in two categories: unilateral cleft lip and palate patients, and bilateral cleft lip and palate patients. The basis of the orthopaedic treatment in these patients are very different as well as the biological and scientific foundations. Many types of maxillary orthopaedic devices can be used, usually considered in three main categories: active, semiactive and passive appliances. Active appliances move the maxillary segment for bringing together the alveolar processes. Semiactive appliances reorient the maxillary segments in a correct position. Passive appliances have different functions, separate the nasal and the oral cavity, allow normal feeding, help breathing and guide the alveolar growth.

The treatment ranges from simple taping (the attachment of patches to bridge between the two segments of the lips) to more complex systems that require the intervention of an orthodontist such as presurgical nasoalveolar molding (PNAM). This is aimed at the active remodelling of nasal cartilage and alveolar processes and elongation of the columella. Different models exist for different conditions depending on whether it is mono or bi-lateral cleft. Generally they consist of a oral plaque that is created from an impression taken in the first days of life that then is changed from week to week in order to bring the bone segments together. A nasal stent is joined to the plaque later when the alveolar defect is already partly reduced. This has the purpose of restoring an adequate arcuate shape to the nasal cartilages which are often crushed and the position of which is modified weekly by means of elastic straps fixed with patches to the cheeks of the child.

Naso Alveolar Molding (NAM) is a protocol introduced in late 1990's using a specific plate to remodel both the nose and the alveolus. The NAM appliance

allows the retraction of the premaxilla and the lengthening of the columella at the same time for the presence of two nasal stents supporting the columella.

Patients with bilateral cleft lip and palate present a severe protrusion of the premaxilla and different techniques have been reported for the retraction of the premaxilla. Presurgical orthodontic treatment is often used to obtain a retraction of the pre-maxilla in bilateral cleft and allow the correction of the defect in a single period of the lip defect. One of the devices used is the one designed by Georgiade and then popularized by Latham and Millard allowing simultaneous retraction of the pre-maxilla and the expansion of the maxillary processes *(Mulliken JB, 2001)*.

In our centre lip taping of prominent premaxilla have proven to reduce the prominence enough to allow a good surgical outcome and we find this easy for parents to manage. It is important, if the premaxilla presents asymmetrically, to correct this bringing the premaxilla to the midline with taping to one side only and after centralizing obtain a symmetric retraction. It costs little in time and supplies, is generally comfortable and does not require any restraints.

Complications related to these systems may be ischemia and consequent ulceration of the soft tissues if the changes are implemented too quickly. Excessive, premature exposure of the deciduous incisors through the gums in the pressure points of the plaque may also occur. Benefit obtained may be lost when parents do not cooperate in maintaining the devices or apply them incorrectly.

2. Treatment in deciduous dentition (before 6 years)

Interceptive therapy before alveolar bone graft

This stage concerns only those cases characterized precisely by anterior and posterior crossbite and retrusion of the facial skeleton (about 20%) and is intended to allow spontaneous eruption and the correct position of the permanent incisors. It involves the use of devices aimed at transverse expansion of the palate and to the protrusion of the jaw (facial mask) for a total period of about 15 months (3 for the first and 12 for the second), followed by the application of the palatal arch fixing apparatus. But orthodontics in the deciduous dentition should be avoided, and the indications for early palate expansion and protraction facemask are extremely rare.

3. Treatment in mixed dentition

Usually the orthodontic treatment starts at this time and includes the palate expansion for managing of the space, alignment of the teeth and preparation for bone grafting. Indications for maxillary orthopaedic protraction could be considered at this age with the use of facial masks, even if the long term effects of these devices are not clear.

4. Treatment in permanent dentition

The objectives in this case are the same as in patients not suffering from cleft and are obtained through the same orthodontic appliances with special attention to the setting of the previously expanded palate to prevent the narrowing *(Berkowitz S, 2013)*. At this stage the orthodontic treatment could be definitive by obtaining a good final occlusion or could be addressed as a preparation for the definitive surgical treatment.

In the case of patients suffering from major discrepancies of the maxillo- facial bones (Class III malocclusion) the surgical advancement of the maxilla should be considered. To do so it is normally expected that the growth of the jaw is almost complete: 14-15 years for females, 16-17 years for males.

Two main end-stage surgical procedures are considered:

1. Traditional Le Fort I Maxillary osteotomy. The jaw is acceded through the buccal groove, and, after the osteotomy and the maxillary advancement a bone graft in the pterygo-maxillary area should be placed if the advancement is greater than 6 mm. The use of rigid internal fixation and bone graft has reduced the instability of this procedure.

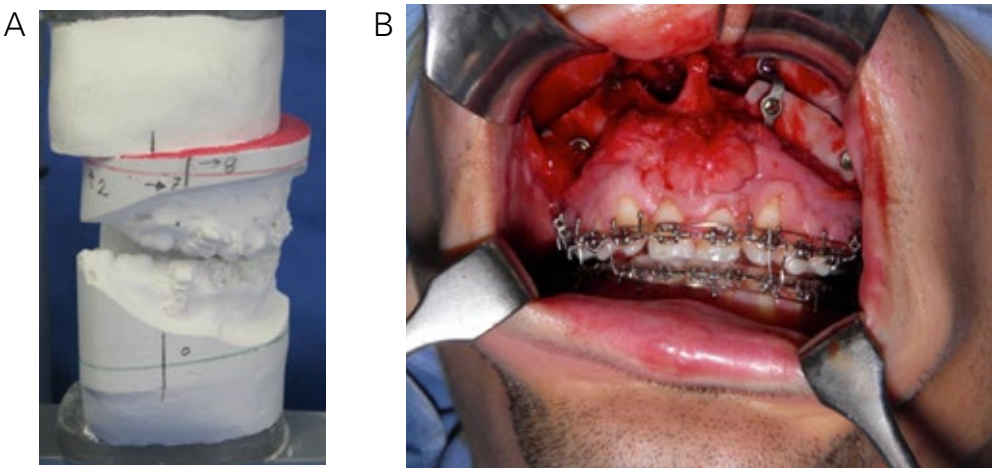


Figure 4.1 Le Fort I schematic. (A) Pre-surgical project. (B) Operation.

2. Distraction osteogenesis. Patients with severe maxillary hypoplasia involving all planes (vertical, horizontal and transverse) and Class III malocclusion that would require an advancement up to 10 mm are considered for this procedure.

After a quarter of century of extensive use, Distraction Osteogenesis now has specific indications for congenital craniofacial and cleft deformities.

Technology has evolved from the first application of external devices to intraoral and hybrid or semiburied techniques. The surgery for all patients is a classic LeFort I osteotomy without repositioning of the fragments or bone grafts. After the vestibular incision and the osteotomy an external or internal device is fixed for the gradual elongation of the maxilla. About five days after surgery the activation phase starts at a rate of 1 mm per day. The principle of the external distraction is to use the skull as the anchor point for the advancement and the stabilization of the maxilla. After the activation period the external device (mask) will be kept in place for 2-3 weeks for activation phase and after obtaining the desired advancement, for a period of 2-3 months for consolidation.

Literature concerning the outcomes traditional osteotomy with advancement of the segments and internal fixation with plates refer that mean values of the jaw advancement is between 5 and 7 mm with a variable degree of recurrence of retrognathia long-term between 20 and 25%. Most favourable are data regarding the use of external distraction, with mean advancing around 11-12 mm and less or no recurrence. The advantages of the second approach are more versatility, the amount and direction of advancement that can be modified during the course of use, and the absence of restrictions on the age indicated to proceed.

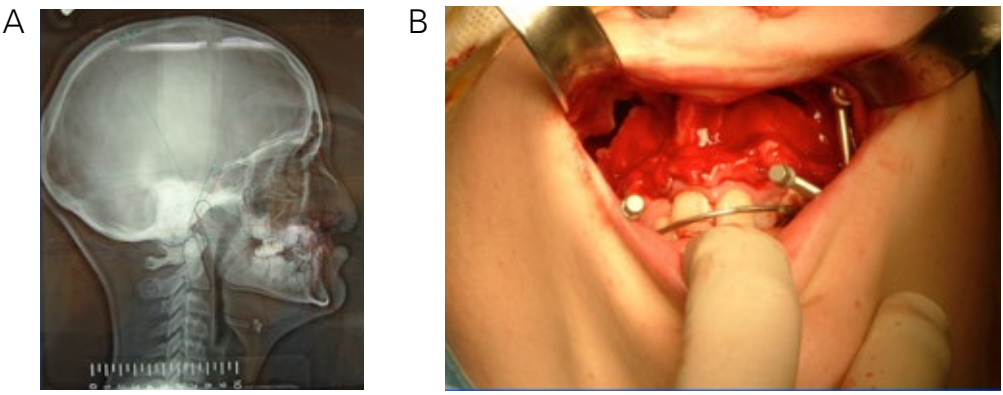


Figure 4.2 Distraction osteogenesis. (A) Pre-surgical radiograph. (B) Operation.

Patients stimulated by the distraction device produce autogenous bone without the need to reposition the segments after osteotomy, nor to insert other means of internal fixation or autologous grafts, with less interference with the facial mass growth. It is sufficient that teething, primary or secondary, is complete, in order to allow the fixing of the device by means of the intraoral bridge. No additional surgical procedures for the removal of the external system are required *(Berkowitz S, 2013).*

5. Secondary surgical treatment

5.1 Cleft lip deformity

A wide variety of deformities can occur after primary repair of the cleft. Scarring and the loss of tissue layers in general make the secondary procedure more difficult than the first operation. The deformity can involve skin, muscle, oral mucosa and the nasal elements.

It is also useful to know which technique was used during the primary repair, for example a short lip more frequently occurs after a Millard repair, a long lip is more commonly associated with a Tennison procedure.

The evaluation of upper lip must include the examination of the adequacy of the buccal vestibule, of the vermillion mucosa, of the Cupid's bow and philtral structures and the location and direction of scars. It is also important to check the alignment of circumoral muscles. Bulging on either side of the scar when the patient purses his or her lips is evidence of orbicolaris muscle discontinuity.

Timing for correction of secondary deformities is still the object of debate, it is important for this kind of surgery that the patient can participate in the choice of whether and how to proceed *(Marsh)*.

Secondary repair occurs most frequently either prior to starting school normally before 5 years of age or during adolescence.

Scar deformities

If a disfiguring scar is the only deformity standard surgical principles of scar revision can be applied. Excision with primary closure along the philtrum columns (with the disadvantage that discarded tissue is useless in case of future surgical procedures), a Z-plasty or diamond shape excision can be considered if some length is needed.

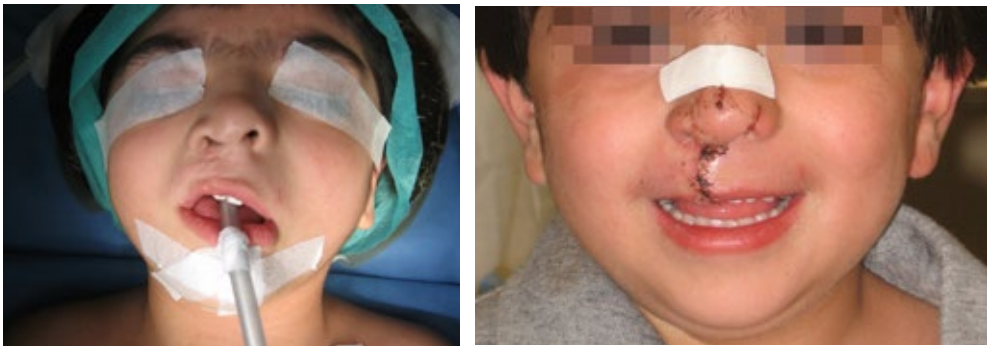


Figure 5.1 Re-do labioplasty in a child with clefl lip and nose deformity.

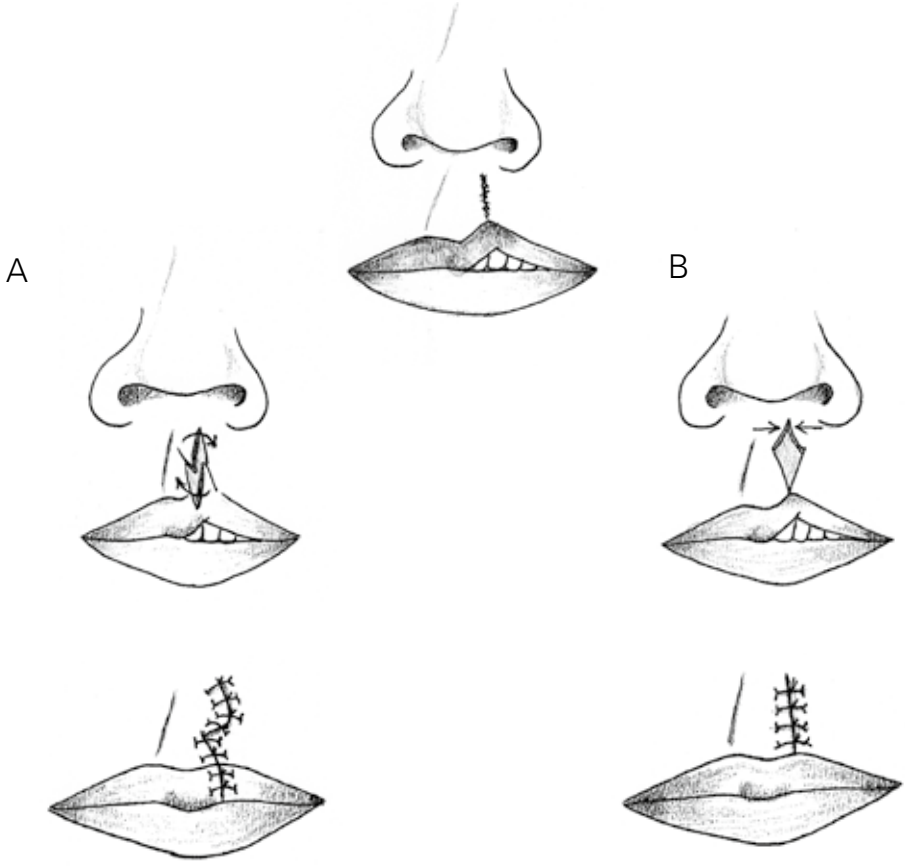


Figure 5.2 Scar lip deformity. (A) Correction with Z-plasty. (B) Correction with diamond shape excision.

Cupid's bow and vermillion deformities

Anomalies of Cupid's bow are the most common deformity observed after cleft lip surgery. A simply vertical deficiency of vermillion lateral to the cleft can be corrected by excision of skin and advancement of mucosa or a Z-plasty.

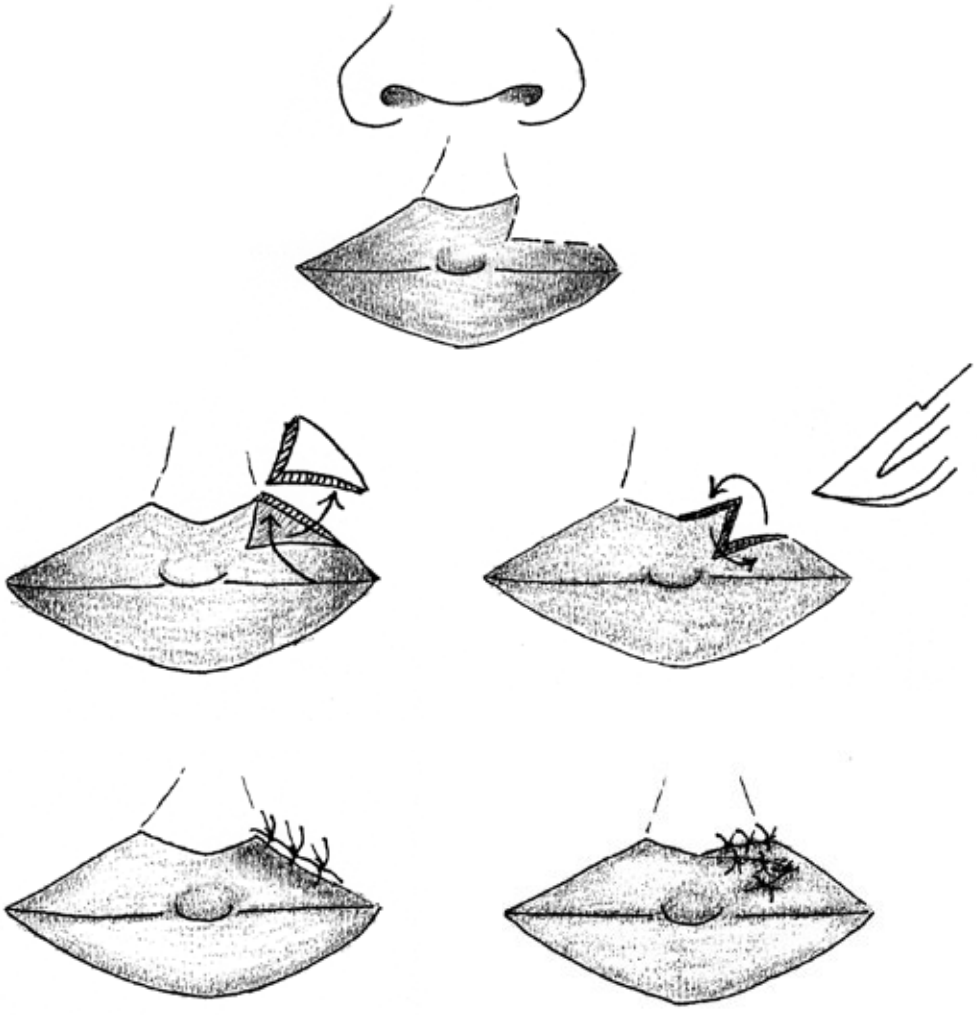


Figure 5.3 Correction of a Cupid's bow deformity with excision and Z-plasty.

When a "notching" defect is associated it can be corrected by a V-Y advancement or a Z-plasty to lengthening the vertical dimension of the vestibular mucosa.

If the Cupid's bow is totally absent it can be recreated by excising the skin bilaterally to the philtrum simulating the arches of the Cupid's bow, cephalic advancing of the mucosa and closing with half-buried mattress sutures. A small modified Abbé flap should also be considered *(see later)*.

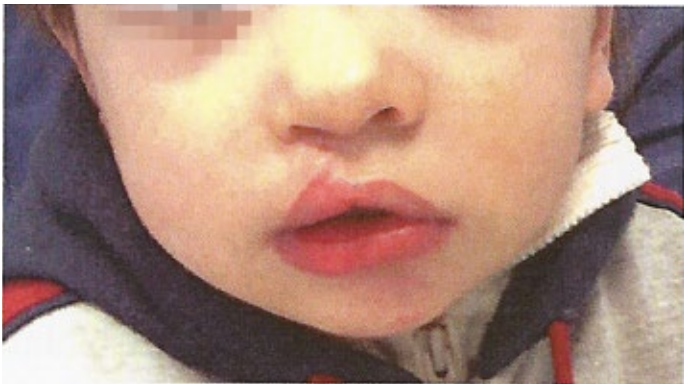


Figure 5.4 Patient with post-labioplasty Cupido's bow deformity.

Anomalies in the length of the upper lip

A long lip was a common finding after Tennison repair, but it is a rare complication today. A short upper lip is more commonly encountered after an insufficient Millard rotation-advancement. In this case the defect can be improved by a re-rotation-advancement procedure with the advantage that simultaneous muscle realignment and bone grafting of the maxilla can be performed.

A vertical midline deficiency can be approached with perialar incisions closed with a V to Y plasty.

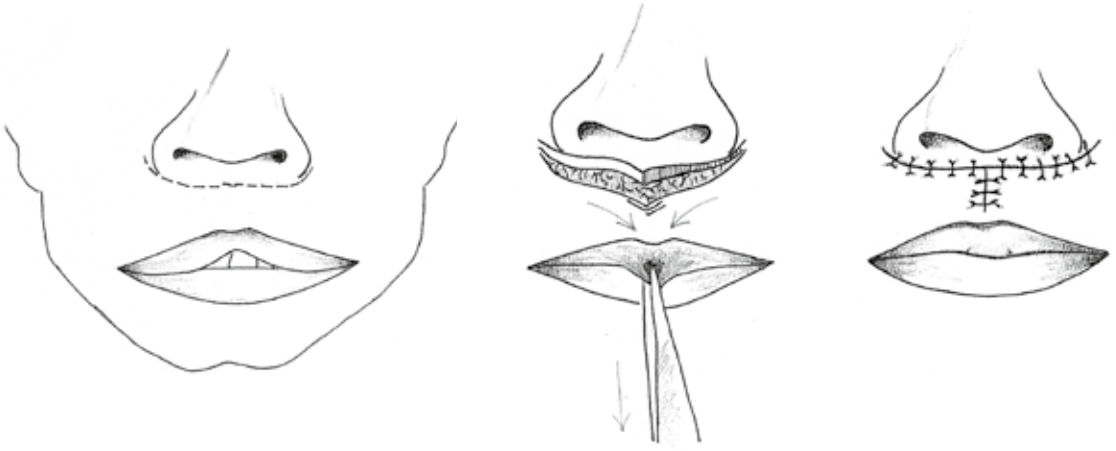


Figure 5.5 Correction of a vertical midline deficiency with perialar incisions and V to Y plasty.

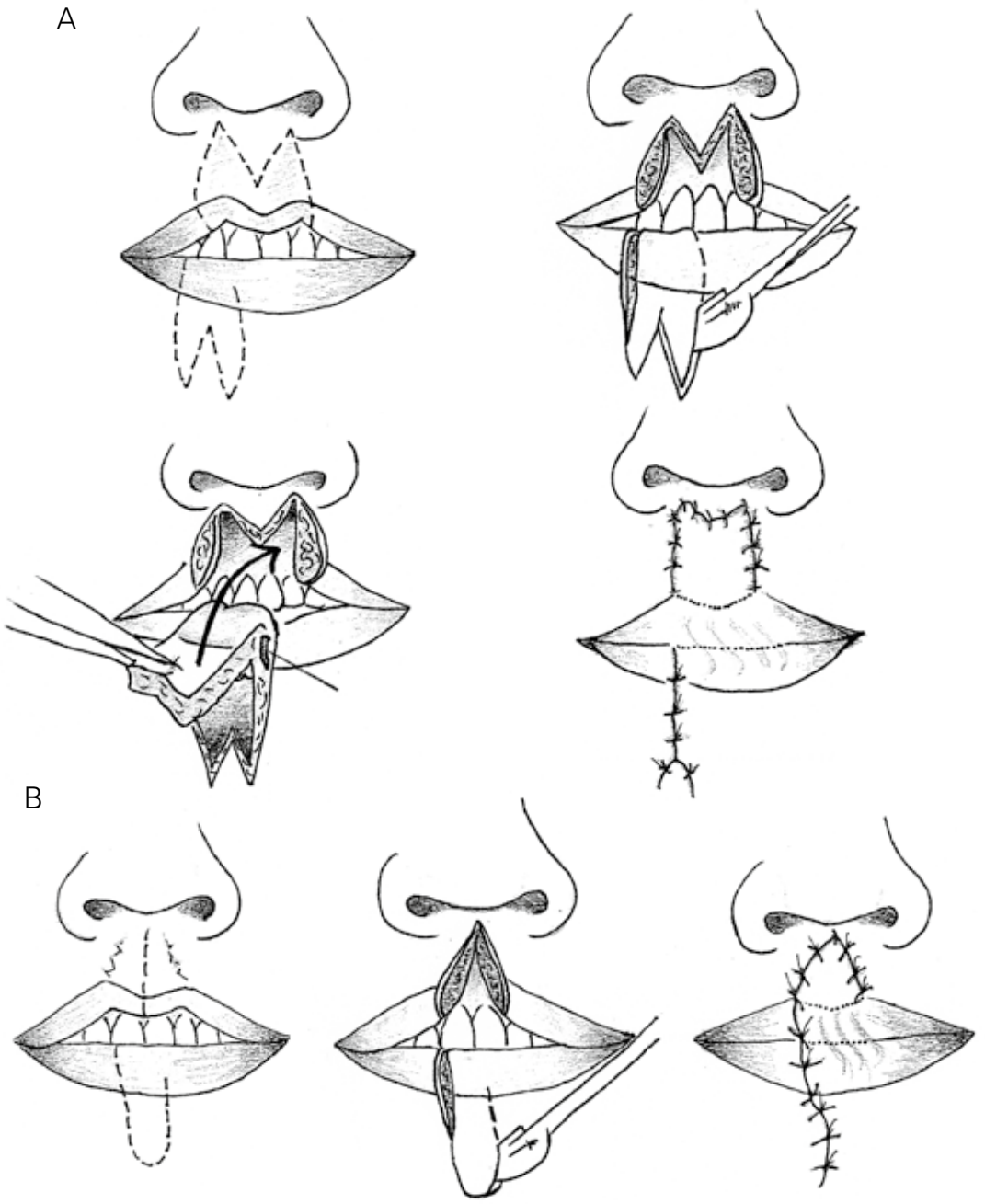


Figure 5.6 Abbe's flap schematic sequence. (A) With M-shaped donor flap. (B) With V-shaped donor flap.

