

Cleft Lip and/or Palate (*Facial Cleft Deformity*)

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www.up.ac.za/up/web/en/academic/maxillo_facial_and_oral_surgery/index.html OR <http://web.up.ac.za>

The multidisciplinary approach and the therapy protocol

Multidisciplinary approach

The facial cleft deformity or cleft lip, alveolus and palate and exceptional cleft deformity cannot be treated successfully by only one discipline or specialty. The interaction and consultation between various disciplines enhances understanding of the possibilities and limitations of the various disciplines involved.

A typical multidisciplinary clinic consists of the following basic members:

- Surgeon [Maxillo-Facial and Oral Surgeon]
- Orthodontist
- Paediatrician
- Speech-language therapist
- Community nurse

Secondary members of the clinic are those members who attend this multidisciplinary clinic or who are on standby:

- Paedodontist
- Prosthodontist
- Ear, nose and throat specialist
- Clinical psychologist
- Social worker
- Human geneticist
- Occupational therapist
- Community dentist or an Oral hygienist
- Dietician

The interdisciplinary referral base becomes broader when a secondary discipline becomes permanently involved in the clinic and is not only on standby.

Therapy protocol

The therapy protocol, treatment protocol or treatment plan table is the time table used for scheduling certain treatment aspects and sequences in a particular facial cleft deformity clinic. Such a therapy protocol must be scientifically motivated, judged and proven. If possible, the team members of a clinic must adhere strictly to such a therapy protocol.

The therapy protocol

<i>TIMING</i>	<i>PROCEDURES</i>	<i>TYPE OF CLEFT</i>
3 ± 3 Days	<p>Consultations Facial deformity examination [team] Advice on feeding Treatment plan</p> <p>Pediatrics Full examination</p> <p>Orthodontics Functional-orthognathic suction and drinking plate Suction and drinking plate Headgear [after 3 weeks]</p> <p>Genetics Parent counseling</p> <p>Psychology Parent counseling</p>	<p>All</p> <p>All</p> <p>LAP hPsP, (sP) LA[b], LAP[b]</p> <p>All</p> <p>All</p>
5±1 Months	<p>Surgery Velum repair [<i>with muscle repair and tensor sling for the Eustachian tube</i>] Hard palate repair [<i>only in bilateral cleft, one side closure</i>] [<i>NO striping of the hard palate muco-periosteum</i>]</p> <p>Ear, Nose and Throat Examination</p>	<p>LAP LAP[b]</p> <p>LAP, hPsP, sP</p>
7±1 Months	<p>Surgery Velum repair <i>with muscle repair and tensor sling for the Eustachian tube</i> Hard palate repair [<i>NO striping of the hard palate muco-periosteum</i>] Anterior nasal floor reconstruction Lip repair [<i>Tight closure from the soft plate, to hard palate, to anterior nasal floor and to the lip can therefore be achieved</i>]</p>	<p>hPsP, sP LAP LA, LAP L, LA, LAP</p>
12±1 Months	<p>Surgery Columella lengthening</p>	<p>LA[b], LAP[b]</p>
18±1 Months	<p>Surgery Hard palate repair</p>	<p>hPsP</p>
1½ - 8 Years	<p>Speech-language therapy Active treatment Fluorographic and/or naso-endoscopic examination [3½ - 4½ years]</p>	<p>LAP, hPsP, sP LAP, hPsP, sP</p>
4½±1 Years	<p>Paedodontics Preventative treatment</p> <p>*Prosthodontics Obturator [<i>should an oro-nasal fistula occur</i>] Speech bulb prosthesis</p> <p>*Surgery Velo-pharyngeal flap [<i>only if indicated by fluorographic examination</i>]</p>	<p>All</p> <p>LAP, hPsP LAP, hPsP, sP</p> <p>LAP, hPsP, sP</p>

7±½ Years	Orthodontics [<i>secondary</i>] Preventive treatment [<i>anterior crossbite - selective extraction</i>]	LA, LAP
12±2 Years	Orthodontics [<i>tertiary</i>] Final treatment [<i>in orthognathic cases, treatment will commence at 15±2 years</i>] Surgery Osteoplasty or osteofusion [<i>secondary</i>]	LA, LAP, L*, hPsP*, sP* LA, LAP
16±2 Years	*Surgery Orthognathic surgery *Prosthodontics Final rehabilitation *Surgery Aesthetic revision [Lip, Nose] Genetics Adolescent counseling	All LA, LAP L, LA, LAP All

* = Indicated for a particular problem only, and not a necessity for that particular cleft.