Deficiency of the horizontal lip or tight lip can occur after excessive resection of normal tissue or when closing an extremely wide defect during primary repair. This defect can be corrected by use of an Abbé lip-switch flap with several modifications described by surgeons like Cannon, Jackson, Hogan and Converse. The procedure is done during or after school age because it requires patient cooperation.

The general principles are:

- Careful evaluation of size of the deficit;
- V or M donor flap from lower lip including vermillion, skin and muscle;
- Central placement of the Flap to ideally recreate the philtral portion of upper lip;
- Division of the flap after 2 weeks.

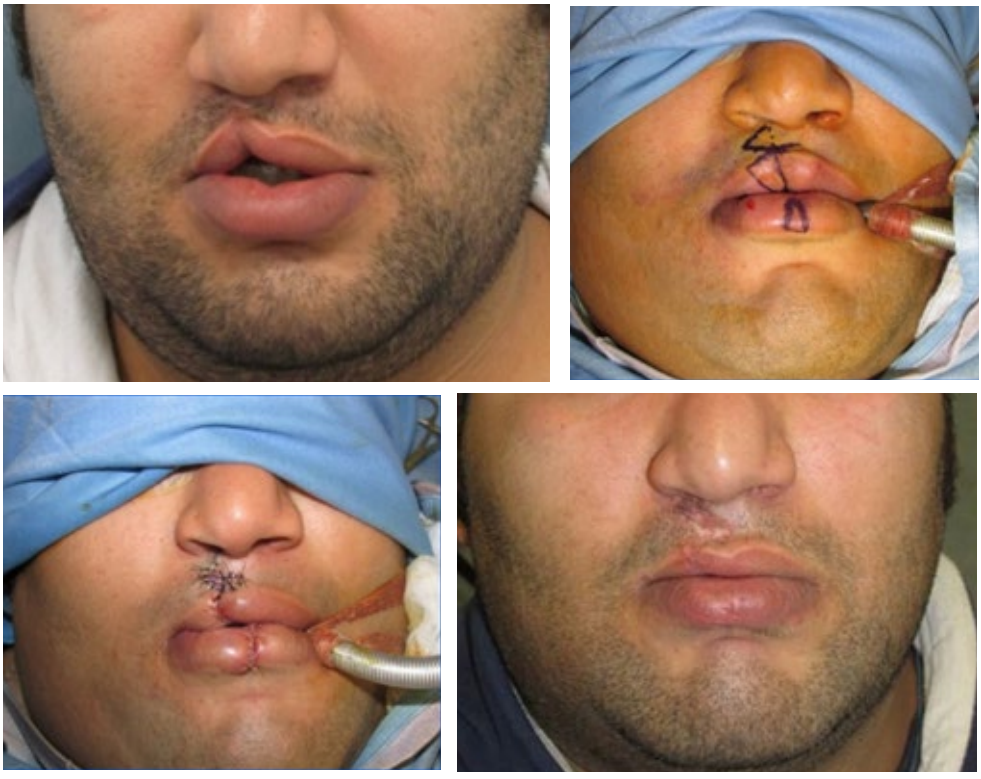


Figure 5.7 Abbe's flap to correct a notch defect.

The three following deformities are typical of bilateral clefts.

Absent philtral ridges

A variety of techniques have been proposed.

O'Connor and McGregor: elevation of prolabial skin, development of subcutaneous flaps based inferiorly and rotated laterally and inferiorly at vermillion border and closure of the skin over the central concavity.

Millard's modification employs medially based de-epithelialized flaps from just above the vermillion ridge of the upper lip, turned medially and upwards and buried in a tunnel to simulate the philtral columns.

Neuner and Schmid: skin and cartilage free graft taken from the ear and positioned into the central lip.

Abbé's flap and modifications of this procedure can create a new philtrum as previously described.

Whistling deformity

The "whistling lip deformity" or central notching defect is the most common vermilion deformity of bilateral cleft. Correction can be achieved by a double V-Y advancement or by a larger V-Y advancement with releasing of the entire superior labial sulcus and closure with cephalical advancement.

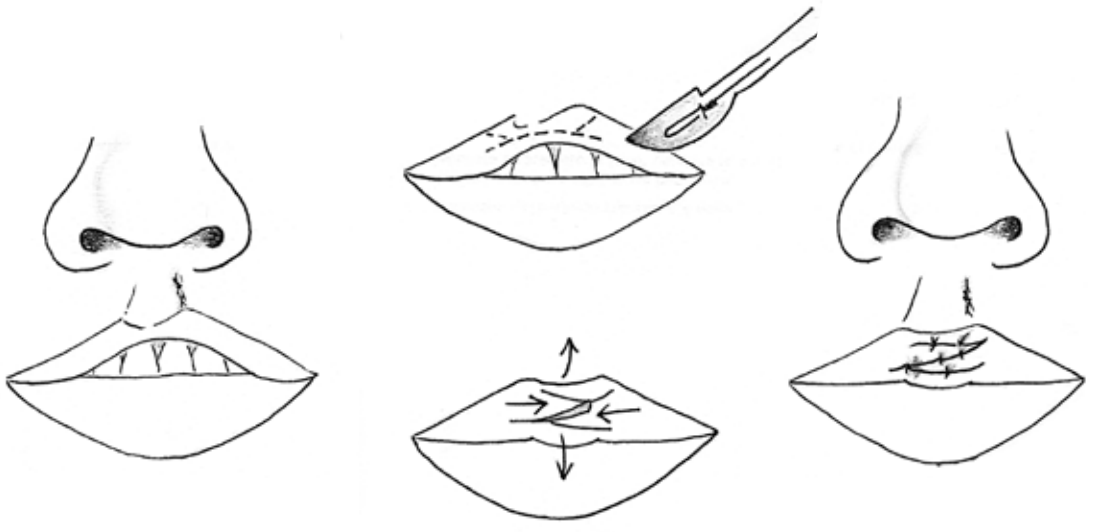


Figure 5.8 Correction of whistling lip with double V-Y advancement.

Other techniques are creation of pyramidal lateral vermilion flap including orbicular muscle to fill the central lack of tissue, that are rotated medially as a pendulum to meet in the prolabium or use of a free composite graft from the central lower lip (described by Flanagin) or of a Abbé flap as previously described.

Columella deformity

There is no consensus concerning timing for secondary correction and the best source of tissue to employ for columella lenghtening.

Forked flaps, originally described by Trauner, have been used effectively to lengthen the columella with upper lip tissue obtaining a narrowed philtrum with acceptable scars: superiorly based flaps were rotated bilaterally and insert transversely into the base, augmenting the columella.

5.2 Complications after cleft palate repair

Oronasal fistula

The oronasal fistula is the most common complication after closure of the hard palate.

It is typically evident a few weeks after the primary surgery. If it is small it can be treated conservatively and some cases can close spontaneously. In other cases, they become apparent with major clinical manifestations such as nasal escape with distortion of speech, regurgitation of food both liquids and solids from the nose.

The incidence is very variable depending on the cases from 2% to 70%, also due to the lack of a standardized classification system. One of the usable schemes is that of Pittsburg, described by Losee and Colleagues that divides palatine fistulas into 7 subtypes:

- Type I, at the level of the uvula or that looks like uvula bifida;
- Type II, in the soft palate;
- Type III, at the junction between the soft palate and hard palate;
- Type IV, in the hard palate;
- Type V, to the incisive foramen level / junction between the primary and secondary palate;
- Type VI, on the lingual aspect of the alveolar process;
- Type VII, on the labial side of the alveole.

A further distinction is that between functional and non-functional depending on the presence or absence of symptoms. Even the smallest fistulas may cause functional problems.

Typically the closure can be performed in the course of secondary surgery of the lip or nose. If the food spill is considerable and makes the patient uncomfortable or if the fistula is likely to interfere with language, the closure must be performed in the preschool years.

Surgical closure of oronasal fistula should be attempted at least six months after the previous surgery.

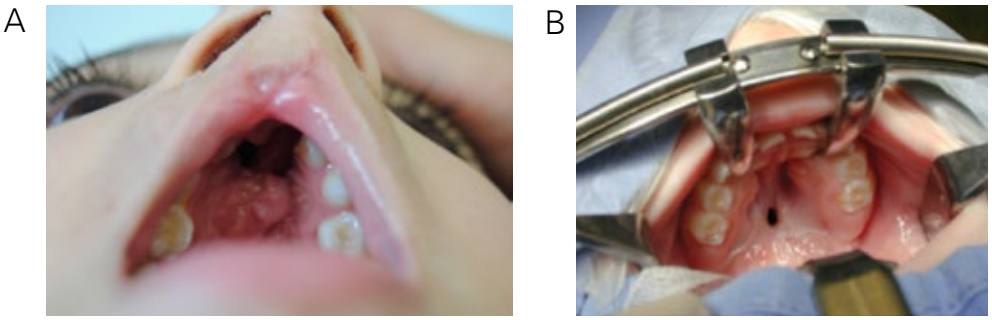


Figure 5.9 (A) Large functional anterior fistula. (B) Small fistula of soft palate.

The way of treating fistulas poses problematic aspects as the percentage of recurrences after a first correction is estimated at 65%. Over the years there have been numerous approaches proposed, from the application of closing devices (Berkman), to the many flaps proposed to close the wide anterior fistulas, particularly lingual (Guerrero-Santos and Altamirano in the 1960s and 1970s, still used in some centres) and mucosa-muscle flaps using facial artery irrigation, especially to free cartilaginous grafts from the ear.

The fistula in the soft palate requires surgery to close it, even if the small local flaps have the disadvantage of being packaged with scar tissue.

Fistulas at the soft palate level may simply be excised and repaired by direct closure in double layer or by performing a palatoplasty elongation (Furlow) if associated with velo-pharyngeal insufficiency. Fistulas of the hard palate pose major difficulties and often the preparation of local transposition flaps is not enough and it becomes necessary to complete with mono or bi-pedicled flaps. The nasal floor is often more difficult to close due to the brittle nature of the tissue constantly exposed to food reflux. Proposals to facilitate the closure have been the creation

of septal flaps, mucus-buccal flaps or the application of an acellular dermal matrix (Kirschner).

It is not possible however to standardize the treatment of this complication, there is not a technique applicable to all cases and the best treatment remains prevention through a primary palatoplasty as much as possible without tension *(Sitzman TJ, Marcus JR, 2014).*

5.3 Velo-pharyngeal insufficiency

The pharynx velum can be considered a dynamic anatomic sphincter composed of the soft palate, the pharyngeal side and rear walls, the closing of which is given by a posterior-superior movement of the velum and medial pharyngeal wall. It is necessary to completely separate the nasal cavity from the oro-pharynx and thus allow an increase in the intraoral pressure necessary for the production of some consonant sounds that are not nasal.

Many factors can be responsible for a dysfunction of this sphincter such as a velum and a hard palate that are too short, a dysfunction of the pharyngeal walls or a paralysis that makes these elements unable to move. For this reason, even if initially terms such as inadequate, incompetent and dysfunction of the soft palate were used indifferently, it is more correct to distinguish between causes related to abnormal anatomy (velo-pharyngeal insufficiency), and altered neurophysiology (velo-pharyngeal incompetence), or articulator errors (erroneous velo-pharyngeal learning or functional dysfunction) *(Berkowitz S, 2013).*

Among the causes of velo-pharyngeal insufficiency cleft palate is definitely at the top, but there are many others such as:

- Submucous cleft palate;
- Deep pharynx, in patients with normal anatomy of the soft palate, but who have cervical abnormalities or of the base of the skull (as in Klippel-Feil syndrome and Down's syndrome);
- Adenoidal atrophy / adenoidectomy, since the nose-pharynx posteriorly lacks the thickness given by adenoid tissue;
- Hypertrophic tonsils;
- Tonsillectomy, linked in this case to the scars on the rear tonsillar pillars which can alter the movements of the lateral walls of the pharynx;
- Maxillary advancement, generally more than causing the insufficiency it accentuates it in patients who already were affected.

Incorrect velo-pharyngeal learning is instead linked to an incorrect verbal articulation that is shown by the inappropriate opening of the velo-pharyngeal valve during verbal production attempts. Examples:

- **Faulty articulation** - during normal joint development some children learn to produce some sounds incorrectly. This causes a nasal resonance phoneme-specific or a nasal air leakage. The most common condition is the substitution of fricatives (f and s) with a hissing sound or the nasalization of the vowel I.
- **Compensatory Verbal Production** - when there is a short velum during development the child learns to compensate for the lack of intraoral pressure producing extra-articulatory sounds at the level of the glottis or the pharynx. When the anatomical defect is corrected this articular pattern remains with resulting hypernasality and nasal leakage of air related to the way in which these sounds are produced. It is vital that these children receive speech and language therapy following surgery.
- **Lack of auditory feedback** - patients with severe / profound hearing loss often have abnormal nasal resonance due to a lack of the auditory feed-back during speech production.

It is estimated that 5-20% of patients who undergo surgery for cleft palate have a velo-pharyngeal insufficiency persistant, even after the primary palatoplasty.

This is manifested by:

- Abnormal phonation and speech articulation associated with severe hypernasality;
- Rhinolalia;
- Audible emissions of air from the nose and learning replacement mechanisms;
- Use of consonants different from those that they can not manage to pronounce (typically a compensatory mechanisms in the pediatric patient, eg. M instead of B, GN in place of G, N in place of D);
- Guttural sounds (glottal and pharyngeal extra articulatory sounds);
- Nasal regurgitation of food or liquids, which generally tends to be reduced, however, after the first year of life.

The approach to a velo-pharyngeal dysfunction is primarily by clinical assessment of the quality of the patient's voice. The phoniatric evaluation is fundamental and must precede any instrumental assessment. Whether the issue of language is tied to hypernasality or hyponasality or whether there are other resonance

disorders must be established. The physical examination of the oral cavity aims to identify any oronasal fistulas (which could cause false positives in the search for velo-pharyngeal insufficiency), indicative signs of submucous cleft (like the uvula bifida, the posterior zona pellucida, the step between the hard palate and soft and the V shape of the velum when this is in elevation), to evaluate the symmetry of the facial bones or the presence of scars in the area of previous surgery (palatoplasty, pharyngoplasty or tonsillectomy) or other abnormalities that may affect the verbal production. The medical history will have to collect information not only on verbal aspects and possible speech therapy cycles but also on the way of eating and swallowing, on the possible history of hearing loss or recurrent ear infections, on the previous surgical history and any concomitant diagnosis of genetic syndromes in the patient or in their family. In particular among children who do not have a history of cleft lip and palate but which are studied for suspected velo-pharyngeal insufficiency the syndrome of chromosome 22q11 deletion is common (Catch 22).

The phoniatric assessment falls within the indirect approach to the problem. Through the evaluation of a standardized sample of language that includes nasal and non nasal sounds, repetition of syllables that stress sphincter function and shift from nasal sounds to sounds that are not nasal, and a fragment of spontaneous speech, the presence of abnormal air leakage can be detected, with which sounds they present themselves and how often. Tools that quantify the air escape such as the Nasometer, which measures the acoustic energy produced in the oral and nasal cavities during phonation, can be used to provide a score on velo-pharyngeal nasality of speech, although the reliability of this and other similar devices has not yet been demonstrated.

The direct approach to the velo-pharyngeal insufficiency is what enables the visualization of the sphincter mechanism during phonation without interfering with the movements of the elements that constitute it; it is reserved for those patients for whom the indirect tests have already confirmed the presence of a dysfunction of the mechanism that needs to be corrected and therefore candidates for the aetiological diagnosis and to a characterization of the defect such as to allow the planning of the treatment.

Mainly used are:

- **Video Rhinopharingoscopy** - viewing the nasal cavity, the nasopharinx and the movements of the soft palate during phonation through the use of a flexible endoscope. This technique shows well the location and extent of the defect and it is important the co-presence of the speech pathologist and the surgeon during the examination.

- **Videofluoroscopy** - displays the velopharyngeal port during its three dimensions by assessing not only the movements of the velum but also those of the side and rear walls of the pharynx at rest and during phonation. It uses different radiographic sections, the main ones are the lateral or medial sagittal, frontal and posterior-anterior and basal or axial.

The first allows an anatomical view, greatly appreciated by the surgeon, of the structures. It also has the advantage of not having time limits as there is no exposure to ionizing radiation. The disadvantages are, however, inability to see the whole vocal tract simultaneously, inability to view the oral articulators and, especially in younger children, the possible lack of compliance. Both tests are essential to complete the diagnosis, but because of the increased invasiveness of videofluoroscopy (although in some cases it has shown greater accuracy), many believe they can collect sufficient data through the exclusive use of the endoscopic technique *(Berkowitz S, 2013).*

The surgical treatment plan for velo-pharyngeal insufficiency is made after careful clinical and instrumental examination that demonstrates the anatomical nature of dysfunction and especially after an appropriate speech therapy. The project is aimed at restoring the closing function of the sphincter.

In general, the approach is decided according to the type of defect, coronal, sagittal, circular and to the size, large or small.

When it comes to a modest gap and the muscle fibres seem to elevate in the sagittal direction, it may be sufficient to use the same techniques of lengthening of the velum employed in primary surgery, in particular the Furlow Double Z technique.

In the case of larger gaps with elevator fibres which run transversly so already in the physiological direction it is more useful to intervene on the pharyngeal part of the sphincter.

The possibilities are as follows.

Augmented Pharyngoplasty

When the distance between the rear edge of the velum and the pharynx is less than one centimeter the thickness at the level of the tubercle of the atlante can be increased using autologous implant material (cartilage, bone) or heterologous (Silastic, Teflon, Goretex) in a pocket obtained through sovrafascial disconnection. The main issue is represented by the long term outcome of the implanted material (displacement, reabsorption, change in shape).

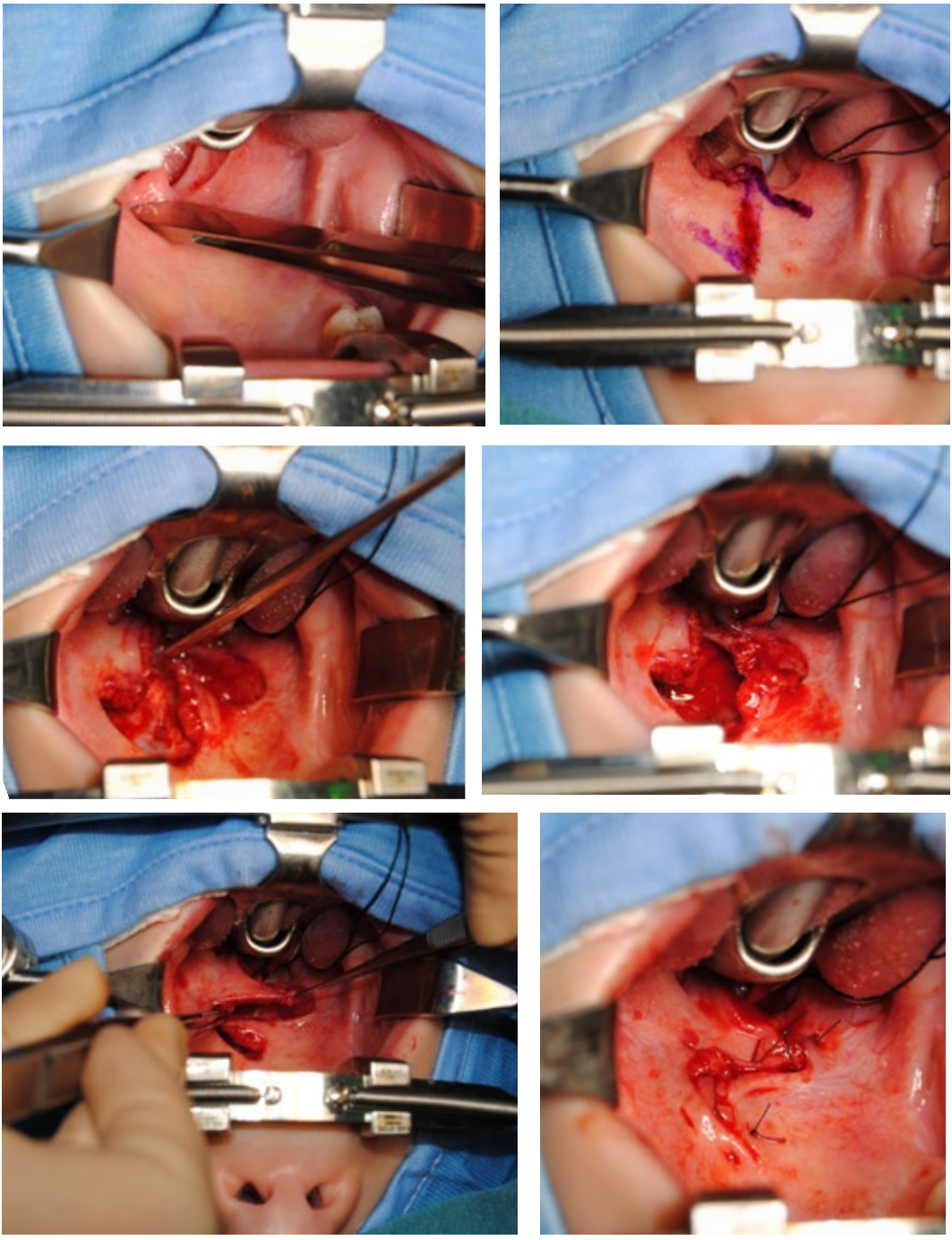


Figure 5.10 Correction of a short palate using the Furlow palate repair.

A further possibility is the injection of autologous adipose tissue taken from the abdomen or the thigh by the Coleman technique. This technique was introduced in 1926 by Von Gaza in which he described an approach by cervical access. More recently (2001) Dejonckere et al have proposed and implemented the transoral nasobroncoscopical implant. The injection of about 12 ml of fat is done using a syringe with a 20 gauge needle at submucosal level of the medial and paramedial of the posterior pharyngeal wall in the contact point between the velum and the pharynx during phonation. The area is exposed during the procedure with the help of a spatula to lower the tongue and endotracheal tube and retract the velum being careful not to inject too deep or laterally risking ostium tubal damage respectively moving the rear spinal ligament *(Leuchter I, Schweizer V, 2010)*. Many uncertainties regarding candidate patients remain for this technique including the risk of complications, the volume and the ideal site for injection, due to the lack of prospective and / or randomized studies comparing this with other methods *(Bishop A, Hong P, 2014)*.

Pharyngoplasty with pharyngeal flap

This technique is indicated especially in patients with a sagittal fault with a deficit close to the centre. The pharyngeal flap which can have a triangular or trapezoidal shape is carved on the back wall of the pharynx and can be lower based, lateral or upper based (the latter is more used today).