Abbreviations:

- L = cleft lip
- LA = cleft lip and alveolus
- LAP = cleft lip, alveolus, hard and soft palate
- hPsP = cleft hard and soft palate
- sP = cleft soft palate
- u = unilateral
- b = bilateral

The purpose of a therapy protocol

1. To treat a baby, child and adolescent at the most optimal time.
2. To treat a baby, child and adolescent with the most optimal surgical technique.
3. To avoid surgical and orthodontic over-treatment. This would in the long-term, result in a dental/functional and/or speech and/or aesthetic cripple (NB: A Facial Cleft Syndrome Appearances).

Classification of clefts

L	unilateral or bilateral	} total or partial [and Simonarts' band]	} complete or incomplete
LA	unilateral or bilateral	} total or partial	} complete or incomplete
LAP	unilateral or bilateral	} total or partial	} complete or incomplete
hPsP	unilateral or bilateral	} total or partial	} complete or incomplete
sP		total or partial [bifid uvula]	} complete or incomplete [submucosal]

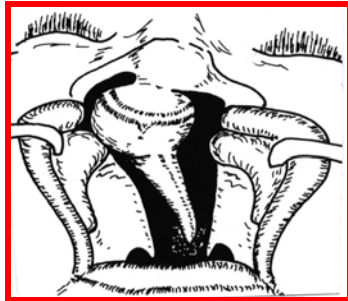
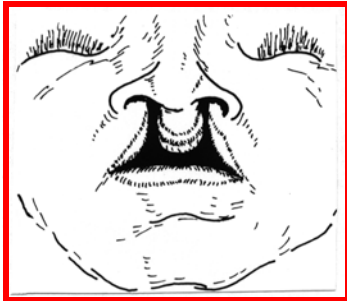
Cleft lip, alveolus and palate [unilateral]

Left: LAP[u] (total), Right: [hP] (partial) [Figure 1]



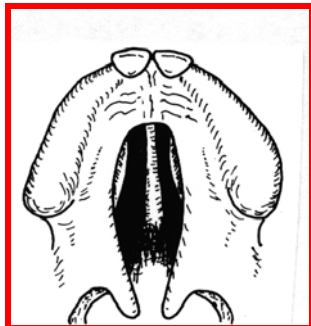
Cleft lip, alveolus and palate [bilateral]

Left and right LAP[b](total) [Figures 2 and 3]



Cleft hard and soft palate

hPsP (hP partial, sP total) [Figure 4]

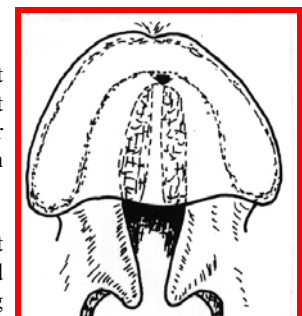


Functional-orthognathic treatment and the pre-surgical phase

Suction and drinking plate

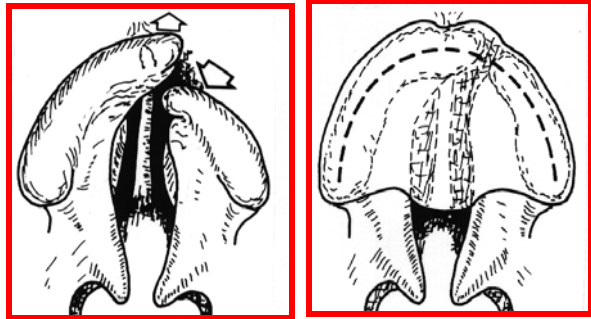
The suction and drinking plate functions as an obturator and is used for the combined hard and soft palate cleft (hPsP) [Figure5]. It is seldom used for a soft palate cleft only (sP). A baby with only a soft palate cleft (sP), but with a relatively severe micrognathia, very often also has a drinking and/or suction problem, and then may also require such a special suction and drinking plate but with an extension into the cleft part.

A Pierre Robin syndrome baby is an infant born with an u-shaped cleft of both the hard and soft palate (hPsP), micrognathia of the mandible and microglossia where the tongue fits into the cleft and the resultant glossoptosis may cause asphyxia. This specific child requires a suction and drinking plate with an extension and should be fed in the upright position.