There are many variations, the most common is that of Sanvenero Rosselli in which a sagittal section to the whole thickness of the velum with the creation of two flaps is performed first. An upper based flap obtained from the posterior wall of the pharynx using the superior constrictor muscle of the pharynx and the mucosa is then prepared. The velum of the flaps are opened and folded and sutured to the mucosa of the pharyngeal flap with the nasal mucosa, while the rear part of the pharyngeal flap is incorporated in the closure of the two palatine flaps. It is important to coat the surface of exposed muscle with the pharyngeal flap to prevent coarse scar retractions. When the suture is completed the oral cavity communicates with the nasal cavity through two lateral orifices. It is a static operation that permanently alters the clearance between the nose and pharynx.

Dynamic Pharyngoplasty

This technique is based on the principle of transferring the lower insertions of the posterior palatine pillars from the lateral walls to the back of the pharynx building a sphincter that can, contracting, separate the oral and nasopharynx.

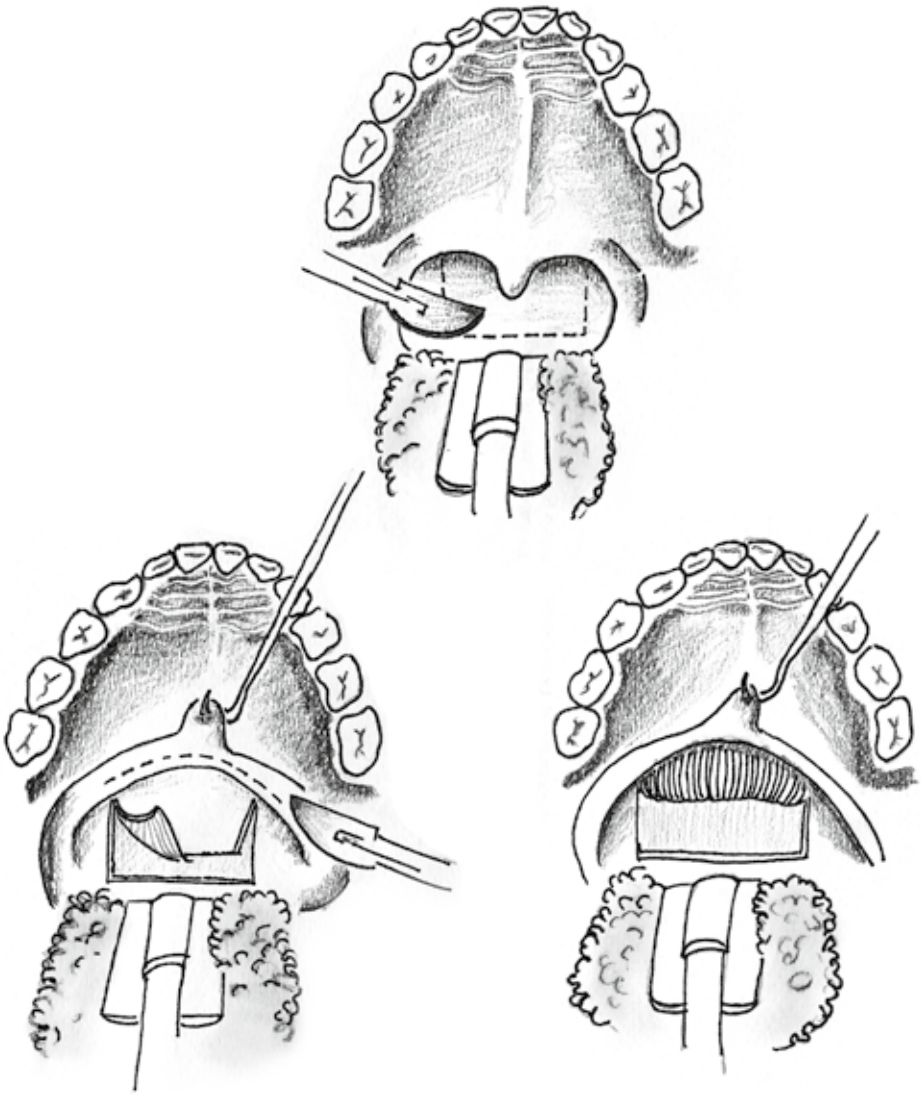


Figure 5.11 Schematic representation of upper based pharyngeal flap for static pharingoplasty.

This creates a mechanism that facilitates the occlusion of the pharyngeal circumferential velum door.

This intervention in its variants (Orticochea, Reichert, Jackson) involves preparation of two upper based flaps obtained by dissecting the posterior palatine pillars and palatopharyngeal muscles that are included in them. These flaps are medialized inferiorly and transposed by 90° and finally sutured on the rear wall of the pharynx making a double Z. The level of this transposition is critical for the

success of the intervention and must match the pharynx point of contact with the velum determined through the preoperative imaging.

There is a modest consensus in the literature about the superiority of the latter technique over previous ones. Failure is often linked to a dehiscence or by improper preparation of the flaps. The most insidious complication in common with static pharyngoplasty is obstruction of the nasal airway with consequent frequent sleep apnea. The onset of hyponasality should still be seen in the context of a bad outcome.

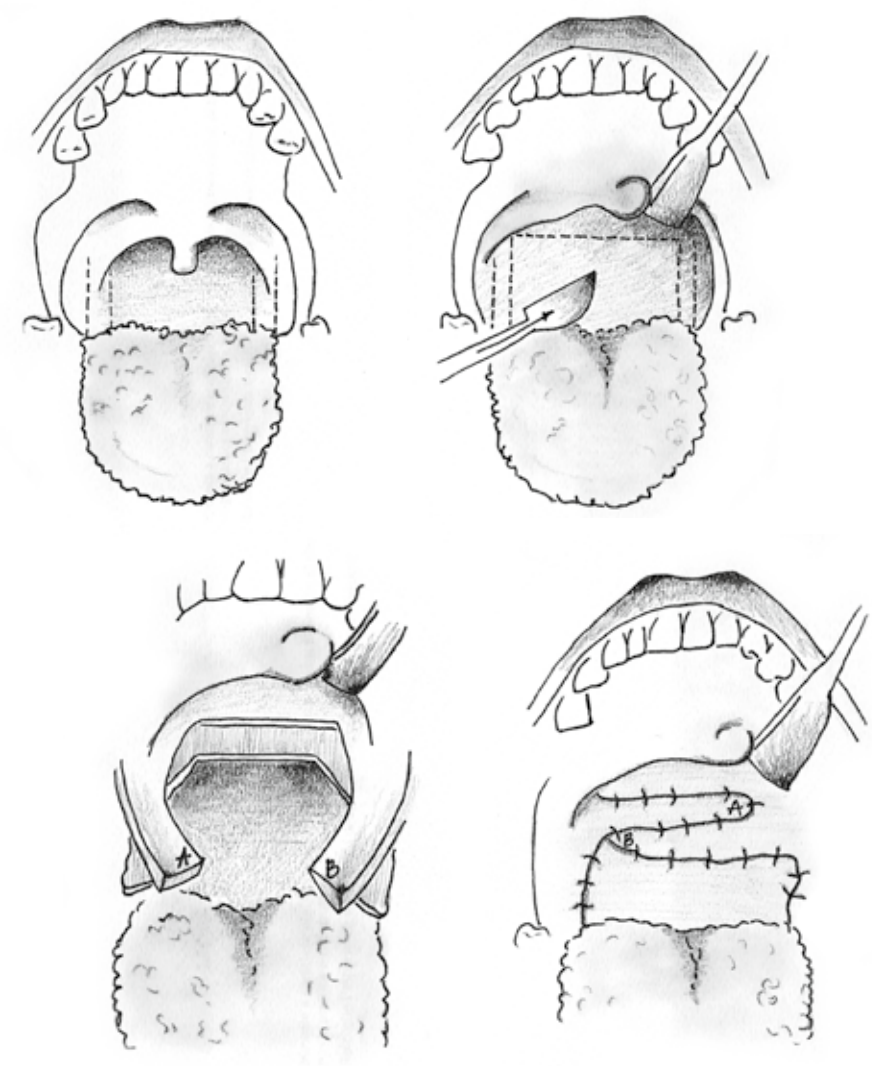


Figure 5.12 Diagram representing the dynamic pharyngoplasty according to Orticochea.

There is disagreement with regard to the most appropriate timing for velo-pharyngeal correction between those who argue that this is closely related to the phonetic outcome (Skoog argued the need to practice surgery before 10 years of age, 1965) and those who believe there is no relationship. In general we can say that it is prudent to plan the surgical correction as soon as it is established that there is the presence of anatomical dysfunction, as long as both the patient and the family are well motivated to take advantage of the benefits that this can bring to speech production *(Berkowitz S, 2013).*

6.
Pierre Robin Sequence

The Pierre Robin syndrome was described for the first time in 1923 as an association of three malformations: micrognathia, glossoptosis and obstruction of the upper airways.

In this first description cleft palate was not considered a significant feature.

From the etiopathogenic point of view, the primary defect consists of a development arrest with consequent mandibular hypoplasia.

Between 1960 and 1970 cleft palate began to be considered as a distinguishing element in this sequence.

The term syndrome was later replaced by the term "sequence" to specify it better as an initial pathogenic factor from which many different clinical pictures can be derived. A specific genetic abnormality responsible for this sequence has not yet been recognized. The mandible originates from the first branchial arch when there is a shutdown of its embryonic growth. The result is a micrognathia which in turn causes backwards displacement of the tongue base. If this occurs before the eighth week of gestation, the anomalous position of the tongue prevents the closing of the palatine lamina resulting in cleft palate.

The incidence varies from 1 in 8500 live births to 1 in 14,000. In 58-70% of cases the malformation is isolated and in the remaining cases it is generally associated with other syndromes (Stickler, Catch 22, Treacher Collins).

It is possible that the factor that has hindered the mandibular growth in utero is less dominant after birth and the jaw recovers in the first year of life to almost physiological dimensions (Figure 6.1). This "catch-up" phenomenon is mainly in the isolated forms of the sequence. For this reason it is important to carefully select patients who need corrective treatment compared to those who can, at least initially, be treated more conservativly.

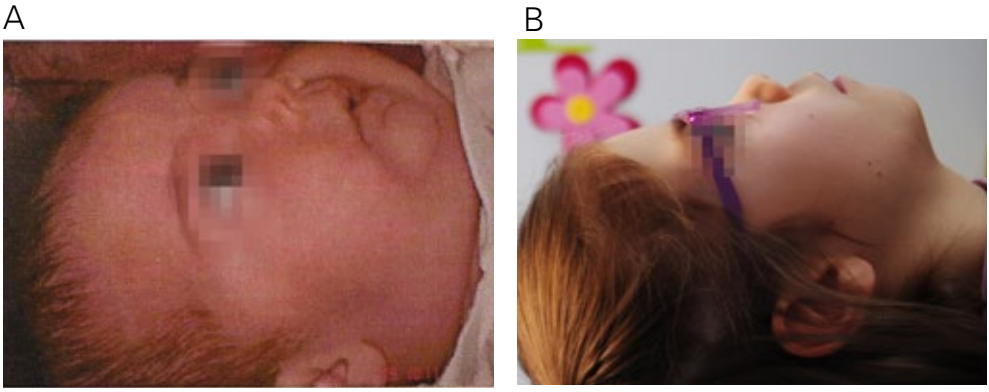


Figure 6.1 Patient with Pierre Robin sequence. (A) First months of life. (B) Nine years old.

Physical examination at birth makes evident the malformation triad of marked mandibular hypoplasia, presence of small tongue with backwards displacement of the tongue base; the cleft palate is only of the secondary palate that runs from the uvula to the incisive foramen and may present as a V-shaped but more frequently in the form of U.

The top priority in patient management is directed to early treatment of respiratory distress, which can be in a more or less severe form. Acidosis from hypercapnia plays a vital role in maintaining a right-left shift (hypoxia and hypercapnia, increase the resistance of pulmonary vessels) that affects further respiratory distress. Increased vascular resistance can also lead to the aggravation of any heart defects. Problems related to nutrition are equally important; facial malformations are responsible for difficulty in sucking and swallowing. The risk of irritation of the airway or aspiration pneumonia is common and worsens the already present respiratory distress.

The possible association with other anomalies requires the execution of a series of imaging studies in the neonatal period including cerebral ultrasound, echocardiogram, electrocardiogram and any neuroimaging.

Treatment

The therapeutic priority is the stabilization of the newborn from the respiratory point of view with the classic procedures of neonatal resuscitation. For patients with moderate defects positioning prone or on their side to keep the airways open may be sufficient. When this is not enough, nasotracheal intubation is required.

In these cases the obstructive problem must be addressed right from the start

by surgery in order to permit the subsequent extubation of the newborn. Over the years different techniques have been developed to correct the glossoptosis. The glossopessia described by Douglas-Routledge using a stitch to fix the tongue to the lower lip, has been abandoned in favor of other approaches; one of the most satisfactory seems to be early palatoplasty.

The complete or partial closure leads to a new intra-oral suspension with front shift of the tongue and increases the ability to suck and swallow avoiding the persistence of pathological reflexes. Early closure of the palate, however, is the treatment of choice in non severe cases and the effectiveness is closely correlated with the degree of micrognathia.

For severe cases, where associated abnormalities that increase the risk of surgery are not present, the last twenty years have seen the emergence of early distraction osteogenesis of the mandible that has proven effective in reducing the proportion of patients requiring tracheostomy. The mandibular osteotomy is performed through an inverted L Risdon incision.

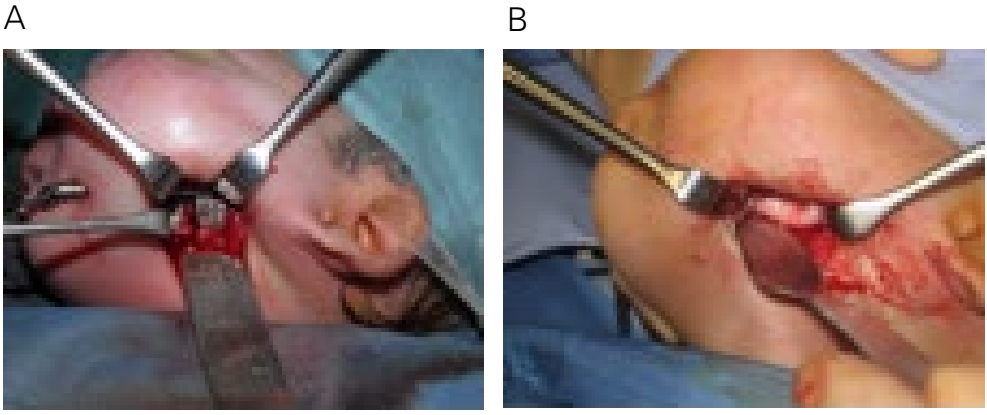


Figure 6.2 Early distraction osteogenesis. (A) Ibrid technique: positioning of the distraction device - Risdon incision. (B) Removal of the distraction device: new bone.

Several clinical studies have demonstrated a better outcome of the mandibular distraction, which today is also applied by means of endo-oral distractors, with respect to tracheostomy in terms of saturation, apneic events, gastro-oesophageal reflux but with an increased risk of ankylosis of the temporo-mandibular joint in the long term.

Increased risk of ankylosis of the temporo-mandibular joint in the long term should be evaluated in relation to the general state of the newborn. In cases of non severe obstruction food gavage may successfully be combined with accurate nursing manoeuvres such as the prone posture during the administration of the meal.

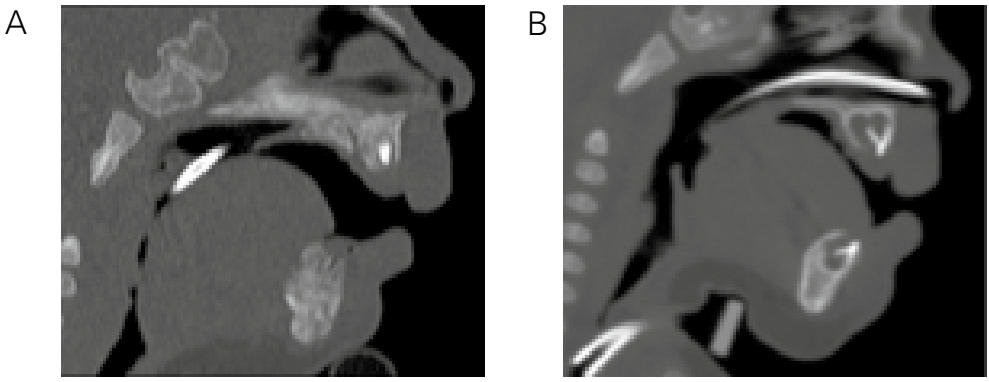


Figure 6.3 (A) The airways before distraction osteogenesis. (B) The airways after distraction osteogenesis.

A palatal plate until the moment in which it is possible to perform the closing operation of the palate can also be useful. In intermediate cases enteral nutrition with feeding tube in cycles are effective. It is critical to retain the reflexes of sucking by stimulating the newborn. In severe cases it may be necessary to use continuous enteral nutrition through gastrostomy or parenteral nutrition.

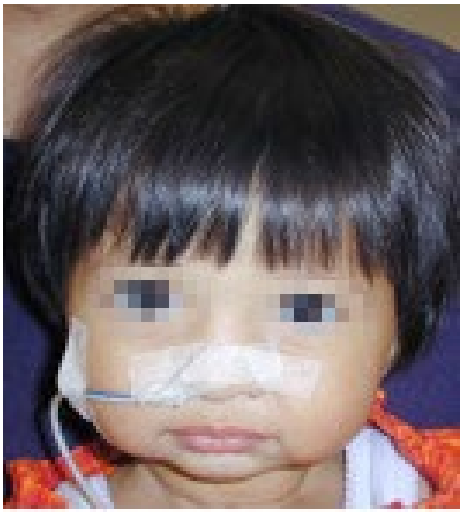


Figure 6.4 Patient affected by Pierre Robin Sequence nourished with a feeding tube.

The presence of a gastro-oesophageal reflux must be early detected and treated with medical therapy to prevent further respiratory complications *(Berkowitz S, 2013; Franchella A, 2007).*

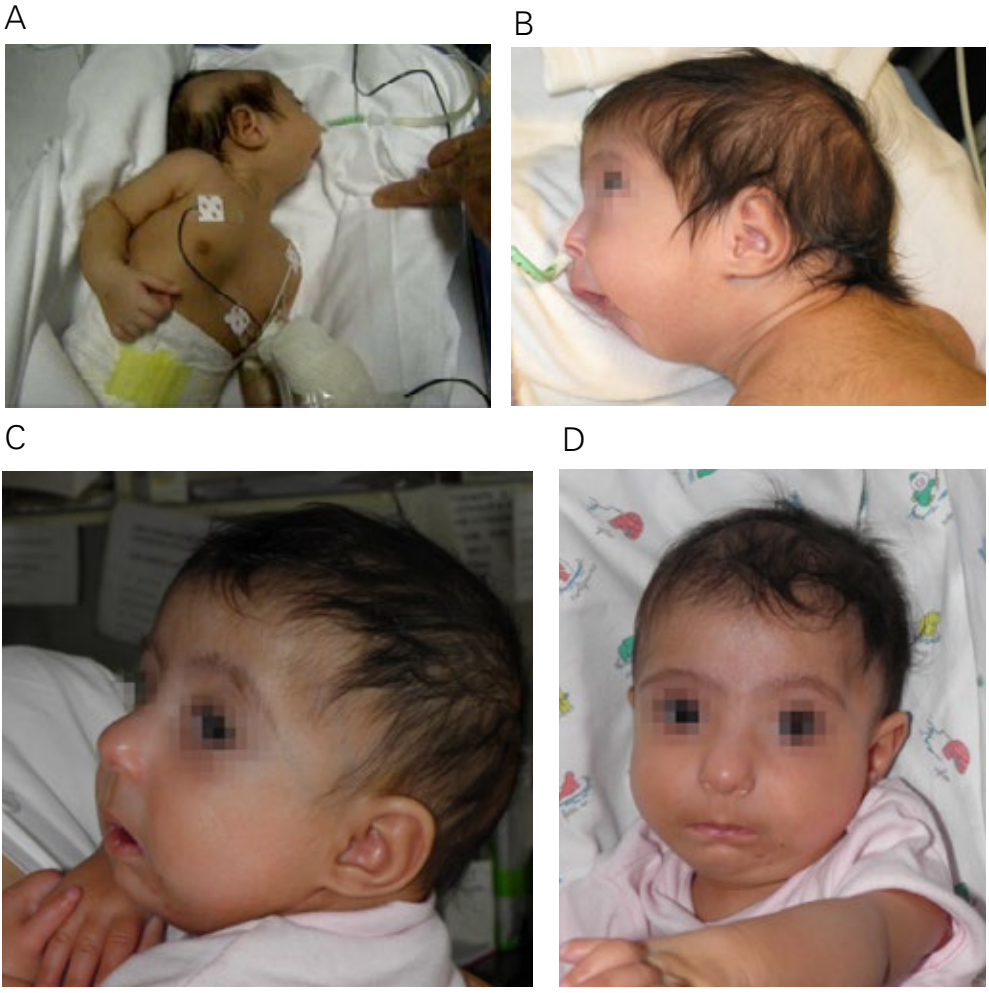


Figure 6.5 Patient affected by a severe form of Pierre Robin Sequence. (A) and (B) Difficult intubation with fibreoptic bronchoscopy. (C) and (D) Results after osteogenetic distraction (mandibular scars) that preceded palatoplasty.

7.
Syndromic cleft lip and palate

Although most facial clefts are isolated anomalies, it is not uncommon for additional anomalies to be present as well. The possible association with other congenital defects may be seen in 13-50% of children with isolated cleft palate. The incidence of associated anomalies is lower in isolated cleft lip (7-15%) and in cleft lip and palate (2-11%) *(Calzolari E, Pierini A, 2007)*.

The association of a specific anomaly/anomalies with a facial cleft may led to the designation of a syndrome with its phenotypic spectrum and genetic involvment.

More than 150 syndromes in which facial cleft occur have been described.

The syndromes associated with cleft lip and palate already characterized by a genetic perspective include:

- Van der Woude syndrome: autosomal dominant, contributes 1% of the syndromic forms, the gene is located on chromosome 1 and encodes interferon 6 (IRF 6) that would seem to have an important role in mediating the craniofacial development interaction between family members TGF beta;
- The CLP Ectodermal dysplasia syndrome: autosomal recessive, associated with mutations in the receptor for polio virus (PVRL1), discovered in a population of Margarita Islands, north of Venezuela. The receptor seems to be involved in the fusion of the palatine processes;
- The ectrodactyly, ectodermal dysplasia, orofacial cleft syndrome (hypoplasia of the hand and foot often associated with syndactyly, dry skin, thinning hair, dental hypoplasia, and cleft lip and palate): autosomal dominant, associated with the mutation of the p63 gene (3q27 cr) expressed in processes maxillary and mandibular development;
- The Ankyloblepharon, ectodermal dysplasia, clefting syndrome or Hay-Wells syndrome: similar to the previous with the addition of fusion eyelid;
- The X-linked Opitz syndrome: gene mutation in MID1 Xp22.

Among the syndromes associated with isolated cleft palate:

• The X-linked cleft palate syndrome that associates cleft palate and ankyloglossia, TBX22 gene mutation encoding a transcription factor family T-box expressed during development of the palate, one of the first genes that has been identified as responsible for cleft palate syndromes;

• Treacher Collins syndrome: malformations of the middle and outer eyelid folded downwards, coloboma of the lower lid, and zygomatic mandibular hypoplasia and cleft palate in 28-35% of cases; the causal mutation concerns TCOF1 gene on chromosome 5q32;

• The Holoprosencephaly syndrome: spectrum ranging from abnormal midline to a complete failure of the forebrain division with cyclopia and often cleft palate, the mutated gene is SHH that code a signal peptide;

• Stickler syndrome (hereditary arthro-ophthalmopathy): autosomal dominant disorder of connective tissue with ocular, auditory, and craniofacial joints (in 25% of cases is associated with cleft palate). The mutation concerns the COL2A1 gene encoding the type II collagen *(Cobourne MT, 2004).*

The recognition of a syndrome has many important implications for the families and for the surgeon.

When a syndrome is identified, the specific defects known to be associated should be sought and children with syndromic orofacial cleft should be evaluated by the Geneticist of the team very early in the course of their care.

The surgical approach to these patients requires timing and techniques personalized for each syndrome.

We describe the approach to one of most frequently diagnosed syndromes in which there is an involvement of the palate.

22q11.2 Deletion Syndrome

The 22q11.2 Deletion Syndrome, also called Di George Syndrome or Velocardiofacial or Catch 22, is a autosomal dominant syndrome characterized by heart defects, facial abnormalities, thymic hypoplasia, cleft palate and hypocalcemia related to a microdeletion of chromosome 22.

This syndrome was first described in 1960 and it is characterized by hypocalcemia, hypoparathyroidism, immune deficiency and heart defects. It is caused by a genetic disorder that almost always includes velo-pharyngeal dysfunction and a real cleft palate or a submucosal cleft, a deep throat and palate-pharyngeal disproportion (skull base obtuse or back) or a hypotonia of the velum.

Associated malformations are heart defects, deficits in cognitive and language

development, dysmorphic facial appearance, vascular abnormalities including tortuosity of the internal carotid and frequent respiratory infections in early life.

The incidence is estimated 1:2,000 to 1:4,000 live births, and is the most frequent syndrome associated with velo-pharyngeal dysfunction or palatine anomalies. The chromosomal deletion that causes this syndrome (22q11.2) is transmitted via autosomal dominance although with variable expressivity. The craniofacial phenotype of affected patients typically incorporates increased size of the jaw vertically and mandibular retrusion, prominent nose with round point and hypoplastic ala, the labial philtrum is long but the upper lip is thin and minor ear malformations can be present.

Language dysfunction includes associated hypernasality, reduced expression of emotions with monotonous language and substitute guttural sounds (the most common compensatory error in patients with cleft palate), articulation skills are more scarce when compared to patients with isolated velo-pharyngeal dysfunction. Since it is a multifactorial defect which probably includes sensorineural loss, these children represent a very serious commitment to both the speech therapist and the surgeon who must therefore work in close collaboration.

Treatment

The ideal timing for patients with deletion of chromosome 22q11.2 syndrome who need a surgical treatment of the cleft palate or of the velo-pharyngeal insufficiency would be between 4 and 6 years. However the intervention is often delayed for the following reasons:

1. Unlike patients with evident anatomical clefts, the malfunction diagnosis requires that these children are at least able to produce a standard language sample in order to be examined;
2. Correction of other associated diseases eg. heart defects can be prioritized;
3. To reach the diagnosis of anatomical velo-pharyngeal insufficiency more prolonged speech therapy cycles are required in order to correct the serious deficit of articulation.

In preparation for surgery, besides the screening and the definition of any other associated malformations, given the high frequency (about 20% in these patients) of abnormal deviations of the internal carotid at risk of surgical damage (close relationship with posterior pharyngeal wall), it is recommended to perform Magnetic Resonance Imaging of the cervical region.

There is no consensus on a specific treatment algorithm, but the literature suggests that in patients with submucosal cleft, palatoplasty alone is not sufficient to correct the velo-pharyngeal dysfunction. For those who have a closure defect of

less than 5 mm, soft palate elongation according to Furlow can be a good compromise to correct at the same time the cleft and the deficit of the palatopharyngeal sphincter. For patients with large defects and for those for whom it is not possible to highlight a submucosal cleft, given the poor surgical outcome and the high incidence persistence of hypernasality it is advisable to carry out pharyngoplasty. The few retrospective works that examined outcomes of phonatory function of different surgical techniques used (in particular posterior pharyngoplasty pharyngeal flap versus dynamic pharyngoplasty) in children with the syndrome have no significant differences. The factors that determine poorer results in children with velo-pharyngeal insufficiency not related to the genetic defect are mainly the greater severity of the initial malfunction and the correction tends to be later *(Sitzman TJ, Marcus JR, 2014)*.

8.
Otologic disease in cleft palate patient

Audiological surveillance

Children with cleft palate or other craniofacial anomalies are patients at risk of otologic diseases. It is important to distinguish patients with isolated cleft palate from those with syndromic forms. The syndromic patients may have audiological issues related to the inner, the middle or the outer ear and usually require a high-level specialist approach in order to decide the best possible therapy for hearing impairment and speech disorders. Patients with isolated cleft palate have different needs, because their otological problems is essentially linked to middle ear disease resulting from a dysfunction of the Eustachian tube.

Syndromic isolated cleft palate

Several syndromes include both cleft lip and palate and otological abnormalities. The most frequent are Treacher Collins syndrome, Pierre Robin sequence, Goldenhar syndrome and DiGeorge syndrome.

Treacher Collins syndrome, also called mandibulofacial dysostosis, has an incidence of approximately 1:50,000 live births and is caused by a gene mutation (TCOF1, POLR1C, POLR1D genes). It has an autosomal dominant inheritance with 90% penetrance and variable expression. The children have facial dysmorphia characterized by bilateral and symmetrical hypoplasia of the zygomatic bone, the infraorbital border or the jaw (retrognathia, retrogenia) which involves a dental malocclusion (characterized by open front bite). In 28% of cases there is a cleft palate and in 60% of cases there are otologic abnormalities, such as anotia or microtia, atresia of the external auditory meatus and anomalies of the ossicular chain; all conditions that result in a conductive hearing loss.

The Pierre Robin sequence has a prevalence of 1:10,000 live births and is characterized by a triad of malformations of the face: retrognathia, glossoptosis and

cleft palate. It is a sequence because the cleft palate is consequent to an abnormal position of the tongue during fetal life that does not allow the fusion of the palate along the midline. The biggest problems that these children have at birth are linked to the retrognathia and the glossoptosis that occlude the airway and prevent a normal diet. If properly handled these issues tend to improve. Unfortunately the risk of ear infections and hearing loss result in difficulty in language development.

Goldenhar syndrome, also called ocular-auricular-vertebral dysplasia, has an incidence of 1:5,000 to 1:25,000 live births, and the cause is unknown. It is characterized by the triad, usually unilateral, of craniofacial macrosomy, ocular dermoid cysts and spinal abnormalities, associated with preauricular tags, ear malformations and sometimes cleft lip and palate.

DiGeorge syndrome, also called 22q11.2 deletion syndrome, has an incidence of 1:2,000-4,000 and is due to a chromosomal aberration. These patients have cardiac abnormalities such as tetralogy of Fallot, defect of the ventricular septum or truncus arteriosus; craniofacial anomalies such as cleft lip +/- cleft palate, velo-pharyngeal insufficiency, hypoplasia of the malar region, hypertelorism, epicanthus, prominent nasal root; vertebral anomalies; immune deficiency associated with aplasia / hypoplasia of the thymus. Other clinical signs can be recognized in these patients as abnormalities of the gastrointestinal tract, deafness, renal anomalies, dental anomalies, learning disabilities and psychological disorders. Given the different clinical forms, the disease has been called by different names like velo-cardio-facial syndrome, cardio-facial, DiGeorge syndrome and 22q11.2 deletion syndrome *(Flynn T, Moller C, 2009)*.

Non Syndromic isolated cleft palate

For isolated cleft palate, the audiological problem is exclusively linked to a lack of tubal function that causes conductive hearing loss and the inner ear or the outer ear are not involved.

Pathophysiology

To understand the disease process that leads to the onset of transmission hearing loss we must first consider the normal physiology of the tubal mechanism. During swallowing the velopharinx sphincter, lifted by the elevator muscles (the levator veli palatini and tensor veli palatini) divide the nasopharynx from the oropharynx so the bolus can not move up to the nasopharynx. The main muscle that regulates the opening of the ostium of the Eustachian tube is the tensor of the soft palate. This muscle originates from the Eustachian tube and inserts into the

palate, where it meets a contralateral homonymous muscle forming a sling part of the soft palate. The middle ear ventilation is related to gas exchange between the mucosa of the mastoid and the blood flow as well as the opening of the Eustachian tube, which in healthy patients happens about 30 times per hour. A consequence of an alteration of the mechanism for opening and closing of the tube (as happens in patients with cleft palate) does not allow a correct ventilation of the middle ear so in these patients there is a higher risk of reflux of nasopharyngeal material towards the tube and then to the middle ear. This situation is common also in healthy children because the tube is horizontal during childhood and it verticalizes only after 7 years of age.

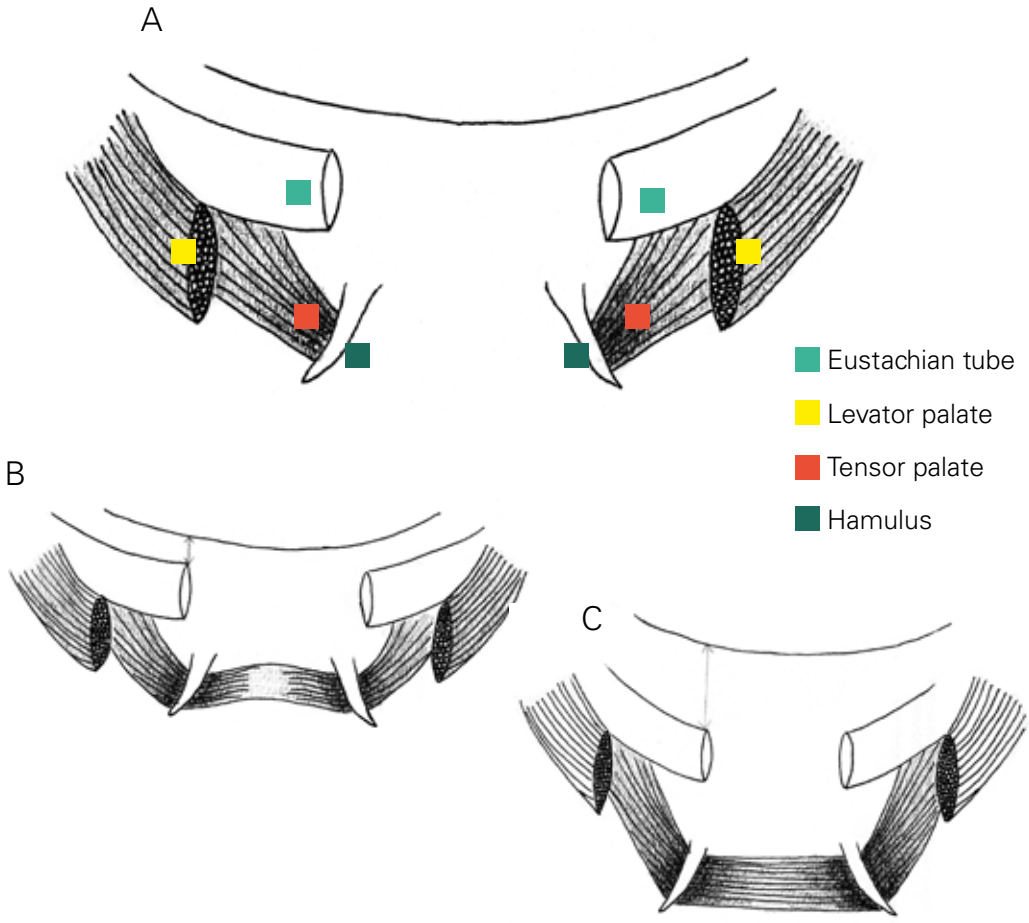


Figure 8.1 Action of palate muscles on Eustachian tube. (A) Child with cleft. (B) Normal infant. (C) Normal child.

The alteration of tubal function predisposes to inflammation and catarrhal persistence. This fluid modifies the impedance of the ossicular transmission system, which is optimal if the medium in which they are inserted is air and decreases if the medium is fluid; this causes a transmission hearing loss. As mentioned it is not uncommon that nasopharyngeal secretions flow up in the tube, thus predispose to infection of the middle ear fluid effusion.

The persistence of otitis media with effusion predisposes to retraction diseases such as the tympanic retraction, atelectasis, adhesive otitis and cholesteatoma.

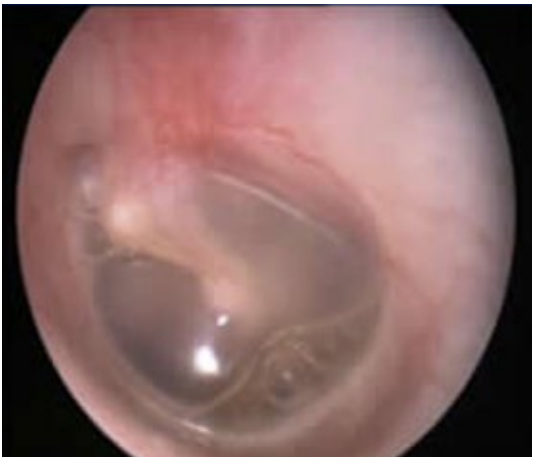


Figure 8.2 Otoscopic aspect of otitis media with effusion.

Diagnosis

Audiological screening is now part of the battery of tests that are performed in newborns in most high income countries. The first audiological test is the otoacoustic emissions (OAEs) that can be performed at the bedside, and, if present, show good hearing level but without giving indications on the state of the central auditory pathways. If the otoacoustic emissions are not present the problem may be at the level of the middle ear or inner ear. An ABR (auditory brainstem response) can then demonstrate the integrity of the acoustic pathways to the brainstem. The evaluation of hearing, however, must always be based on the correlation of more electrophysiological and behavioural tests.

Hearing loss can be measured by about 6 months of age with childhood audiometry, however it requires an audiometrist experienced with children. Normally the tone audiometry that is also used in adults can be performed from 5 years of age.

The diagnosis of acute otitis media (AOM) and otitis media with effusion (OME) is not particularly difficult. Acute otitis media is an acute middle ear inflammation that manifests as pain, fever, ear fullness and objectively manifested by hyperemia of the tympanic membrane, bulging of the eardrum or possibly purulent otorrhea. Otitis media with effusion, shows a more blurred situation: pain and fever are absent while there is auricolar fullness. Objectively it is manifested by shades from yellow to blue of the tympanic membrane depending on the type of exudate, which can be serous, mucous, serum-mucous or chronic mucoid (glue ear).

While in older cooperative children the diagnosis is much simplified by history and can be easily confirmed by a simple Weber test (if unilateral); in small babies OME diagnosis can be confirmed with tympanometry, which in the case of endotympanic exudate will be flat, or with a peak on negative values.

The microorganisms most frequently involved in the acute inflammatory process are *Haemophilus influenzae*, *Streptococcus pneumoniae*, *Streptococcus group A beta-hemolytic*, *Moraxella catarrhalis*, all sensitive to penicillins such as amoxicillin, or possibly the association of amoxicillin and clavulanic acid.

Treatment

The treatment for this kind of problem is not simple, because we have to remember that the main problem in these children is audiological hearing loss linked to the tympanic effusion. The consequences of hearing loss are seen in learning ability and the possibility of use of auditory feedback can correct this issue.

Amplifying sound through a hearing aid would appear to be more effective and less invasive method that could lead to the resolution of the problem. Unfortunately, in most countries the hearing aids are paid by the patient and require experts hearing aid specialist, that can set the best prostheses for the specific problem of the child.

Another option is to act directly on the tympanic exudates through the positioning of a grommet which allows, through this tube, to eliminate the liquid of the middle ear and the system can function properly, at the right impedance. Unfortunately, this procedure is not without risk, since in small children the drainages should be positioned under general anaesthesia and it may be necessary to put them several times as they have an average life of about 6 months. We must also remember that many of the procedures on the tympanic membrane to which these children are subjected often leave scars on the membrane and increase the risk of cholesteatoma.

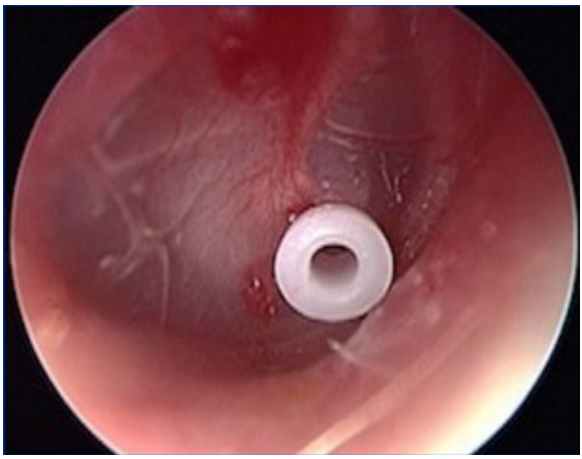


Figure 8.3 Treatment of recurrent otitis with grommet.

Normally, in healthy children (not malformed) suffering from otitis media with effusion it is a common practice to carry out an adenoidectomy if there is evidence of adenoid hypertrophy. In children affected by cleft palate this intervention must be avoided because it will almost certainly lead to a velo pharangeal insufficiency as the adenoids offer good support to allow a good closing of the nasopharynx by the soft palate.

Nasal saline washes can help cleaning the nasopharynx and hence help to reduce the number of middle-ear infections *(American Academy of Pediatrics, 2004; Drake AF, Roush J, 2013)*.

Follow up

The first audiological examinations in these children begin at birth with the otoacoustic emission and possibly the ABR. Often the otoacoustic emissions are absent because of tympanic effusion that does not allow the execution of a correct examination. ABR, in the case of tympanic effusion may also be difficult to perform. This examination is performed with the child sleeping and as they often snore due to the anatomical defect, the noise of the snoring can alter the results.

The first audiometry with a threshold will be performed at 6 months. The child with conductive hearing loss will have to take a conservative path (with hearing aids) or interventional one (with placement of grommets). This treatment is obviously linked to the extent of hearing loss because up to 30-40 dB in a child who does not have other learning problems can be tolerated and well managed with simple environmental measures (such as make him sit in the front rows).

Children who undergo surgery or who use hearing aids should be seen every 6 months to evaluate the clinical outcome. Usually after 7 years of age there is an improvement of transmissive hearing loss, because there is a physiological maturation of the tubal function.

Conclusions

The child with cleft palate is subject to frequent episodes of acute otitis media and often presents persistent otitis media with effusion that causes a transmission hearing loss of varying degrees. The tympanic exudate is related to a defective opening of the Eustachian tube and not always resolves with the repairs of the palatal defect. The management of children suffering hearing loss should be entrusted to an otolaryngologist or an audiologist who should follow the patient from the start of life. Treatment options are essentially two: the use of hearing aids or the placement of grommets *(Lehtonen V, Lithovius RH, 2016)*.

9.

Secondary repair of cleft lip nasal deformity

Correction of nasal deformity associated to unilateral/ bilateral cleft lip

The objective of the secondary surgery in this type of deformity has been described by Padgett in 1929. A good repair of cleft lip and the resulting nasal deformities is important in relation to the final result and is often the most difficult to achieve. The operation becomes an art when the nostrils are similar and well balanced and the lip has a perfect Cupid's bow *(Padgett EC, 1929)*.

Normal anatomy

The visual characteristics of the nose are determined by the skeleton bone and cartilage, covered by soft tissue. The bony portion consists of the nasal bones, equal and symmetrical, which articulate with the frontal bone superiorly and the frontal processes of the maxilla laterally, caudally are in contact with the triangular cartilage. The two triangular cartilages, which again are equal and symmetrical, are divided medially by the quadrangular cartilage, unique and medial, which is part of the nasal septum. More inferiorly there are the alar cartilages, equal and symmetrical, formed by a lateral crus and a medial crus. The medial crus are juxtaposed medially and form part of the columella.

The nasal septum is formed anteriorly by the quadrangular cartilage that articulates postero -inferiorly with the vomer and postero-superiorly with the perpendicular lamina of the ethmoid bone.

Anatomy in the malformation

The anatomy of the malformation is obviously modified by the type of malformation.

In unilateral cleft you can find a deviated nasal tip on the opposite side of the cleft, a retro positioned narinal dome on the side of the cleft, an obtuse angle between the medial and lateral crura, a lack of nose-genien groove with an appearance (or real) bone defect below the fibro-fatty tissue of the wing that is in contact with the cheek. The affected nostril appears bigger than the other one and it is retro positioned. The columella appears shorter on the side with the cleft and the medial crus of the alar cartilage is retro positioned *(Huffman WC, Lierle DM, 1949)*.

In bilateral cleft the columella is very short and the medial crura are badly positioned into the philtrum. An inadequate tip projection with a bifid appearance is clear. The axis of the nostrils is more horizontal than vertical. From the intranasal point of view there is a fault in the vestibular tissue with a flattening of the nasal floor and a retro position of the maxillary bone. As already mentioned the main problem is the length of the columella, shorter than normal, and which is further shortened during the repair operation of the lip to allow the correct setting of the filter.

The nasal septum, both in unilateral and bilateral malformations, almost always appears deviated. The magnitude of the deviation is different from case to case and can lead to unilateral respiratory obstruction.

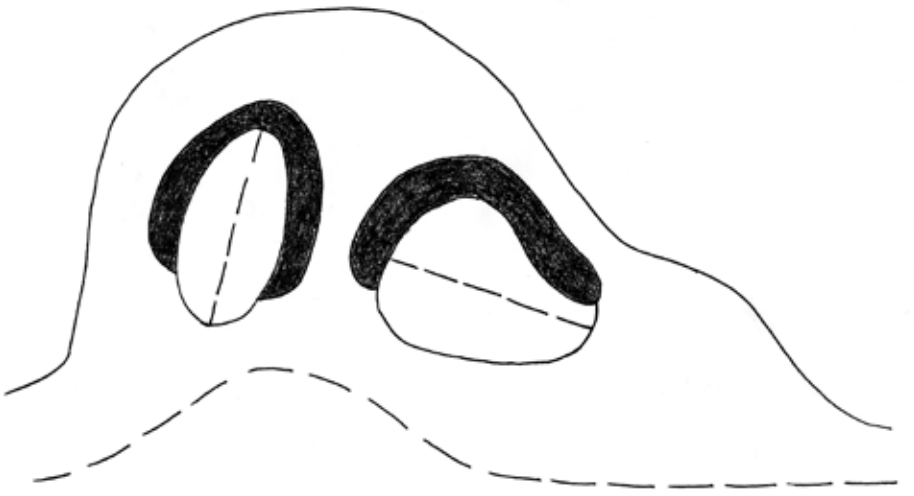


Figure 9.1 Nasal deformity in unilateral cleft lip.

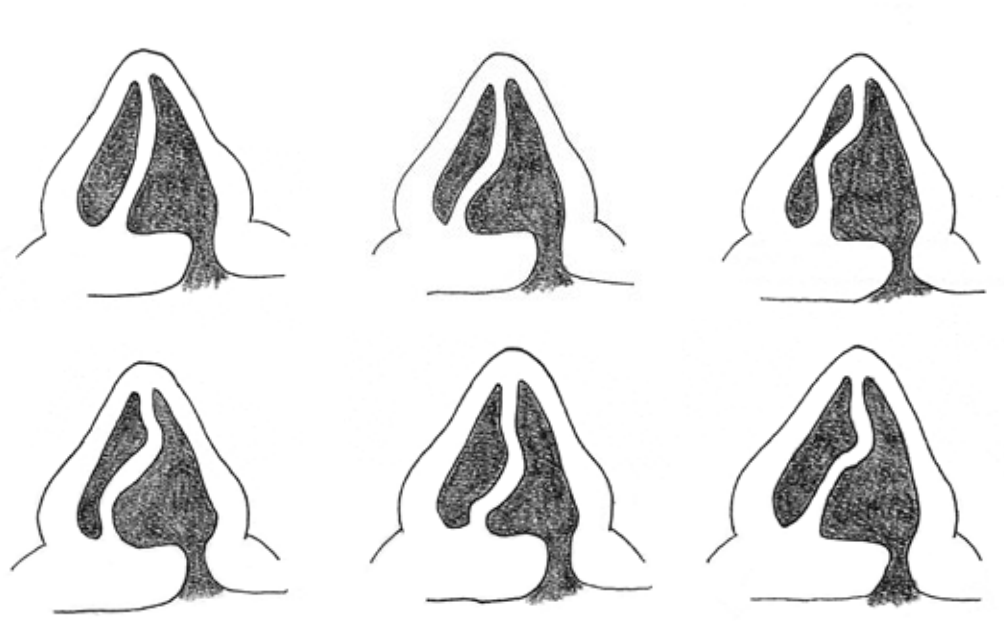


Figure 9.2 Various types of septal deformity associated with unilateral cleft lip.