Functional-orthognathic movement

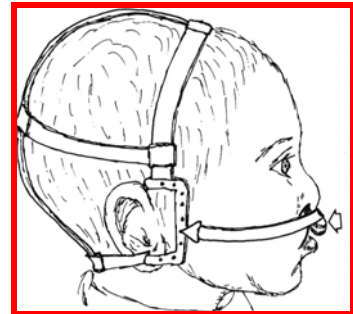
The functional-orthognathic movement, also known as the primary orthopedic or jaw-orthognathic movement, is the description used for alveolar or dento-alveolar segment movement of the individual alveolar segments before the first surgical intervention. The individual segments should be as perfectly aligned as possible [Figure 6, un-aligned alveolar segments]. The functional-orthognathic suction and drinking plate therefore has a dual purpose, namely to initiate functional-orthognathic movement of the lateral alveolar segments [Figure 7], as well as to create a temporary closure between the nasal and oral cavities. The functional-orthognathic treatment lasts up to the first surgical intervention which is at about five months of age.



Repositioning of the pre-maxilla

The bilateral facial cleft deformity

Active repositioning of the pre-maxilla is required in cases of a bilateral cleft lip and alveolus, and bilateral cleft lip, alveolus and palate where there is no partial fusion between the lateral lip segments and pro-labium or where a Simonarts' band is present. The baby born with a bilateral facial cleft deformity usually has a protruding pre-maxilla-pro-labium immediately visible after birth. A special head-gear with an elastic band positioned over the upper lip is necessary for the repositioning of the pro-labium-pre-maxilla part of the middle facial structures [Figure 8].



Pre-surgical phase

Functional-orthognathic appliance

The main purposes of the functional-orthognathic suction and drinking plate are:

- to achieve functional-orthognathic movement, thus movement with growth [lateral alveolar segments], and movement against growth [a separate premaxilla as in a bilateral cleft],
- to achieve a normal suction and drinking pattern [creating a border between the nasal and oral cavities],
- to discourage any abnormal habits [example: placing a thumb, finger or the tongue into the cleft, thus preventing the natural narrowing of the cleft],
- to enhance therapeutic speech muscle development [normal suction allows better development of muscles used for speech],
- to facilitate normal breathing during suction and drinking as well as during sleep [this is of utmost importance in a baby with the Pierre Robin syndrome],
- to support the parent psychologically, in that the treatment of their baby has been started without delay.

General

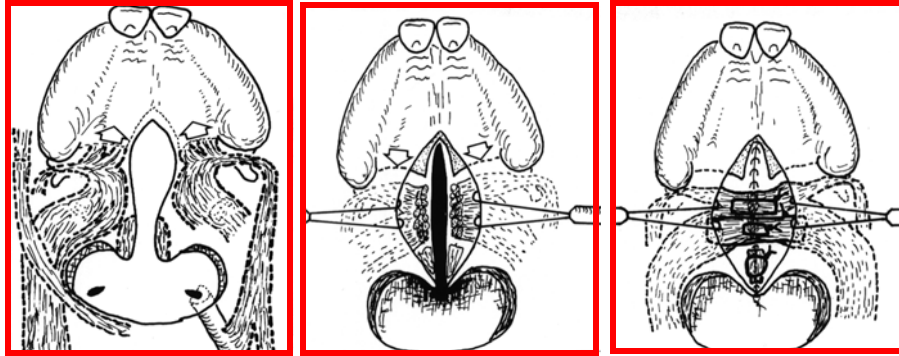
The pre-surgical phase is the time-span from birth up to the first surgical intervention. During this very important period the parent experiences the trauma of having a child with a facial cleft deformity, but also receives support through counseling, as well as through physical means such as positioning of the functional-orthognathic feeding plate.

It is imperative that every clinic member should know the treatment protocol, as the parent will address his/her questions to any clinic member, irrespective of their particular specialty or training. During this pre-surgical phase a patient may be referred to an ear-, nose- and throat specialist, a social worker or some other specialist, or sub-specialist for a particular aspect.