The surgical objective

The goal in this type of surgery is the same for both unilateral and bilateral malformations: a nose that allows correct breathing, is symmetrical and seemingly normal, in harmony with the rest of the face and compatible with the patient's ethnicity.

Surgical timing

There are no shared guidelines regarding timeliness and standardization to perform this type of surgery. Each surgeon has his preferences and each patient has his or her needs.

A large number of studies show that there is no adverse growth effect in spite of extensive surgical dissection performed during the primary rhinoplasty *(McComb H, 1990)*, only a nasal deformity can persist but no adverse growth effect has been demonstrated *(Salyer KE, Genecov ER, 2003; Henry C, Samson T, 2014)*.

Nevertheless it is generally recommended to perform the rhinoplasty not before the age of 16 for women and 17 years for men. At this age these young adults can decide for themselves if they really want to undergo this surgical procedure. Normally these children have already been subjected to a lot of surgery, including primary correction operations, ortognatic operations, orthodontics and alveoloplasty.

The rhinoseptoplasty should be the operation that closes the course of treatment of this malformation, definitely rehabilitating the patient's face. There are also concerns that not every patient is ready to undergo surgery that changes the morphology and appearance of their image. Patients should therefore be informed that this type of intervention can also be performed later.

Regarding septoplasty it has been demonstrated that the correction of the deviated septum in unilateral cleft during the primary rhinoplasty is effective, and the improved septal position is maintained into adulthood *(Anderl H, Hussl H, Ninkovic M, 2008; Gawrych E, Janiszewska-Olszowska J, 2009)*.

The functional repair of the nasal septum, to allow proper breathing, is better done in one operation along with the aesthetic nose correction, performing a rhinoseptoplasty. This reduces the number of procedures in general anaesthesia and allows autologous septal cartilage for any aesthetic corrections to be available.

Surgery

Septoplasty: the purpose of the septoplasty is to put straight the nasal septum, that does not allow (or reduces) the ability to breathe through one of the two nostrils. This obviously creates a considerable problem for the patient that, especially during hyperventilation, must breathe with the aid of the mouth bypassing the nasal filter.

The operation is performed under general anaesthesia with the help of a front light.

Gawrych and Janiszewska-Olszowska *(McComb HK, 2009; Sykes JM, Tasman AJ, 1990)* perform a limited septoplasty during the primary rhinoplasty; they release the septum from the nasal spine and the maxillary groove on the non-cleft side and then allow to straighten out toward the side of the cleft.

Instead the classical intervention can be performed by either open or endonasal approach including an adequate exposure and a complete breakdown of the ligamentous attachment that contribute to the septal deviation. The complete transfixion incision is used if the rhinoplasty is performed at the same time. Once identified the perichondrium of the quadrangular cartilage it is detached from the cartilage bilaterally; now is possible to resect a strip of cartilage inferiorly, allowing the septum to swing over the nasal spine, and this position can be maintened by suturing the cartilage to the spine; the removed strip of cartilage can be utilised to support the nasal septum or to fill a defect for the rhinoplasty. It is important to position silicone plates on the two sides of the septum to hold it in place for 10-15 days with anchor point; the nasal packing is to maintained for 1-2 days *(Xu H, Salyer KE, 2009; Price JC, 1990)*.

Rhinoplasty: the purpose of rhinoplasty is to improve the aesthetics of the nose. Normally during the primary lip correction also the primary nose correction is performed. In this kind of patient it is generally recommended to perform an open rhinoplasty, rather than the close technique that often threatens to leave behind deformities or areas incorrect. In unilateral cleft the incision will have to cover the bottom margin of the alar cartilages to join up at the level of colummellar margin, from the side of the cleft the incision must continue into the philtrum. With this incision you can then lift the tip up to the nasion and better expose the entire osteo-cartilaginous nasal structure and its deformities. It is then necessary to free the alar cartilage of the side with the cleft from the nasal vestibule in order to reshape the lateral portion of the cartilage and to give symmetry to the two nostrils. The skin will be turned over and the suture completed. Another technique that may be used in a unilateral malformation was described by Dibbel *(Van der Heijden P, Korsten-meijer AG, 2008; Dibbel DG, 1982)* which provides a unique approach using an external incision with reduction of the skin on the side of the cleft, changing the long axis of the nostril on the side of the cleft. Essentially the nostril on the side of the cleft is completely freed and rotated to the new position and a part of the nostril skin is removed. The rotation of the nostril brings the alar cartilage to its normal position, lengthening the columella from the side of the cleft and thus creating a symmetrical tip. By turning the nostril also the cheek could move and the removal of a small piece of tissue in the lower level of the vermillion may be needed. This step provides the opportunity for a revision of the lip scar. Obviously this type of technique can be combined with a true open technique to allow a better exposure of the osteo-cartilaginous structures.

The surgical approach in bilateral malformations will be different, since the main problem in this type of malformation is the short columella, which often has already been further shortened during the repair of the lip. In addition the tip of the nose is badly positioned following the separation and lowering of the dome of the ala. Various techniques are used depending on the type of problem. Cronin's technique, for example, can be used when the columella is not too short, the arc of the Cupid's bow is normal and when it is not necessary to revise the lip scar. The incision extends from the groove between the cheek and wing cartilage, immediately passes inferiorly to the nostrils and is joined medially at the level of columella with an inverted V. It then clears the entire base of the nose that allows the rotation and the advancement of the nostrils. This allows a lengthening of the columella and a repositioning of the tip. Autologous cartilage can also be used to give an additional support *(Cronin TD, Upton J, 1978; Cronin TD, 1958).*

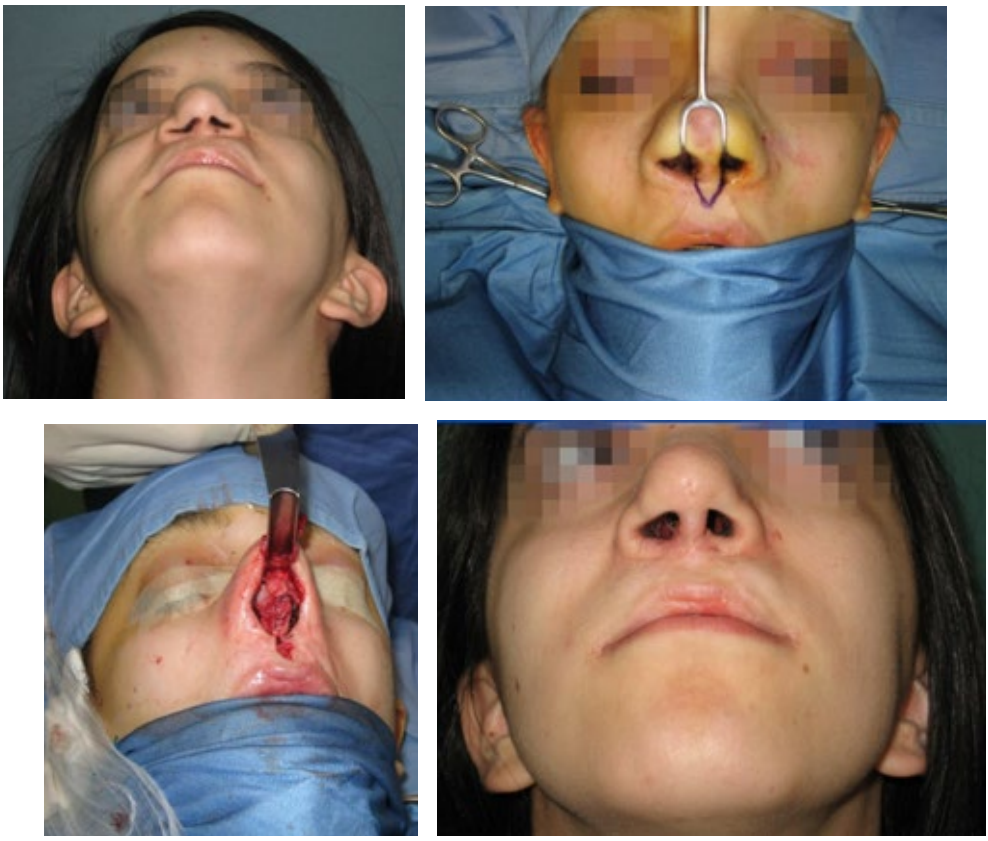


Figure 9.3 Bilateral cleft lip and palate Open rhinoplasty with auricular autologous cartilage graft.

The forked flap is another technique described by Millard *(Millard DR Jr, 1958),* mainly used when the philtrum is rather wide, when it is necessary to revise the scar on the lip and when the skin of the nasal floor is insufficient. The incision will trace the scars of the lip and will extend along the base of the nostrils, forming two forked flaps in the upper base. This allows the revision of the lip scars, the improvement of the filter and the repositioning of the alar cartilages. Often this procedure requires a columellar revision after some time.

The V-Y technique is another procedure used in bilateral cleft, allowing a lengthening of the columella and an advancement of the philtrum. The incisions run along the labial scars and are extended along the margin of the alar cartilages and along the base of the nose. This allows a rotation which causes the columellar to lengthen, and the lip is resutured with a V-Y shape. If a further lengthening of

the columella is required it can be associated with a strip of Abbé *(Abbé R, 1968)*, which provides for the preparation of a pedicle flap of the lower lip that goes to fill the defect of the upper lip, the stalk is then cut about 2-3 weeks after surgery.

Conclusions

The secondary surgery of the nasal deformities should be kept to a minimum thanks to a correct surgical planning during the primary surgery. However, deformities and blemishes are hard to avoid and present an ongoing challenge for the surgeon who reconstructs the nose. This surgery should be the end point of the course of treatment, and should be performed by experienced surgeons who deal frequently with such problems.

10.

Retrospective Analysis

of cases treated by the Pediatric Surgery Department, Saint Anna Hospital, Ferrara

10.1 Sequence of treatment procedures used at the Pediatric Surgery department of University Hospital of Ferrara

Over the years a selection of treatment techniques and their timing has been developed by our department. The following table shows the timing and techniques adopted for different forms of cleft lip with and without cleft palate.

Unilateral cleft lip repair

Timing: *3-6 months of age*

Technique: Millard technique / Skoog labioplasty

Bilateral cleft lip repair

Timing: *3-6 months of age*

Technique: Salyer modification of the Millard technique

Primary correction for nasal deformity

Timing: *3-6 months of age (at the time of primary repair)*

Technique: McComb technique

Palate repair

Timing: *8-18 months of age*

Technique: Two flap palate repair / Furlow Z-plasty palate repair / Widmayer Perko two stage palate repair

Correction for secondary lip deformity

Timing: *before 5 years of age*

Technique: Various techniques

Oronasal fistula

Timing: 6 months after the previous surgery

Technique: Transposion flap / Re-do palatoplasty

Velo-pharyngeal insufficiency

Timing: related to the phonetic outcome, 4-6 years of age

Technique: Pharyngeal flap / Orticochea Dynamic pharingoplasty / Pharingeal lipofilling

Alveolar cleft

Timing: 7-9 years of age

Technique: Cancellous bone graft from iliac crest

Correction for secondary nasal deformity and septoplasty

Timing: 16 years for females, 17 years for males

Technique: Various techniques

Submucous cleft palate

Timing: at the age of diagnosis

Technique: Furlow Z-plasty palate repair

Orthognatic surgery

Timing: 14-15 years for females, 16-17 years for males

Technique: Le Fort I Maxillary osteotomy / Distraction osteogenesis

10.2 Pre and postoperative management

The blood tests are performed as required by our usual preoperative screening for major surgery (blood count, coagulation structure, cholinesterase and creatine) and an anaesthesist evaluation is also carried out. The anaesthesist may engage the bronchoscopist in cases of possible difficult intubation primarily related to retrognathia, particularly present in the Pierre Robin Sequence.

Whatever the patient's age, the minimum hospitalization is 5-7 days and includes 24-48 hours of fasting with an intravenous hydro-electrolytic infusion and then a gradual reintroduction of food starting with clear liquids, then with milk or semi-liquid diet or pureed if the child is already weaned. The liquids and the

milk are administered by means of a syringe with a short piece of soft tubing, the tip of the tubing is inserted into the mouth and 3-5 ml are delivered to the back of the tongue, at which point the swallowing reflex takes place. This method is necessary until the infant is able to feed from a cup or using a soft spoon, avoiding a bottle, pacifier and straws. This regime is generally followed when the child goes home under guidance for at least 20 days after admission. Before awakening in the operating room semi-rigid bracelets which interfere with the flexion of the elbow to bring the hands to the mouth are applied if the child is 3 years or less. These are kept in place during the hospital stay and if possible for the first few days after discharge if the child tolerates this.

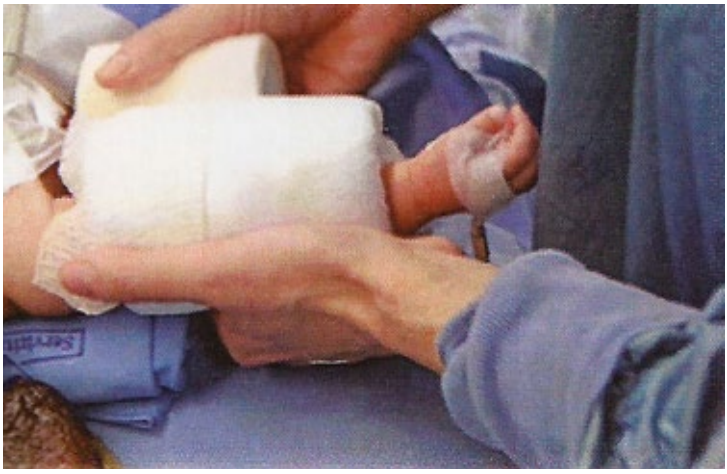


Figure 10.1 Semi rigid bracelets.

For the first 48 hours analgesic therapy provides strong painkillers (opioids such as morphine 0.1-0.2 mg/kg or Tramadol 1.2 mg/kg at fixed times or continuous infusion via elastomeric pump) alternating with acetaminophen at 15 mg/kg associating corticosteroids at low doses if surgery also involved the palate and pharynx to reduce oedema and facilitate the respiratory function. A gradual transition is followed towards paracetamol administered either endorectally or as an oral syrup at fixed times and then as needed. Prophylactic antibiotics are administered intra and post-operativly for at least 7 days with broad-spectrum antibiotics (usually amoxicillin and clavulanic acid) preferred.

A now consolidtate practice is that the lip and palate in the area that will be affected by the incisions is treated with local anaesthetic (usually Mepivacaine, 3mg/kg) associated with adrenaline (0.5 to 0.6 mcg/kg) to reduce bleeding (Figure

10.2 A). The sutures on the palatine mucosa, labial and nasal floor are made with a braided fast absorbable 4.0 or 5.0 thread. At the end of the palatoplasty the suture lines and any raw areas left uncovered are protected with fibrin glue (Figure 10.2 B).

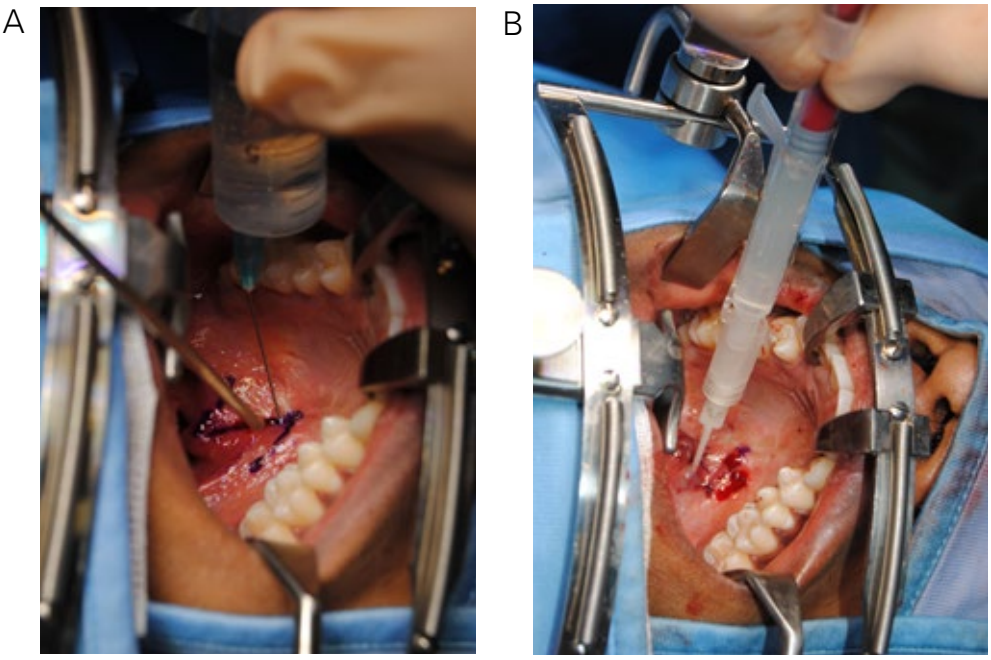


Figure 10.2 (A) Injection of mepivacaine-adrenaline solution. (B) Sealant application.

Since for skin closure in labioplasty non-absorbable thread is used, the patients are returned to the operating room five days post-operative to allow the dressing and the removal of the suture material under sedation.

Periodic appointments are organized in an Integrated Ambulatory Package scheme which provides as well as surgical outcomes and check ups, the evaluation by the other specialists involved, in particular:

- Dentist and Orthodontist;
- Audiologist;
- Speech Pathologist;
- Speech Therapist.

The frequency of the controls will be established by each specialist according to the needs of the individual patient. The Maxillofacial Surgeon is involved by

the Pediatric Surgeon or directly by the dentist if maxillary surgery is needed to correct large discrepancies of the facial skeleton.

10.3 Patients

The graphs below show the percentage distribution of primary surgery indications, associated malformations and surgical techniques employed, observed in patient with orofacial cleft referred to our Department since 1980.

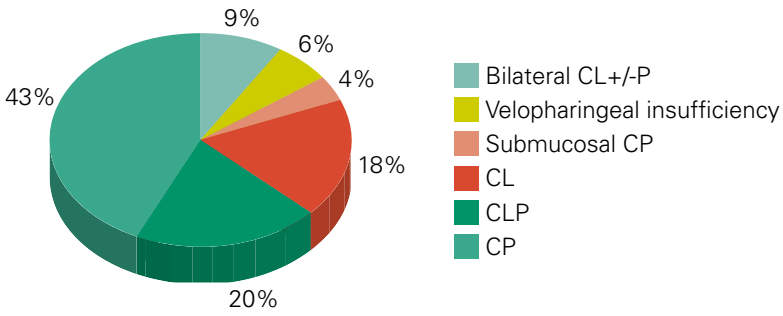


Chart 10.3 Chart percentage of cleft defects observed in our department (CL=cleft lip, CLP=cleft lip and palate, CP= cleft palate).

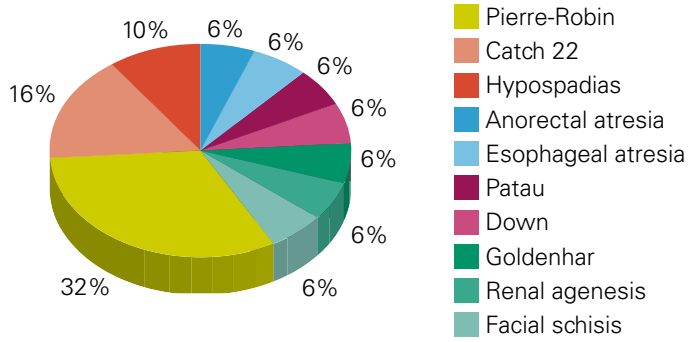


Chart 10.4 Associated diseases.

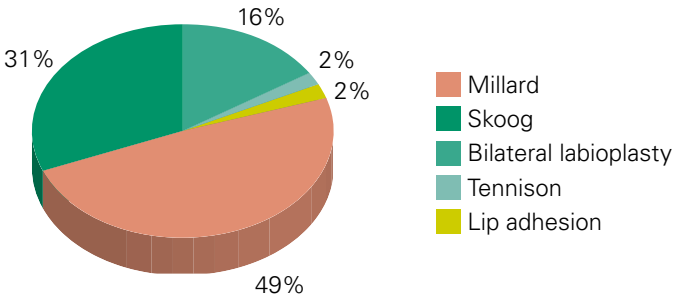


Chart 10.5 The surgical techniques employed for lip repair.

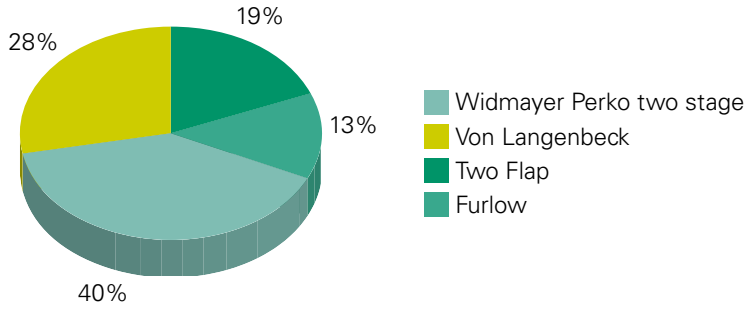


Chart 10.6 The surgical techniques employed for palate repair.

The average age for the execution of labioplasty was 5.2 months, for posterior palatoplasty 16.3 months, anterior 23.1 months. A patient with posterior cleft palate was referred to us at 13 years old and was simultaneously subjected to posterior palatoplasty and pharyngoplasty according to Sanvenero-Rosselli as phonetic results typically are inferior when obtained by performing the correction in adolescence. The primary palatoplasty cases also include interventions to correct the submucosal cleft. For this correction an average age of 54 months or 4.5 years, confirms the later diagnosis of this defect.

The majority of complete cleft palate was corrected in 2 surgical operations, usually Widmayer-Perko for the first time (secondary palate) and Von Langenbeck for the primary palate in case of good reduction of the defect following the first intervention or new Widmayer-Perko in the event of persistence of wide cleft of the hard palate, which has occurred in only a third of the cases. In cases of closure in one operation preferentially the Two flaps technique was used and to a lesser extent the Von Langenbeck.

10.4 The outcomes

The goals of cleft care are to obtain optimal aesthetic, speech, growth and hearing functions according to American Cleft-Palate Association (1993). The outcome treatment focuses on two areas:

1. Speech development.
2. Facial growth.

The primary goal of cleft palate repair is to provide for the development of normal speech without interference on facial growth.

1. Speech development

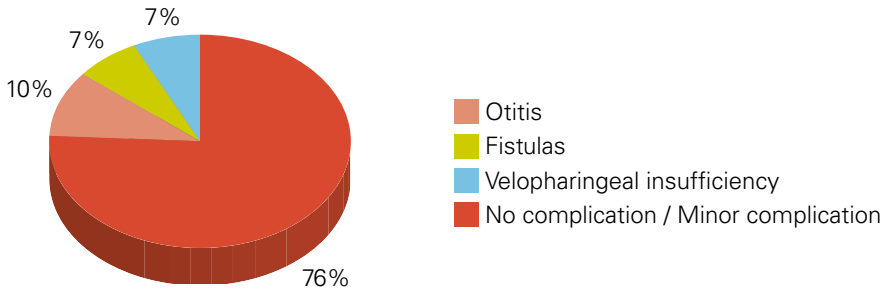


Chart 10.7 Report of the percentage of complications, related to speech development.

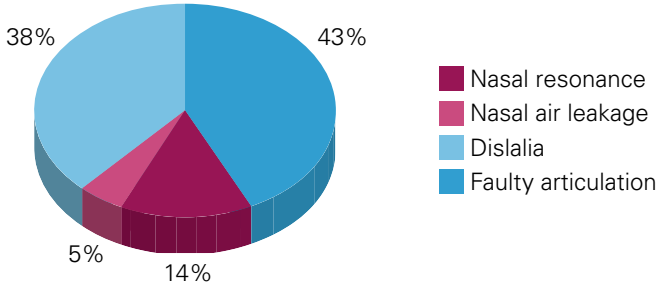


Chart 10.8 Report of the percentage of speech disorders.

In 28.6% of patients who followed the proposed follow-up a conductive hearing loss, unilateral or bilateral or mild, linked to recurrent ear infections was reported.

Placement of endotympanic drainage was not found to be an indication rou-

tine in these cases. Two patients were found to have hearing abnormalities not dependent on the cleft, one with sensorineural bilateral hearing loss in Pierre Robin sequence and one with unilateral ear atresia in Goldenhar syndrome.

Patients treated for primary velo-pharyngeal insufficiency, divided between the techniques used in a fairly uniform manner between dynamic pharingoplasty (Orticochea) and plastic elongation of the soft palate (Furlow). Only one patient in the first instance was operated with a static pharyngoplasty using a superior based Pharyngeal flap (according to Sanvenero-Rosselli).

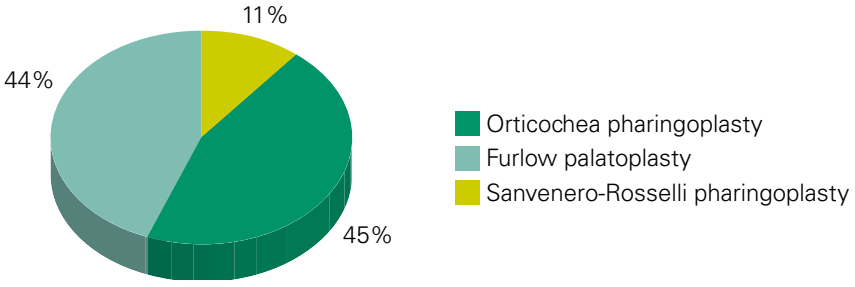


Chart 10.9 Percentage of surgical techniques for primary velo-pharyngeal insufficiency.

Leading the therapeutic and surgical choice in these cases is almost always a rhino-pharingoscopy and inter-disciplinary assessment with the speech therapist who follows the patient and who will have oriented the diagnosis towards a dysfunction of an anatomical type. Two patients were affected by the 22q11 deletion syndrome that is typically associated with this disease, a third patient had Trisomy 21.

2. Facial growth

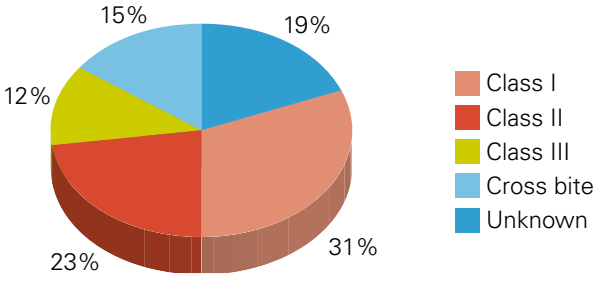


Chart 10.10 Percentage of malocclusion classes and crossbite.

Patients who necessitated postsurgical orthopaedics treatment suffered mainly from Class III malocclusion associated with anterior cross bite and / or lateral or upper dental arch overcrowding or need for extraction of palatine teeth at the defect.

3. Secondary surgery

The rate of patients that required at least one secondary surgery intervention was 16.5%. These included closure of fistulas in the palate, lip revision, re-do palatoplasty, pharyngoplasty to deal with velo-pharyngeal insufficiency, revision of the lip scar, autologous injection of adipose tissue in the retropharynge (lipofilling) and secondary rhinoplasty (Chart 10.11).

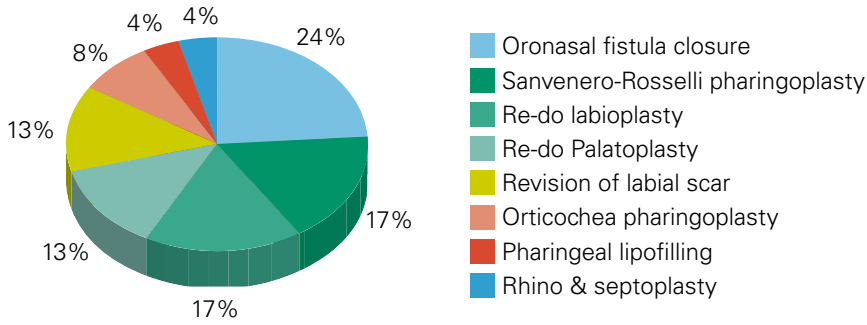


Chart 10.11 Secondary surgery in patients previously treated with primary surgery

Among patients who underwent complete correction of cleft palate with a single operation, 13% have needed secondary surgery, among those who had corrective surgery in two stages, 16.1% were then subjected to further secondary surgery.

The incidence of velo-pharyngeal insufficiency which required secondary surgical correction was 5.8% (7 patients).

From the total of patients undergoing re-do palatoplasty we excluded those patients undergoing surgery for submucous cleft and velopharingeal insufficiency which as previously mentioned has a higher average age for correction and analyzed the outcome of a homogeneous group of patients to check if there were any significant changes between those who have not presented any kind of complication, those that have been subjected to a second surgery for correction of velo-pharyngeal insufficiency, and those undergoing minor surgery for other complications.

The median age in months for soft palate closure or full closure of the palate was

analyzed for patients operated only once. The Mann-Whitney test for indipendent variables was used to compare the group of patients who did not have complications with the group of those who underwent secondary surgery for palatal fistula before (Chart 10.12) and velo-pharyngeal insufficiency later (Chart 10.13).

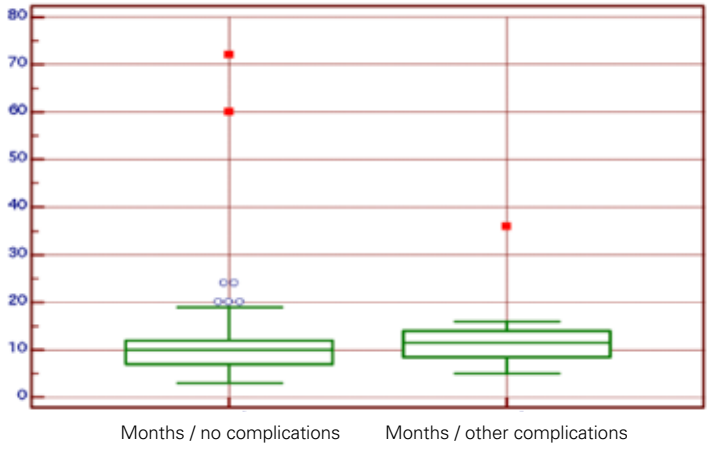


Chart 10.12 Comparison of median age at operation in the group of patients who did not have complications, left (median 10.0 months) and reoperated group for complications (palatal fistula) right (median 11.5 months): P = 0.48, NOT SIGNIFICANT.

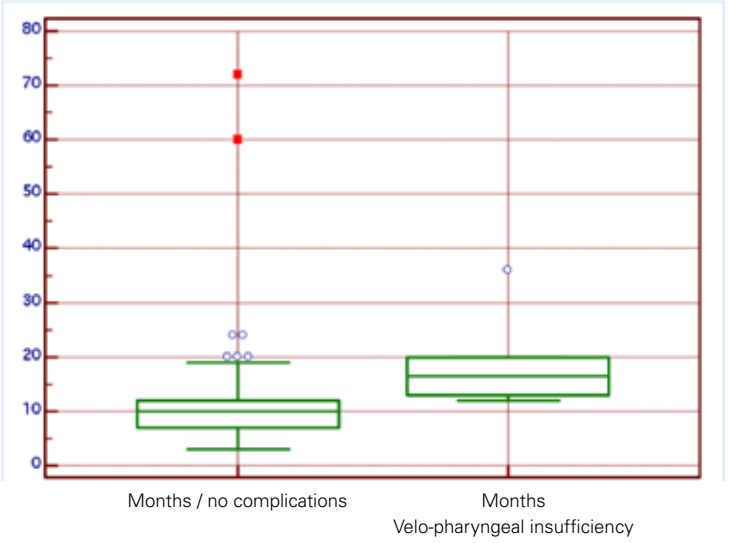


Chart 10.13 Comparison of median age at operation in the group of patients who did not have complications, left (median 10.0 months) and in the group that underwent secondary surgery to velo-pharyngeal insufficiency right (median 16.5 months): P = 0.05, SIGNIFICANT.

The difference between the medians was significant (P 0,05) only in the second comparison with mean age at the primary palatoplasty significantly higher in patients who later needed velo-pharyngeal insufficiency surgical correction.

The three groups were then analyzed according to the surgical technique used by chi-square test and there was no statistically significant difference, although the complication rate is higher in patients undergoing correction by Widmayer Perko and Von Langenbeck technique.

No difference in incidence of major complications was seen in patients with craniofacial malformations associated with cleft palate (Pierre-Robin, DiGeorge, Treacher-Collins).

Among patients who needed revision of the Labioplasty most had undergone primary surgery with the Millard technique, the technique which is most practised in all cleft lip patients.

Section 2

Surgical partnership. Cleft care in Low and Middle Income Countries

1.
General considerations

When considering the needs and health priorities in developing countries, congenital malformations appear low on the list, not only in terms of prevalence, but also because their treatment requires unsustainable human and financial resources and resources are orientated to large-scale major killers of these populations such as HIV, tuberculosis and malaria.

The weight in terms of DALYs (disability-adjusted life year) of cleft lip and palate is globally 657,000, or less than 1%, divided into 123,000 in the African region, 44,000 in the Americas, 240,000 in southeast Asia, 35,000 in the European region, 69,000 in the Eastern Mediterranean, 144,000 in the Western Pacific region (Global Health Estimates 2014, based on 2012 data from the World Health Organization report). One of the major projects for the treatment of cleft lip and palate was launched in Sri-Lanka, against 8,592,864 (0.3%) of DALYs attributable neural tube defects, which are more frequent congenital malformations. In order to understand the importance of Clefts compared to other health issues a good example is Africa where in 2012 nearly 67 million DALYs were due to HIV / AIDS compared to 123,000 due to cleft lip and palate.

Despite the low global burden that emerges from these statistics it is estimated that in India alone 35,000 children are born every year with cleft lip and that the figure of individuals not operated in the same country is said to be around one million (Nagarajan, 2006).

To treat the widespread communicable diseases that afflict countries with few resources means essentially to establish programmes of distribution of anti-microbial drugs. Dealing with the treatment of cleft lip and palate in these countries, if you do not want to fall into a mere "new humanitarian colonialism", as already reported, requires a broader vision of the pathology and consideration of some issues closely related to it.

Cleft lip and palate, like most congenital malformations, are primarily a source of disability and are also a stigma for who bear this hallmark to talk about it in polite terms. Whilst the rights of the disabled or handicapped and the definition of disability has already been extensively addressed in the Western world leading to the United Nations signing a convention on the rights of people with intellectual disabilities (Enable Convention on the Rights of Persons with Disabilities 2006), the same can not be said for developing countries. In many societies, a child born with a malformation, especially if it disfigures the face, is seen as a sign of the punishment of the gods to the mother or the community to which it belongs. The wrath of God or the influence of evil beings on the expectant mother of the baby is apparent in the birth of a creature to be considered "hybrid", neither man nor animal *(Mars M, Sell D, 2008)*, often to kept at the edge of the community because of fear.

From the modern view of disability sanctioned by the United Nations to the extreme mentioned above many variations exist. In both the Hindu and Islamic religions for example, the disability is seen as God's will and both the bearer or who is close to the patient, should accept it without shame and do everything possible to promote healing and care of those who suffer. It goes without saying that 8 out of 10 individuals belonging to these societies, faced with a congenital malformation, turn primarily to the religious leader of the community or to the "traditional healers" and only secondarily or if directed by the latter, to doctors practicing Western medicine.

Team care in Low and Middle Income Countries

To emphasize the complexity of the problem is the recognition, relatively recently (from 2000 onwards), of the need to encourage, even in countries with limited resources, an interdisciplinary management of cleft lip and palate. This team will take charge of long-term follow-up not only of surgical problems but also those related to the development of language and hearing and orthodontic care. It is irresponsible to think that it is sufficient to ensure more surgical interventions and just the transfer of surgical skills. The few studies available on the long term outcome of patients operated as part of humanitarian missions have shown that surgical correction alone does not affect the articulation of language and in these contexts the acceptance by the community passes through the improvement of communication skills out of necessity.

In many countries with limited resources there are no speech therapy rehabilitation services, in some cases there may be a single expert speech therapist or speech pathologist for millions of people and they do not always have the specific

experience to care for patients with cleft lip and palate. Also the equipment needed to carry out tests and audiological screening between patients with conductive hearing loss linked to endotympanic exudates and sensorineural hearing loss or linked to ear malformations is expensive and not sustainable for most of the health facilities.

To cope with this series of barriers different approaches have been attempted over the past 20 years. In China for example maxillofacial surgeons and nurses who also take care of phoniatric assessment and speech therapy have been trained (WHO Report 2002). The Sri Lankan programme Cleft Lip and Palate Project has instead initially planned the training of local personnel in the principles of the phono-articulatory therapy. Problems experienced in the latter case were represented mainly by the lack of an institutional and economic recognition by the local social health system of these figures who were therefore not motivated to continue their efforts. In addition, the training periods were too limited in time to make local staff autonomous in the management of all the problems associated with cleft palate.

In the wake of this trial and error approach and in order to meet a constantly growing need in a sustainable way a proposal created by the United Nations Development Programme in the 90s, the Rehabilitation System based on the Community (Community-based rehabilitation, CBR) was created. It is a strategy of common sense that aims to improve the quality of life of persons with disabilities through the involvement and empowerment of themselves and their families and the usage of resources that are realistic, reproducible and sustainable in all countries. The system must be supported at several levels: that of the Community, with the mobilization of local resources and technologies and support from the community to the families as regards the basic needs and protection of human rights; the intermediate level, through the creation of a network of services which can capture all persons with disabilities who require more specialized interventions than those provided by the community; through the national planning policies, implementation, coordination and evaluation of the CBR system *(Helander E, 1993).*

In the specific case of speech therapy rehabilitation there are already pilot projects in which members of a community in South India were instructed to identify and select children with communication errors suffering from cleft palate. Prathanee et al in 2006 designed workshops aimed at developing a basic model of speech therapy rehabilitation that was community-based. Projects can be very different, what is important is the involvement of participants in the CBR system, where this has already been developed, in all cooperation programmes concerning the treatment of cleft lip and palate.

2.
Surgery and Anaesthesia

In countries where most of the already limited financial and human resource is devoted to endemic diseases that endanger survival, an obvious discrepancy between patients with cleft lip and professionals occurs. The result is that the majority of patients will not have access to adequate care and will have to live with an uncorrected defect or one corrected by inexperienced hands.

This discrepancy obviously attracts missions of qualified surgeons from countries with high resources, but because of its educational potential can unfortunately attract younger surgeons still in training wanting to gain experience on less fortunate patients, who by nature are content with little. This bad practice, which is fortunately not universal, is encouraged by the fact that operated patients, if not clearly instructed to do so, are unlikely to return for a long-term evaluation of outcome and, even when they do, they tend to be uncritical.

Very negative effects are also created by missions with teams that bypass local surgeons and staff and leave out all types of training. However, most humanitarian organisations now work towards creating co-operation and training of the local community as one of their primary aims.

The participation of local surgeons with the visiting team is absolutely essential. Local surgeons should participate in the work so acquiring the skills to assume the role of first operator and work towards self-sufficient medical communities and medical structures.

Ideally, surgical teams should follow the highest medical standards to "guarantee that the quality of treatment and the safety standards are as high as in any other surgical unit in countries with better resources" *(Ward & James, 1990).*

Aspects of high quality care for cleft lip and palate that are largely uncontested include:

1. Comprehensive and multidisciplinary treatment;
2. Experienced health-care providers.

At attempt should be made to establish centres that resemble as closely as possible those in the Developed World. At these centres, longitudinal, multidisciplinary care of the child should be done.

Those who decide to take care of cleft lip and palate in the developing countries must be able to adapt to local practices, including surgical equipment and anaesthetic technique, without taking anything for granted.

The operating programme must be based on the type of hospital and the presence or absence of services such as a blood bank and intensive care, but above all patient safety must always be the priority.

Objectives to be taken into consideration are also:

1. Do not leave fistulas in order to reduce drastically the need for secondary interventions;
2. To ensure an improvement of the articulatory ability to facilitate insertion in the patient community.

The scheduled mission, usually concentrated in a period of 2-3 weeks should have a first phase for the proper preparation of patients.

Ideally for ensuring patient safety and quality of care the hospital must have:

- Operating theatre with operating table, operating lamp, surgical equipment, anaesthesia equipment, autoclaves and sterilizers;
- Post surgical and emergency room;
- Post operative ward.

The selection of patients

Every patient undergoing cleft surgery must have a complete history and be examined by a pediatrician or general practitioner, familiar with the common health problems of the locality in which the surgical mission is situated.

- The history and physical exam should include basic lab work to rule out anaemia, respiratory or urinary tract infection and the presence of associated malformations especially cardiac ones. Severely underweight children should be examined for gastrointestinal parasites and treated preoperatively if possible. Consideration should be given to preoperative malaria screening and prophylaxis in endemic areas.
- All patients undergoing surgeries must qualify for American Society of Anaesthesiology (ASA) class 1 or class 2. ASA class 1 patients have no organic, physiologic, biochemical, or psychiatric disturbance and the pathologic process for which the operation is to be performed is localized and does not entail a systemic disturbance. ASA class 2 patients are those with mild to moderate systemic disturbance caused either by the condition to be treated

surgically or by other pathophysiologic processes, including the otherwise healthy child with cleft lip or palate.

The aim is to select only those patients who are not at high risk of intra and post-operative complications and so do not need intensive care facilities as these are often not available. In the wards of many hospitals visited by this kind of missions the same intravenous infusion of liquids or analgesics can pose a risk if there is no experience with pediatric patients and care is largely entrusted to parents. Unlike in high resource countries where operated children are kept fasting for a few days, rapid oral rehydration is encouraged even with breast milk if available.

For the same reason the administration of opioids or other drugs that may induce respiratory depression should be avoided. If used intraoperatively, it is appropriate to keep the patient under observation until they are fully awake before returning him to the ward.

The preoperative tests should have at least a CBC exam for all children and screening for malaria in those with fever. In the absence of a laboratory it is possible to use portable instruments such as Hemo Cue for the evaluation of the haemoglobin concentration in blood.

Anaesthesia

Anaesthesia must be provided by an anaesthesiologist with experience caring for small children using anaesthesia machines and carbon dioxide monitors or having, at a minimum:

- Vaporizers for Halothane;
- A functioning oxygen supply;
- A sufficient drug formulary including antibiotics, I.V. hypnotics, I.V. and oral analgesics, muscle relaxants and emergency drugs;
- Using pulse oximeters, appropriately sized for children, during surgery and having appropriately sized blood pressure cuffs and precordial stethoscopes;
- Having and using other anaesthesia equipment (including endotracheal tubing, IV catheters and tubing, oral airways, masks, laryngoscopes and laryngoscopic blades, stylettes, circuits, suction catheters, disposable needles and syringes) sized appropriately for the age of the child;
- Recording the details (heart rate, blood pressure, ventilatory data, agents).

To reduce the number of general anaesthetics, adults and older children able to cooperate which require only the lip repair, they can be treated under sedation

(local anaesthesia with lidocaine + epinephrine 1: 200,000, 7 mg/kg or lidocaine alone 3 mg/kg bupivacaine or 3 mg/kg + midazolam or ketamine).

Operation Smile has published a report on 5000 general anaesthetics carried out between the years 1998-99, denouncing a high incidence of complications, especially in younger children. Mortality (4 deaths in more than 9,000 interventions) was entirely attributable to preventable causes: co-morbidity not seen at diagnosis and anamnesis, depletion of oxygen stocks, the absence of intra and post-operative monitoring.

Therefore, even if the surgeon approves surgery, the anaesthetist and possibly the pediatrician have the last word on patient operability.

Surgery

The functional goals of cleft palate surgery are to facilitate normal speech and hearing without significantly affecting the facial growth of the child.

To reach these aims the surgeon has to partially modify his surgical approach:

- Cleft palate repair is more important from a functional stand point: surgical restoration of all components of an abnormal velopharyngeal mechanism at an early age increases the patient's chances of developing normal speech and hearing;
- Single-stage lip and palate repair avoids the risk of failure to complete palatoplasty after lip repair that may be attributable to cost as well as concerns by parents about aesthetic problems: there is a high percentage of drop-outs in follow-up surgery.

Surgeons in developing countries modify the timing of their repairs to improve results.

There are indications that early repair of both lip and palate in one surgical session is not inferior in outcome to the traditional staged procedure, for this reason it is considered appropriate that all children in good health, over six months and who have no easy access to a secondary surgery receive simultaneous correction of cleft lip and cleft palate *(Onah I.I.* et al.*, 2008)*.

Surgery must be a part of an ongoing surgical programme, and performed by surgeons who are trained and have experience in surgery for cleft lip and palate.

In missions limited to 2 weeks, many pre-surgical procedures such as lip-taping, lip-adhesions, the use of palatine or nasoalveolar molding plaques to facilitate the combination of bone segments, and post-surgical removal of stitches not absorbable or separation of lingual flaps prepared for closing palatine fistulas (usually done at 15 days after primary surgery) is not practicable without an adequatly trained local surgeon.

The surgical technique recommended for the closure of the cleft lip is the one described by Millard as it ensures adequate correction of nasal deformities. The closure of the hard palate with vomer flap in primary surgery as suggested by the Oslo group and Lehman reduces the need for secondary intervention. The correction of the bilateral cleft can present further difficulties because the presence of a pre-maxilla not properly repositioned can prevent the primary Labioplasty. The most used techniques are those described by Millard and Mulliken, in this case avoiding the simultaneous closure of the front palate to prevent undermining the integrity of the pre-maxilla vascular intake.

The first choice for the closure of the palate instead is the closure in a single operation by means of a two flap technique due to its reproducibility and safety, because it is easy to teach and applicable to almost any type of cleft. The Furlow technique also receives widespread support but is less easy to learn and then teach and can not be used in a single operation for wide cleft.

One stage Cleft lip and palate repair is a therapeutic option that can find an indication in patients in poor countries with limited health resources, where they are taken into account factors not only related to the development of the facial skeleton but which also take account of economic and social situations cultural.

The components which justify this option may be summarized in:

- A moderate or minimum extent of the defect;
- A poor patient motivation for surgery;
- Satisfactory possibilities to a postoperative intensive surveillance;
- A state of good health.

If closing the cleft palate in a single procedure is contraindicated, many families consider aesthetic repair to enable social acceptance. The defect in the palate and labial and alveolar deformities are repaired in subsequent operations.

In unoperated patients with cleft palate and more than 15 years old, the speech articulation remains very unsatisfactory performing only palatoplasty and many authors suggest that an associate of emblée pharyngoplasty thus improving the competence of the sphincter velo-pharyngeal.

The incidence of fistulas after palatoplasty can reach 40% and the failure of the secondary closure of fistulas varies between 12 and 65%. Hence some principles to be observed to increase the percentage of success are:

1. Dissection of wide palatine flaps;
2. Excision of all scar tissue present at the edge of the fistula;

3. Non-voltage double-layer of the nasal and oral mucosa of the planes;
4. Use for the front flaws additional tissue by wrapping buccal mucosal flaps or lingual;
5. Any bone graft.