Primary surgical repair of the palate

Primary repair of the soft palate

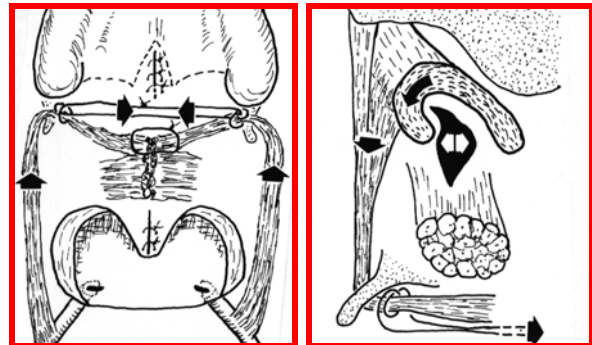
The soft palate cleft {in cleft lip, alveolus and palate, unilateral and bilateral types (LAP[u+b])} is repaired at the age of five months. Where there is only a soft palate cleft (sP) or only a soft and hard palate cleft (hPsP), the first intervention is done at the age of seven months.



The intravelar veloplasty (with certain modifications) is the surgical technique used for the closure of the soft palate cleft [Figures 9, 10 and 11]. A tensor sling around the tensor veli palatini muscle is also incorporated during each procedure for better middle ear ventilation [Figures 12 and 13]. The tensor tendon should never be explored or cut. This may lead to severe middle ear effusion. Where the cleft is complete and bilateral

(LAP[b]), one half of the hard palate cleft (and the soft palate cleft) is repaired by means of an inferiorly-based vomer-septum flap. Where there is a hard and soft palate cleft (hPsP) only [either a partial or complete hard palate cleft], the hard palate is closed at 18 months only.

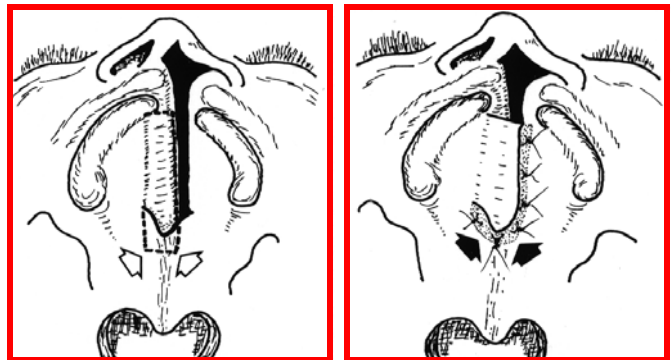
The intravelar veloplasty is therefore an anatomical-functional approach for the primary closure of the soft palate cleft. This surgical closure involves very careful dissection and alignment of all the relevant muscle components, namely, the uvula, palato-pharyngeus, levator veli palatini and tensor veli palatini.



During the intravelar veloplasty procedure a tensor veli palatini muscle tension sling for the improved function of the Eustachian [pharyngo-tympanic or auditory] tube is inserted in the soft palate. The tissue adjacent to the hamulus process is neither dissected, nor fractured to relieve the tension of the mucosa, nor is the tendon of the tensor veli palatini cut. The negative sequella of persistent middle ear effusion in the cleft lip and/or palate infant, who is already at risk of developing a communication disorder, necessitates improved patency of the Eustachian tube.

Primary repair of the hard palate

The surgical repair of the cleft hard palate is done normally at the age of seven months (LAP[u+b]). The hard palate cleft is usually closed after the repair of the soft palate cleft [Figures 14 and 15], and it is one of three procedures done at the age of seven months in a complete cleft lip, alveolus and palate (LAP[u+b]) case. The cleft hard palate closure precedes the anterior nasal floor reconstruction and the cleft lip repair, which are done during the same procedure, to achieve a tied closure of the oral cavity. No mucoperiosteal stripping is done of the hard palate, as this causes in the majority of cases relative severe midfacial growth disturbances (known as a Facial Cleft Syndrome).



The exceptions to this general rule of seven months is the bilateral cleft (LAP[b]), where one half of the cleft has already been closed during the primary closure of the soft palate cleft. The other half is then repaired at seven months. The other exception is a cleft and especially a u-shaped cleft of the hard palate, without an alveolus cleft (hPsP), where the hard palate cleft is closed at 18 months. By this stage, considerable narrowing of the hard palate cleft defect has occurred.

Various procedures for the closure of the hard palate have been published. It is vitally important that the particular procedure chosen should not lead to long-term mid-facial growth disturbances. The only surgical procedures which should be used in the infant are the vomer techniques, and then only those where no stripping of the hard palate mucosa is done. However, the cleft hard palate should be closed as early as possible, as an open hard palate cleft is detrimental to normal speech development.