Secondary surgery such as bone graft to close the alveolar defect or the correction of maxillary retrusion is conceivable and programmable only in the presence of a good orthodontic service that ensures the management of pre- and post-surgical care *(Mars M, Sell D, 2008).*

In a ideal foreign mission the staff must foresse:

- Having experienced operating room personnel;
- Sufficient quantities of instruments and sutures, with type and quantity appropriate for the planned procedure;
- Having on-site a suctioning machine;
- Electrocautery machine and equipment;
- Adequate lighting for illumination of the surgical field;
- Equipment for proper sterilization of surgical equipment;
- Preventing transmission of blood borne pathogens;
- Post-Surgical and Emergency Care;
- Safe post-anaesthesia care, *by:*
  1. Having a policy and procedure that anaesthesiologists extubate patients when they are awake enough to have a return of normal upper airway reflexes.
  2. Having a surgeon immediately available in the operating room suite until the patient is breathing spontaneously, is extubated, and has a clear airway.
  3. Having a designated unit for post-anaesthesia care which is adjacent to or in the OR suite.
  4. Having a clearly delineated medical chain of command, communication and responsibility for care of children in the first 24 hours after cleft surgery. This includes the ready availability of a physician capable of treating any complications that might occur.
  5. Having and using pulse oximeters (again, appropriately sized for children) to monitor post-anaesthesia care patients.
  6. Staffing the post-anaesthesia care unit with clinical staff with training in recovery care and who have post-anaesthesia care as a regular part of their job. The training in recovery care must include how to recognize hypo/hypertension, airway obstruction, respiratory depression and hypoxemia as detected by a pulse oximeter.

7. Having sufficient numbers of skilled post-anaesthesia staff that individualized observation is possible the first night after surgery.
8. Specifically, all patients in the recovery area must be monitored by a nurse until they are fully awake and crying and all patients must be assessed at regular, frequent intervals for post-operative bleeding.

- Be able to intervene to provide intensive care if a patient requires it, *by:*
  1. Having written protocols in place and known by the staff for emergency care, triage, CPR, and blood transfusions.
  2. Having on-site and immediately available a suctioning machine, resuscitative medicines, an oxygen delivery system and oxygen supply, an ECG and blood pressure monitors, and resuscitation equipment.
  3. Having the ability to intubate children and support their breathing with mechanical ventilators and provide 24-hour monitoring by trained clinical staff; **or by**
  4. Having a current, functioning transfer agreement with a health care facility that can provide this type of intensive care.

Operating room surgical set up

The patient is positioned at the end (top edge) of the operatory table.

The head is extended gently by having the baby lengthwise on a pillow.

The head is over the pillow resting on the table close to its top edge.

The general anaesthetic is given through an intratracheal tube in the mouth, a nasal tube would distort both nostril and lip in the mouth.

The buccal tube has no such effect and does not interfere with the operation if kept in the midline and brought over the chin.

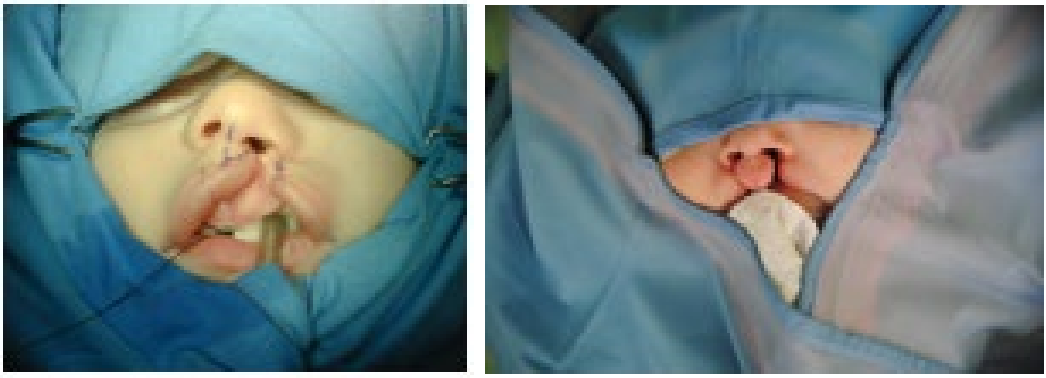


Figure 2.1 Position of sterile drapes.

The skin is cleansed with chlorhexidine 0.05% aqueous solution.

A mouth gag can be placed in case of palatoplasty.

A

B

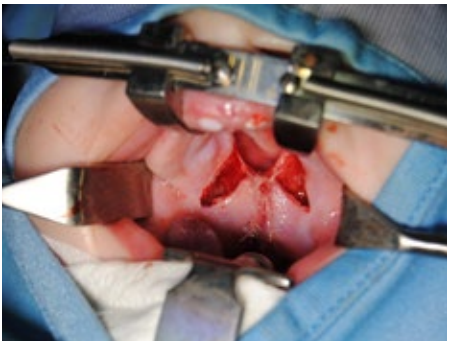


Figure 2.2 (A) Position of the patient on the operating table with hyper-extension of the neck and slight Trendelemburg. (B) Mouth gag, Digmann type.

Then is important to clean inside the mouth and to remove the mucus from the nostrils with damp ribbon.

Two sterile towels are laid underneath the head; the sides of the towel are wrapped around to meet on the forehead in a V, where they are secured with a clip.

Leaving the eyes exposed is probably less dangerous than having them covered

The surgeon sits at the head, but he must be able to get up during the operation and inspects his work from other angles.

A small gauze or a small pack of ribbon wet gauze is inserted in the pharynx to prevent aspiration of blood.

A pack of dry gauzes can become abrasive.

Only with magnification we can have a true picture of how the lip, nose and palate are evolving.

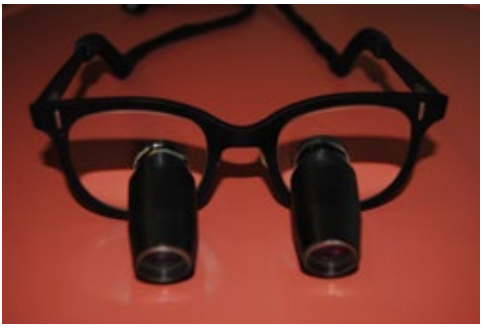


Figure 2.3 Magnifying surgical loupes.

The assistant works on the other side and keeps the lip and nose free of blood with gauze and aspirator.

Suturing

The closure of a cleft can depend from the quality of suturing. If edges are not in apposition healing will be retarded, but if they lie nearly together the process is rapid in the infant and the line of closure may become nearly invisible.

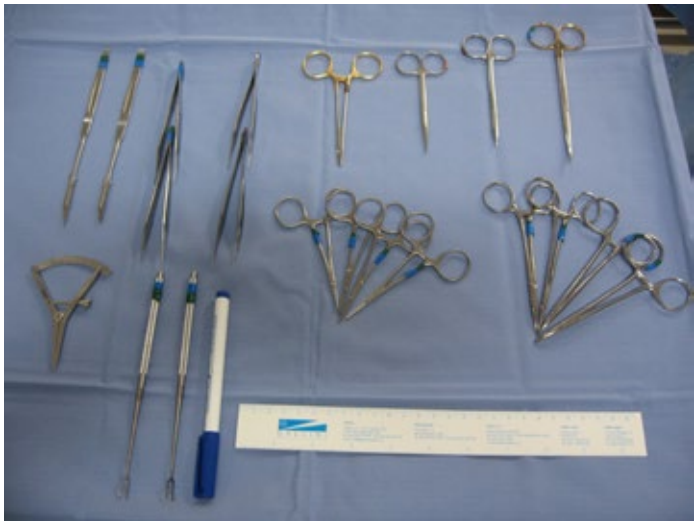


Figure 2.4 Cleft lip instruments.

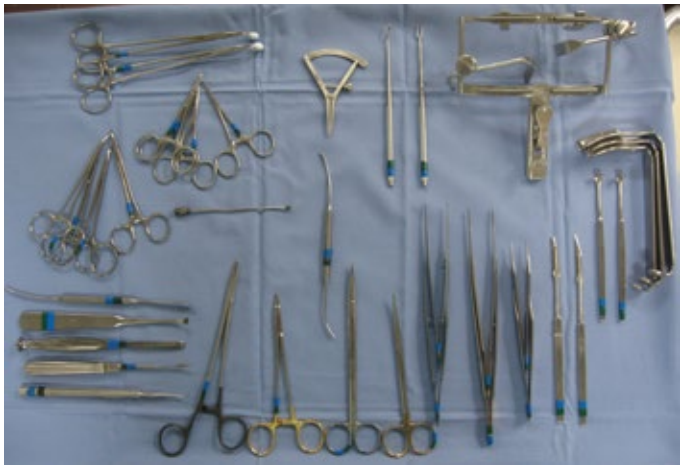


Figure 2.5 Cleft palate instruments.

Materials

	Suture	*Needle*
Soft palate mucosa	Absorbable polyglactin 910 4.0	*13 mm, 3/8 c*
Hard palate fibromucosa	Absorbable polyglactin 910 4.0	*Compound curve reverse cutting*
Muscle	Absorbable polyglactin 910 4.0	*17 mm, ½ c*
Lip vermillion/skin	Polyglycolic acid, braided, coated, fast absorbable 5.0 or 6.0	*11 mm, 3/8 c, cutting*
	Polypropilene, monofil, non absorbable 5.0	*13 mm, 3/8 c*

Chart 2.6 Suture material utilised in cleft lip and palate closure.

Technique

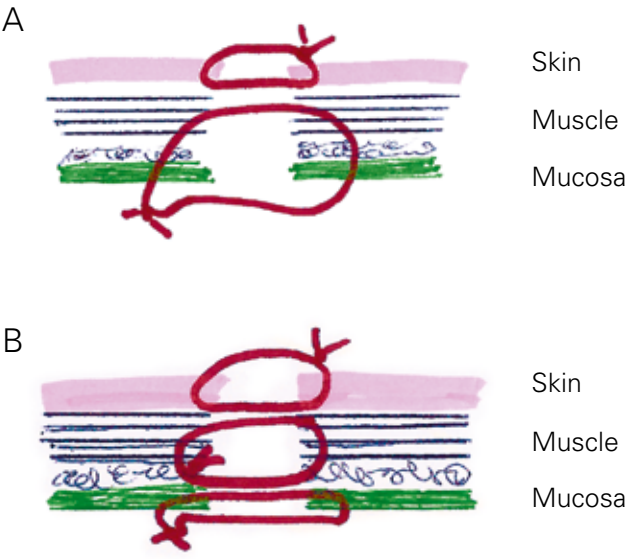


Figure 2.7 Method of suturing the lip. (A) Muscle closed along with the mucosa. (B) Muscle closed as a separate layer.

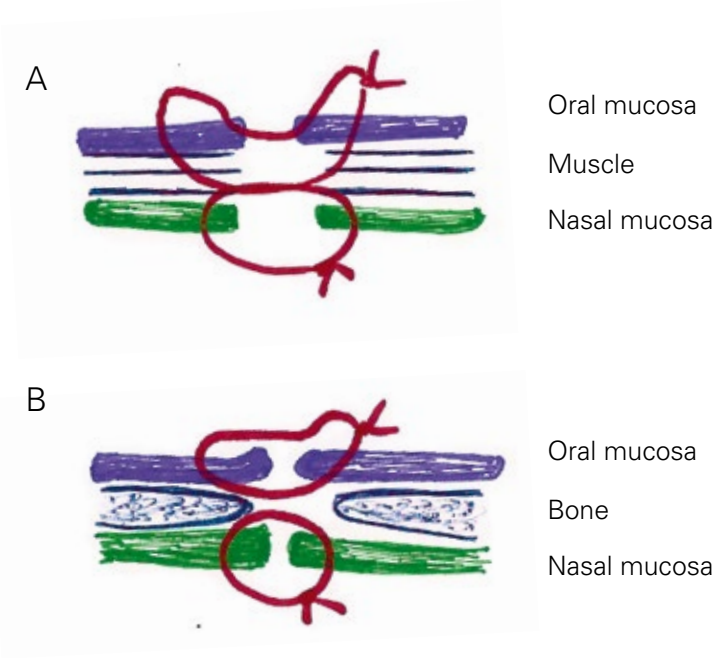


Figure 2.8 Method of suturing the palate. (A) Soft palate. (B) Hard palate.

Postoperative

- Every patient admitted to the postoperative ward will have vital signs monitored (blood pressure, respiratory rate, heart rate, oxygen saturation and temperature).
- An area on the postoperative ward is designated and equipped for resuscitation including emergency drugs.
- Suction equipment and oxygen must be available on the postoperative ward.
- 24-hour nursing care must be provided on the postoperative ward.
- Deliver optimal pain management through proper assessment and application of pharmacology.
- Postoperative patient education programmes will be administered by nursing staff and delivered in the local language with written instructions using words and pictographs.
- Comprehensive discharge instructions will be administered to patients/family, including medications and feeding.

3.

The treatment of cleft lip and palate in Low and Middle Income Countries: 20 years of experience through humanitarian missions

The Pediatric Surgery Department of the University Hospital of Ferrara has been involved since 1995 in many humanitarian missions. The "Chirurgo e Bambino Onlus -Surgeon for Children" a non-profit organization, was founded with the intent to form projects for medical aid and to create a sustainable pediatric surgery in poor countries. Sustainable pediatric surgery means that the visiting team should teach more than treat, training local surgeons, nurses and paramedical personnel the standards of pediatric surgery. Training local medical personnel empowers the community to be self sufficient and care for their own children. The Association has also established close cooperation with charities dedicated specifically to the treatment of children with surgical diseases, in particular these with oral cleft.

The following data were extracted mainly from the Reports of the Missions.

1. The first missions were carried out between 1995 and 2002 in Guatemala in Antigua, in close cooperation with WOPSEC (World Organization of Pediatric Surgeons for Emerging Countries) at the institute for the disabled poor people "Las Obras Sociales Hermano Pedro", where we worked together with local staff, doctor (a general surgeon) and paramedics (scrub nurses and support staff) to make operational and autonomous a centre specialized in the correction of cleft lip and palate.

Guatemala 1995-2002

Total number of surgeries: 406
Operations for cleft lip and palate: 46%

Figure 3.1 Antigua, Guatemala, Obras Sociales dell'Hermanno Pedro, a child with Left cleft lip and palate.

2. In 2003 our group began travelling to Kenya in order to build a sustainable pediatric surgery unit at the newly founded hospital of Matiri Taraka. After only five years severe economic problems have led to a significant reduction of the hospital activities interrupting our training project.

Kenya 2003-2008
Matiri, Tharaka

Total number of surgeries: 292
Operations for cleft lip and palate: 16%

3. In collaboration with Operation Smile Italia, two humanitarian missions were undertaken:

Russia 2005
Novosibirsk

Total number of surgeries: 62
Operations for cleft lip and palate: 79%

- 6 unilateral labioplasties
- 17 palatoplasties
- 15 revisions of labioplasty
- 11 fistula closure

Armenia 2006
Yerevan

Total number of surgeries: 25
Operations for cleft lip and palate: 100%
- 8 unilateral labioplasties
- 1 bilateral labioplasty
- 3 palatoplasties
- 6 revisions of labioplasty
- 2 revisions of palatoplasty
- 4 fistulas closure

4. In collaboration with Smile Train Italia the following missions were carried out:

Georgia 2008
Tbilisi

Total number of surgeries: 28
Operations for cleft lip and palate: 100%
- 3 unilateral labioplasties
- 3 palatoplasties
- 10 revisions of labioplasty
- 2 revisions of palatopalsty
- 10 nasal deformity corrections

Tanzania 2008-2009
Mbweni

Total number of surgeries: 111 (2008: 70, 2009: 41)
Operations for cleft lip and palate:

97%
- 22 unilateral labioplasties
- 2 bilateral labioplasties
- 4 palatoplasties
- 16 nasal deformity corrections
- 15 revisions of labioplasty

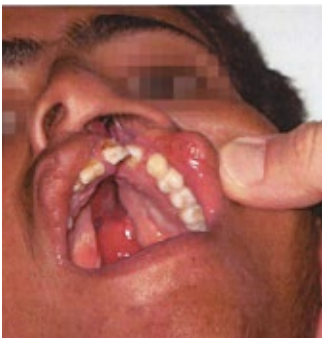
98%
- 13 unilateral labioplasties
- 1 bilateral labioplasties
- 6 palatoplasties
- 8 nasal deformity corrections
- 2 revisions of labioplasties

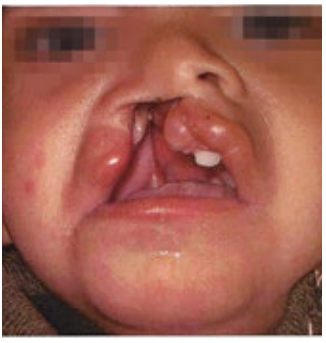
Yemen 2008-2009
Sana'a

Total number of surgeries: 19 (2008: 16, 2009: 3)
Operations for cleft lip and palate:

100%
- 5 unilateral labioplasties
- 1 bilateral labioplasty
- 6 palatoplasties
- 2 revisions of labioplasty
- 1 revisions of palatoplasty
- 4 fistulas closure

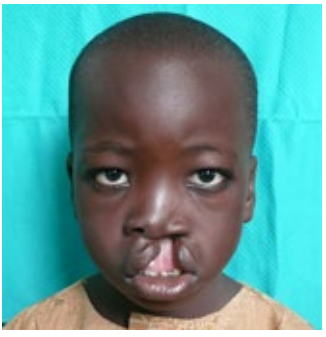
100%
- 1 bilateral labioplasty
- 2 palatoplasties

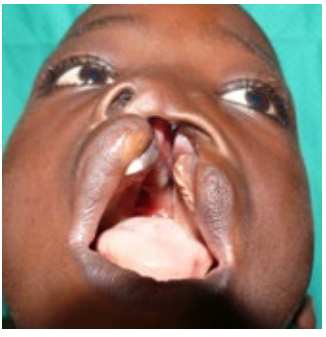




Ivory Coast 2009
Abidjan

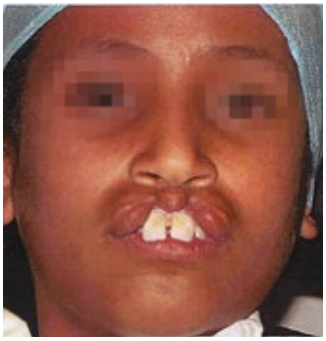
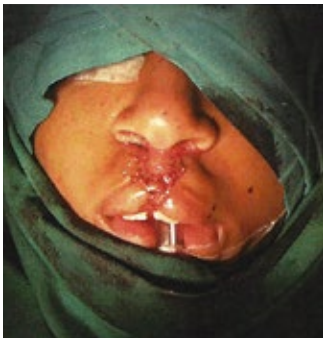
Total number of surgeries: 75
Operations for cleft lip and palate: 88%
- 21 unilateral labioplasties
- 5 bilateral labioplasty
- 11 palatoplasties
- 8 alveoloplasties
- 14 nasal deformity corrections
- 1 columella lenghtening
- 3 revision of labioplasty
- 3 fistulas closure





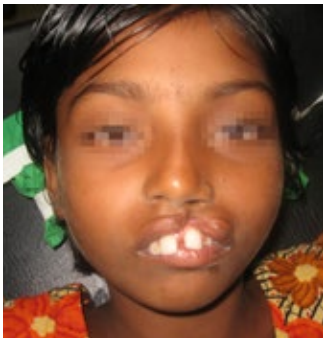
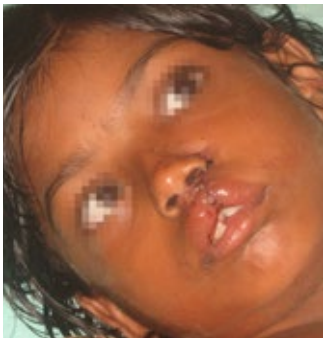
Ethiopia 2009
Makallé

Total number of surgeries: 62
Operations for cleft lip and palate: 97%
- 18 unilateral labioplasties
- 5 bilateral labioplasty
- 9 palatoplasties
- 6 nasal deformity corrections
- 1 columella lenghtening
- 1 fistulas closure

Bangladesh 2010
Gaibanda
Emirates Friendship Boat

Total number of surgeries: 53 in first mission / 23 in second mission
Operations for cleft lip and palate: 100%
- 17 unilateral labioplasties
- 3 bilateral labioplasties
- 3 palatoplasties

Benin 2011
Cotonou

Total number of surgeries: 18
Operations for cleft lip and palate: 100%
- 10 unilateral labioplasties
- 2 bilateral labioplasties
- 4 palatoplasties
- 1 revisions of labioplasty
- 1 revisions of palatoplasty

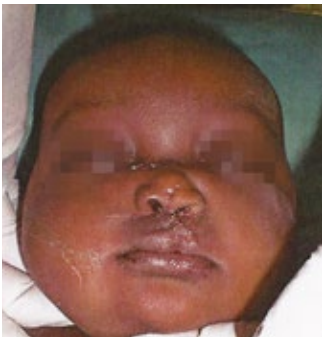
Iraq 2011
Mittica

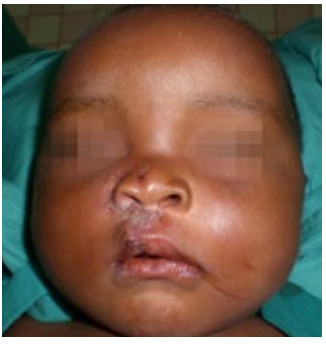
Total number of surgeries: 151
Operations for cleft lip and palate: 90%
- 43 unilateral labioplasties
- 4 bilateral labioplasties
- 62 palatoplasties
- 30 revisions of labioplasty
- 10 revisions of palatoplasty
- 11 nasal deformity corrections
- 11 alveolar fistula correction

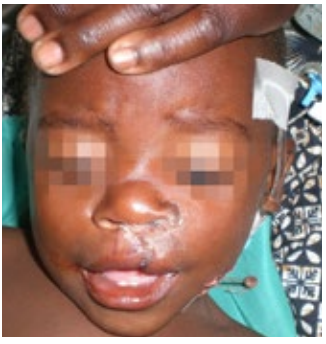
5. A Pediatric surgery teaching programme in 2010 started at Mbweni Hospital Kinondoni District and at St.Gaspar Hospital Itigi,Singhida,in close cooperation with Ruvuma Onlus, St. Joseph University and Ospedale Bambino Gesù Roma.

Tanzania 2010-2018
Mbweni
Itigi

Total number of surgeries: 255
Operations for cleft lip and palate: 15%

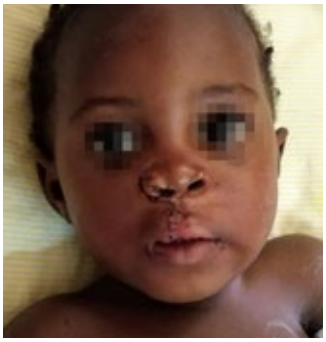






Guinea Bissau 2017
Bissau

Total number of surgeries: 33
Operations for cleft lip and palate: 28%
- 6 unilateral cleft lip
- 7 cleft palate



Percentage distribution of indications for surgery in the patients with cleft lip and palate during our humanitarian missions

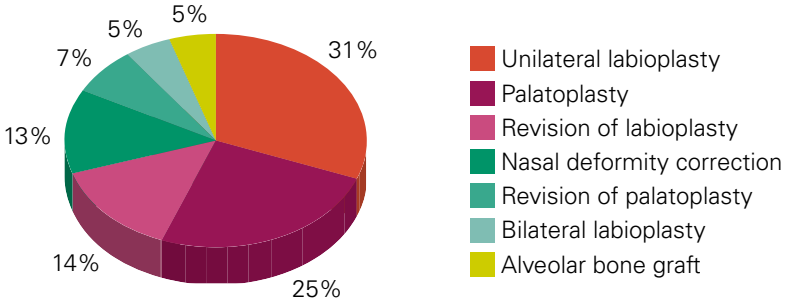


Chart 3.2 Indications for surgery 1995-2015.

The same surgical correction procedures used in our Department in Italy were used in all cases. However the age of the patients operated was higher.

Age distribution of patients at operation (percentage)

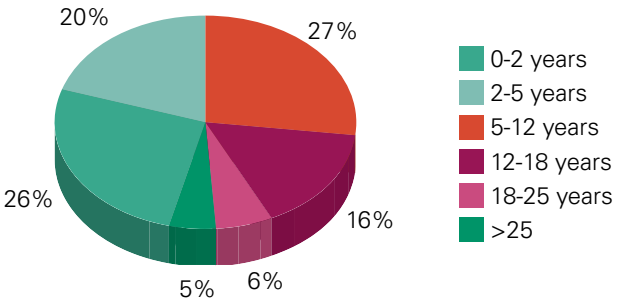


Chart 3.3 Age of presentation of the patients 1995-2015.

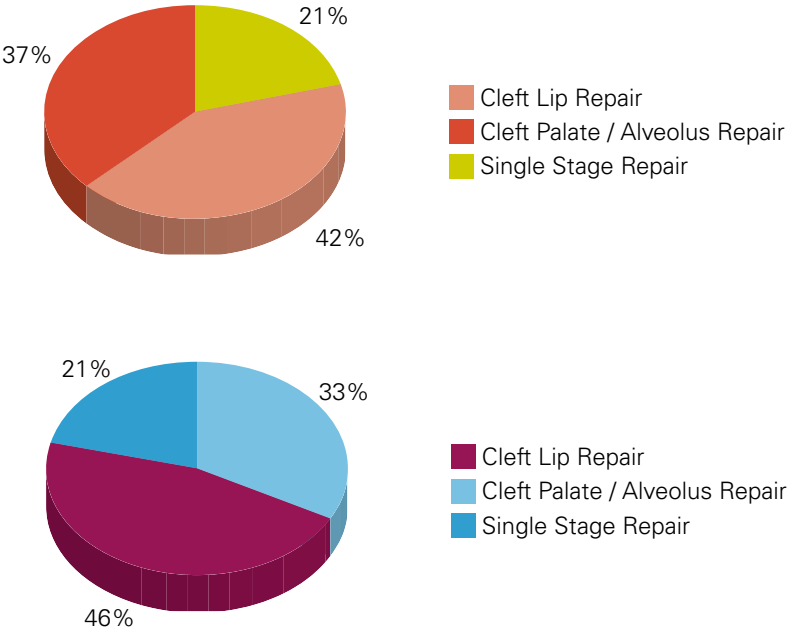


Chart 3.4 Tendency to prefer one stage cleft lip and palate repair by years.
(A) 1995-2005. (B) 2005-2015.

The late presentation for care in children and adults observed in our experience led us to apply in selected cases a single stage tecnique for complete cleft lip and palate repair (Chart 3.4).

Comparing the first and second period of the twenty years of experience in the repair of clefts a one stage approach requires:

- A moderate or minimal extension of the defect;
- A poor motivation of the parents or of the patient;
- An adequate post operative care;
- A good general health status.

The Authors thank Dr. F.M. Abenavoli, President of the NGO Doctors for smiling children, Dr. R. Rodriquez, President and Dr. G. Travaglini, Scientific Director of the Ruvuma Onlus Association and Dr. D. Cumba Pediatric Surgeon of the Bor Hospital in Guinea Bissau for their fundamental contribution to the implementation of the above-mentioned humanitarian missions.

Concluding remarks

One of the biggest studies concerning the treatment of cleft lip and palate, Eurocleft (1996-2000), was born with the intention to overcome the major limitations of the analysis carried out on outcome in a single centre. These are multifactorial in origin combining the difficulty of comparing within the same group different protocols of differing types. In the case of our centre the tendency is to adopt one with only a few variations on the theme. Hence it is difficult to distinguish between good outcomes resulting from a good protocol or a surgeon with good experience. There remains also the numerical limit linked to the large variability of the malformation presentation, to the knowledge of the operator as to how to act in different ways depending on, for example, whether mono or bilateral form, complete or incomplete. It becomes therefore impossible to study large homogeneous groups. This difficulty also exists in creating prospective randomized studies, leaving many issues unresolved *(Shaw WC, Brattstromm V, 2005)*.

In addition to the above limits, the great difficulty is to obtain comprehensive data on the follow up of patients. Many of them decide to suspend check ups or continue them in another place distant from the centre in which the primary treatment has been carried out. Incompleteness of such data requires the results obtained to be considered carefully.

The Eurocleft research has revealed the coexistence of 194 different protocols at 201 centres in Europe, only designed for the correction of unilateral forms of cleft palate *(Shaw WC, Semb G, 2001),* other surveys have shown that the age for correction of cleft soft palate varies from 3 to 24 months while for the hard palate from 6 months to 16 years *(Berkowitz S, 2013)*.

Despite the awareness of the limitations of a retrospective analysis covering patients in a single centre and far from wanting to add something new to the still unresolved debate about what appears to be the best protocol, the present study

has the advantage that almost all patients were treated by a single operator, to eliminate at least one of the variables in play. Its extension in time has allowed, when available, follow up of long duration to be examined.

The propensity towards an early palate repair gave good phonetic outcome, now a concept given for granted in the literature *(Berkowitz S, 2013),* confirmed in our case by a relatively low incidence of post-surgical velo-pharyngeal insufficiency and an average age at palatoplasty significantly higher in patients that then underwent secondary surgery for velo-pharyngeal insufficiency.

The first choice of protocol adopted for complete cleft palate has been the palatoplasty in 2 stages with closing relatively early the posterior palate as also indicated by the Swedish Gothenburg Protocol as long as this can be obtained with minimally invasive surgery or leaving the least amount of hard palate with denuded areas. The main motivation to support this choice was the finding of a partial spontaneous reduction of the anterior defect that has allowed in 2/3 of patients to perform a closure without large mobilization of the flaps.

Despite the early closure of the hard palate there were no large discrepancies of the facial bones. No patients among those who followed the follow-up required a maxillary distraction.

No significant differences were recorded in terms of complications between the different techniques used in palatoplasty nor between patients treated in two stages compared to those treated in a single intervention.

The relative homogeneity of complications in patients treated with different techniques and protocols gives an account of the importance that individual factors and the operator's experience play, together with the Protocol adopted, in determining the outcome.

The incidence of associated anomalies, laterality and the distribution of the individual variants coincide with those reported in the literature except for the prevalence of patients treated in the series of isolated cleft of the palate with respect to the forms associated with cleft lip. That finding is not easily explainable based on the data in our possession.

It is well established that the gold standard for treating the complexity of the patient with orofacial cleft is the establishment of a multidisciplinary team that includes at least one surgeon (of any specialisation provided he/she has extensive experience in the treatment of cleft lip and palate), an orthodontist, a speech therapist and a Coordinator. The Department of Pediatric Surgery of Ferrara for this type of patient use a interdisciplinary team in which several specialists will individually deal with problems related to their discipline and formally communicate through medical reports or sometimes in person or by telephone their feedback

in order to take a joint decision if it is necessary to continue the therapeutic procedure.

More effective is the current trend, especially in centres dedicated more specifically to the treatment of these malformations, towards the creation of an interdisciplinary model in which individual specialists acquire some knowledge of the management of other problems afflicting the patient outside their specialisation and informally discuss with other members of the team through regular meetings. The latter approach seems to give better results with respect to both the patient outcome and job satisfaction of individual members *(Berkowitz S, 2013)*.

Other figures that should be part of the team are the Psychologist and Psychiatrist, Audiologist, Geneticist, Otolaryngologist and Pediatrician. The follow-up proposed by our Department does not include a routine psychological assessment so it was not possible to consider this aspect in patients' outcome. Living with a malformation or with the results of this, especially when they involve a fundamental function such as language, involves adaptation of the social implications of school-age children, which would seem to make it still more evident in young adults in terms of low perceived quality of life compared to healthy subjects. Studies conducted to date have focused on school-aged children and adolescents but the few studies available on adult patients with cleft lip and palate reveal a history of delays in the emancipation from the family of origin, less inclination to marriage and motherhood or paternity. Current research studies are trying to identify possible relationships between the development of the facial skeleton and the brain.

Such studies are limited in that they consider only population groups from a country with High Socio-Economic Resources. Given the peculiarities already listed in section 2 (popular religious beliefs related to facial malformations, higher prevalence of adolescent patients or young adults not operated or operated belatedly), it would be interesting to extend these studies of psychosocial outcome to groups of patients treated in the Low Resource Countries.

For the same reasons in the planning of missions abroad among the top targets together with the surgical correction is the creation of a multi-disciplinary team able to meet on site all issues involving the patient especially the development or rehabilitation of language so the role of the foreign surgeon will become in time only one member of the team or a consultant for training.

Lessons from Low and Middle Income Countries

The increase of missions and projects for the treatment of cleft lip and palate in the developing countries on the one hand allows the discussion of critical issues

in correcting surgical pathologies in a manner different from how it has been done for decades in higher socio-economically developed countries and on the other hand it has revealed realities that no longer exist in the western world such as the large pool of adults or young adults with non operated cleft lip and palate, and worthy of accurate research. In particular this has been done in the context of the Sri Lankan Cleft Lip and Palate Project to evaluate the separate influence on the growth of the maxilla and the palatoplasty of the defect when this is not corrected early in life. The data were collected over 23 years in which the progression of facial growth of more than 500 patients from birth to maturity, through the skull X-rays and dental arch impressions acquired under strictly controlled conditions were recorded to ensure their quality and validity. In particular, 30 subjects suffering from not operated cleft, aged over 17 were compared with 52 healthy control subjects and this has highlighted the intrinsic influence of the malformation on the vertical development of the maxilla anteriorly and posteriorly, but not on the development of anteroposterior dimension and the bone relationships. It has not been observed in patients with maxillary retrusion *(Liao YF, Mars M, 2005)*. In addition 48 patients who underwent only repair for the cleft lip were compared with 23 non-operated peers and it was found the Labioplasty influences solely the anteroposterior and vertical position of the socket and incisor, not the jaw itself *(Liao YF, Mars M, 2005)*. Finally 48 patients undergoing palatoplasty alone were compared with 58 patients who underwent complete correction of the lip and palate and results showed that the correction of cleft palate inhibits slipping outwards and anteroposterior growth of the jaw and the alveolus *(Liao YF, Mars M, 2005)*.

Data stored thanks to this project were then analyzed using the Goslon Yardstick scale for the analysis of maxillary deformities.

What has emerged is that the majority of non-operated patients are part of Goslon group one and none can be attributed to Goslon group four or five. This contrasts with those operated in the first year of life where 70% are among groups 4 and 5. These data on patients undergoing correction correlate closely with those revealed by a study carried out in Britain in 1992 *(Mars M, Asher C, 1992)*.

These studies have a significant value but it must be emphasized that these patients are from areas where there are still infectious endemic diseases and malnutrition that can affect the results. It has been also widely recognized that growth can be hindered by the emotional and affective deprivation generated by social exclusion in which these young adults with craniofacial malformations often find themselves especially as cleft palate, can make their speech unintelligible cancelling their communication skills.

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Useful websites

http://store.enfamil.com

www.acpa-cpf.org (American Cleft Palate-Craniofacial Association)

www.bottlesandfeeders.respironics.com

www.eurocat-network.eu/content/eurocat-guide-1.4-section-3

www.medelabreastfeedingus.com

www.phgfoundation.org

www.silps.it

www.who.int/heathinfo/global_burden_disease/en/

Glossary

A

Adenoids: a normal collection of unencapsulated lymphoid tissue in the nasopharynx, also called pharyngeal tonsils.

Adenoidectomy: surgical removal of adenoid tissue in the nasopharynx.

Adenotonsillectomy: surgical removal of tonsils and adenoids.

Ala nasi: the lateral wall of each naris.

Alar base: the area where the ala meets the upper lip.

Alar rim: the part of the nose that surrounds the opening to the nostril on either side.

Alveolar bone graft procedure: a surgical procedure of grafting bone into the cleft site to stimulate new bone formation.

Alveolar ridge: the portion of the maxilla and mandible that surrounds and supports teeth.

Alveolus: the socket of the tooth.

Audiologist: specialist in evaluation and rehabilitation of communication disorders centre and hearing function.

B

Backing of phonemes: a compensatory articulation strategy characterized by the production of most phonemes with the back of the tongue and with the velum or with the posterior pharyngeal wall.

Base view: x-ray view visualizing the entire velopharyngeal sphincter during speech, as if looking up through the port, also called en face view.

Bifid uvula: a congenital split or cleft in the uvula.

Biofeedback: a training technique that enables a person to perceive body functions in order to voluntarily manipulate them by conscious mental control.

C

Cephalogram: a lateral radiograph of the craniofacial skeleton.

Choana: the opening into the nasopharynx of the nasal cavity on either side.

Circular pattern: a pattern of velopharyngeal closure, where the closure pattern resembles a true sphincter.

Class I occlusion: normal dental arch relationship where the mesiobuccal cusp of the first maxillary molar fits in the buccal groove of the first mandibular molar.

Class II malocclusion: abnormal dental arch relationship where the mesiobuccal cusp of the first maxillary molar is anterior to the buccal groove of the first mandibular molar.

Class III malocclusion: abnormal dental arch relationship where the mesiobuccal cusp of the first maxillary molar is posterior to the buccal grove of the first mandibular molar.

Cleft: an abnormal opening or fissure in an anatomical structure.

Cleft lip: a congenital facial defect of the lip due to failure of fusion of the medial and lateral nasal prominences and maxillary prominence.

Cleft palate: a congenital fissure in the medial line of the palate.

Columella: a small column at the lower portion of the nose that separates the two nostrils.

Compensatory errors: articulation gestures that are the individual's response to velopharyngeal dysfunction rather than the direct result of velopharyngeal dysfunction.

Complete cleft lip: a cleft condition that involves the entire lip through the nostril floor and the alveolus to the area of the incisive foramen.

Concha: part of outer ear similar to a shell in shape.

Congenital: existing at birth, referring to mental or physical traits, anomalies, malformations, or diseases, which may be either hereditary or due to an influence occurring during gestation up to the moment of birth.

Congenital palatal insufficiency: velopharyngeal dysfunction with no history of cleft palate or other known aetiology.

Coronal pattern: a pattern of velopharyngeal closure that is accomplished primarily by the posterior movement of the velum against a broad area of the posterior pharyngeal wall and the possible anterior movement of the posterior pharyngeal wall.

Craniofacial anomaly: a structural or functional abnormality that affects the cranium or face.

Cul-de-sac resonance: abnormal resonance perceived as muffled due to the resonating sound trapped in the vocal tract with no outlet.

Cupid's bow: the contour of the superior margin of the upper lip.

D

Dehiscence: a breakdown of sutured lines.

Denasality: abnormal resonance due to lack of vibration of the sound energy in the nasal cavity.

Distraction osteogenesis: a technique of inducing new bone formation by dividing a bone and applying tension through an external fixation device to lengthen the bone.

Dysphonia: any disorder of phonation affecting voice quality or ability to produce voice.

E

Eustachian tube: the tube connecting the middle ear with the nasopharynx, allowing middle ear aeration and drainage.

F

Fistula: an abnormal passage from one epithelialized surface to another, either congenital, caused by disease or injury, or created surgically.

G

Glossoptosis: downward displacement of the tongue.

Glottal stop: a compensatory articulation production characterized by forceful adduction of the vocal folds and the build-up and release of air pressure under the glottis.

H

Hard palate: the anterior part of the palate, consisting of the bony palate covered above by the mucous membrane of the floor of the nasal cavity and below by the mucoperiosteum of the roof of the mouth.

Hypernasality: a resonance disorder characterized by excessive resonance in the nasal cavities, often due to velopharyngeal dysfunction, particularly perceptible on vowel productions.

Hypodontia: missing teeth as a result of their failure to develop.

Hyponasality: insufficient nasal resonance during speech, usually due to obstruction of the nasal tract.

Hypoplasia: underdevelopment of a tissue or organ, usually due to a decrease in the number of cells.

I

Incisive foramen: a hole in the bone that is located in the alveolar ridge area of the maxillary arch.

Incisive suture lines: the suture lines that separate the premaxilla from the rest of the hard palate, which run between the lateral incisors and canines and meet posteriorly at the area of the incisive foramen.

Intraoral air pressure: a build-up of air pressure in the oral cavity that provides the force for the production of oral consonants.

Intravelar veloplasty: a surgical reconstruction of the levator veli palatini muscle sling during palatoplasty for correction of a cleft of the velum.

L

Lateral pharyngeal walls: the side walls of the throat.

Latham appliance: a two-piece acrylic dental appliance used prior to the lip and alveolus repair to close the gap between the greater and lesser maxillary segments from a wide cleft of the primary palate.

Levator veli palatini: paired muscle forming the main muscle mass of the velum, primarily responsible for velar elevation.

Lip adhesion: a simple, straight-line surgical procedure to temporally repair a cleft lip.

M

Malocclusion: any deviation from a physiologically acceptable contact of opposing dentitions.

Mandibular hypoplasia: lack of mandibular development, resulting in a small, retrusive mandible.

Maxillary hypoplasia: lack of maxillary development, resulting in midface retrusion.

Maxillary retrusion: a common anomaly in individuals with repaired cleft lip and palate secondary to the inherent deficiency in the maxilla due to the cleft and the possible restriction in maxillary growth with the surgical repair; typically characterized by a small maxilla relative to the mandible.

Micrognathia: abnormally small lower jaw, receding lower jaw.

Midface retrusion: concavity of the midface due to maxillary hypoplasia.

Mixed resonance: a combination of hypernasality, hyponasality, or cul-de-sac resonance during connected speech.

Mucoperiosteum: mucous membrane and periosteum so intimately united as practically to form a single membrane, as that covering the hard palate.

Mucous membrane: a mucous tissue lining various structures, consisting of epithelium and lamina propria, often called mucosa.

Multifactorial inheritance: inheritance involving many factors, of which at least one is genetic but none is of overwhelming importance, as in the causation of a disease by multiple genetic and environmental factors.

Musculus uvula: a paired muscle that forms chief bulk on the nasal surface of the velum.

N

Nasal air emission: the sound of air forcefully flowing through the nose during speech due to poor valving between the oral and nasal cavities.

Nasal grimace: a muscle contraction during speech that is typically noted either above the nasal bridge or around the nares and is usually accompanied by nasal air emission.

Nasal molding: a method of repositioning the nasal septum and ala in the infant prior to cleft lip repair.

Nasal regurgitation: reflux of fluids into the nasopharynx and nasal cavities during drinking or vomiting.

Nasal septum: a wall dividing the nasal cavities into two halves.

Nasal turbulence: a fricative turbulent sound generated by a partially opened velopharyngeal port, often called nasal rustle.

Nasalance: the ratio of nasal over nasal plus oral acoustic energy during speech as determined through the use of the Nasometer; represents the relative amount of nasal acoustic energy in the patient's speech.

Nasalization of oral phonemes: an obligatory error due to an open velopharyngeal port.

Nasolabial fistula: a fistula (opening) in the alveolus that is often deliberately left by the surgeon during the initial repair to allow for maxillary growth.

Nasometer: a computer-assisted instrument (KayPENTAXTM, Lincoln Park, NJ) that measures the relative amount of nasal acoustic energy in a patient's speech.

Nasopharyngoscopy: an endoscopic procedure that allows visual observation of the velopharyngeal mechanism or larynx during speech, where a nasopharyngoscope is inserted through the nose until it reaches the nasopharynx.

O

Obligatory errors: speech characteristics that are the product of structural abnormality or dysfunction.

Occult submucous cleft: a defect in the velum that is under the mucous membrane and not visible on the oral surface; this defect can usually be viewed on the nasal surface of the velum through nasopharyngoscopy.
Otitis media: infection and inflammation of the middle ear.

P

Palatal fistula: an opening in the palate which may be the result of a breakdown of the area of a previously repaired cleft, the result of maxillary expansion, or even growth.
Palatal lift: a prosthetic device used to assist raising the velum in cases where the velum is long enough to achieve velopharyngeal closure but does not move well.
Palatal obturator: a prosthetic appliance used to cover an open palatal defect such as an unrepaired cleft palate or a palatal fistula.
Passavant's ridge: a prominence on the posterior wall of the nasopharynx formed by contraction of the superior constrictor muscle.
Pharyngeal affricate: a compensatory articulation production that is a combination of pharyngeal or glottal stop and a pharyngeal fricative.
Pharyngeal fricative: a compensatory articulation production while air escapes through a small opening between the base of the tongue and the posterior pharyngeal wall.
Pharyngeal stop: a compensatory articulation production that is produced by the base of the tongue articulating against the pharyngeal wall.
Pharyngeal wall augmentation: an implant surgically performed in the posterior pharyngeal wall to correct velopharyngeal dysfunction.
Pharyngoplasty: a surgical procedure of the pharynx that is designed to correct velopharyngeal dysfunction.
Posterior nasal fricative: an abnormal articulation production due to an audible friction sound of air escaping through a velopharyngeal opening.
Posterior pharyngeal wall: back wall of the throat.
Premaxilla: a triangular-shaped bone that is bordered on either side by the incisive suture lines.
Primary palate: the lip and palate anterior to the incisive foramen.
Prolabium: the isolated central soft-tissue segment of the upper lip in an unrepaired bilateral cleft palate.

R

Relative prognathism: Retruded upper jaw causes the lower jaw to appear overly large.

Resonance: the quality of the voice that results from the vibration of sound in the pharynx, oral cavity, and nasal cavities.
Retrognathia: recession of one or both of the jaws.
Rolled flap: a flap of tissue that is surgically raised from the posterior pharyngeal wall to form a bulge.
Rule of 10s: a guideline for the appropriate time for a cleft lip repair; the infant must be at least 10 weeks old, weighs at least 10 pounds, and have a haemoglobin of 10g/dL.

S

Sagittal pattern: the least common pattern of velopharyngeal closure where the closure is primarily achieved by the medial displacement of the lateral pharyngeal walls in the midline.
Secondary palate: structures that are posterior to the incisive foramen, including the velum and the portion of the hard palate posterior to the premaxilla.
Speech bulb obturator: a prosthetic device that aids to fill in the space between the velum and the posterior pharyngeal wall to achieve velopharyngeal closure.
Sphincter pharyngoplasty: a type of pharyngoplasty to create a dynamic sphincter that encircles the velopharyngeal port.
Submucous cleft palate: a type of cleft palate that is characterized by a midline deficiency in the bony structures of the hard palate or the muscles of the velum with intact oral surface by the covering of the mucous membrane.
Superior pharyngeal constrictor: paired muscles in the upper pharynx that is responsible for displacing the lateral pharyngeal walls medially to assist velopharyngeal closure.

T

Tensor veli palatini: paired muscles that contributes to opening the Eustachian tube to enhance middle ear aeration and drainage.
Teratogen: a drug or other agent that can produce congenital anomalies or birth defects or increase the incidence of an anomaly in the population.
Tonsillectomy: a surgical removal of the tonsils.
Turbinates: bony structures inside the nose (superior, middle, and inferior turbinates).

U

Uvula: a teardrop-shaped structure projected from the posterior edge of the velum.

V

Velar dimple: the area on the oral surface of the velum where it bends during velopharyngeal closure.

Velar eminence: a bulge on the nasal surface of the velum during phonation.

Velar fricative: a compensatory articulation production that is produced while air escapes through a small opening between the back of the tongue and the velum.

Velar stretch: the process where the velum elongates as it elevates to achieve velopharyngeal closure.

Velopharyngeal dysfunction (VPD): a generic term used to describe velopharyngeal malfunction.

Velopharyngeal inadequacy (VPI): a generic term used to describe velopharyngeal malfunction.

Velopharyngeal incompetence (VPI): a physiologic disorder resulting in poor movement of the velopharyngeal structures.

Velopharyngeal insufficiency (VPI): an anatomical defect that precludes adequate velopharyngeal closure by causing the velum to be short relative to the posterior pharyngeal wall.

Velopharyngeal mislearning: inadequate velopharyngeal closure due to faulty learning of appropriate articulation patterns.

Velum: the posterior muscular portion of the palate covered by the mucous membrane.

Vermilion: the red pigmented portion of the upper and lower lips.

Videofluoroscopy: a radiographic procedure commonly used to examine internal structures of the body during movement, which allows assessing the velopharyngeal mechanism and other oral and pharyngeal structures during speech.

Vomer: the inferior and posterior portion of the nasal septum.