Primary surgical repair of the anterior nasal floor

At seven months of age, the closure of the hard palate cleft precedes the reconstruction of the anterior nasal floor performed

during the same surgical procedure. The surgical repair of the cleft lip then follows. All three techniques are therefore executed during a single surgical session.

The purpose of an anterior nasal floor reconstruction is twofold:

- the tight closure between the dento-alveolar arches nearly always inhibits the formation of an anterior oral nasal fistula,
- to prevent collapse of a near-ideal dento-alveolar arch alignment.

This arch alignment was achieved by means of the functional-orthognathic treatment. The excess lip tissue remaining after the lip has been dissected, may be successfully used for the creation of an anterior nasal floor at the buccal sulcus and between the dento-alveolar arches. The double-layered anterior nasal floor closure, as well as the supra-periosteal dissection, prevents mid-facial growth disturbances in the long-term.

Primary surgical repair of the lip and nose

Primary surgical repair of the unilateral cleft lip

The primary repair or reconstruction of a unilateral cleft lip is done at the age of seven months. The complete cleft, thus a cleft lip, alveolus and palate (LAP[u]), as well as the cleft lip and alveolus only (LA[u]), are repaired. The exception is where there is only a cleft lip (L[u]), which may already be reconstructed at the age of five months. It is essential that the cleft soft palate, the cleft hard palate, as well as the anterior nasal floor are reconstructed before the lip is closed. This will ensure a tight closure of the whole palate, with a decreased possibility of the development of an oro-nasal fistula during and after the initial repair stage. When the reconstruction is done from the posterior towards the anterior part of the cleft defect, in other words, from the soft palate towards the lip, the chances of the development of an oro-nasal fistula are also diminished.

When repairing the cleft lip, the following aspects must be taken into consideration:

- the aesthetic appearance of the lip, including the lip skin with the philtrum edges, the lip white-red junction, the lip red dry-wet junction, and the nasal sill with the alar base rotation,
- the function of the lip, with the correct muscle alignment; this should also automatically improve the aesthetic appearance,
- the long-term mid-facial growth disturbance or dysgnathial effect on the mid-face should determine the choice of surgical technique used for the reconstruction of the cleft lip,
- how the closure of the cleft lip will influence the "cleft" nasal structure, especially the effect it will have on the nasal sill and the alar base.

Anatomical considerations



The basic anatomy should be considered in the anatomical and functional repair of the cleft lip [Figure 16]:

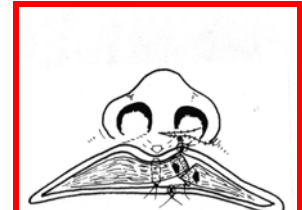
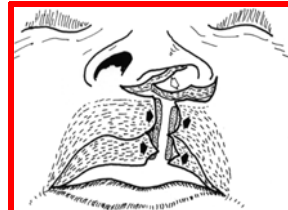
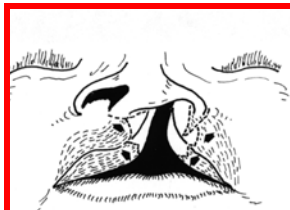
- the philtral column,
- the C-junction of the alar base (nostril sill),
- the slight bulging and continuation of the nasal floor,
- the vermilion-cutaneous junction (white skin roll),
- the vermilion moist-dry line (muco-vermilion border).

An anatomical and functional surgical reconstruction of the cleft lip therefore involves the following aspects:



- The lip scar should be located in the following anatomical regions [Figure 17]:
- the philtral column is the only natural fold or bulge which can be used to resemble a vertical scar in the lip,
- a horizontal alignment of the vermilion- cutaneous junction and moist-dry line would interrupt any vertical scar and lead to a balanced vermilion.
- The inferior part of the nasal structure should be aligned for maximal aesthetics:
- the alar base is curved medially and positioned to form a C-junction; this is the natural naso-labial crease, and
- this C-junction with its extended tissue creates a natural nasal floor due to the slight bulging of the tissue band lying across the inferior nasal orifice.

All the above-mentioned considerations should be incorporated in an anatomical lip-plasty design. However, the functional repair is achieved by the proper reconstruction of the muscle segments [Figures 18 to 20].

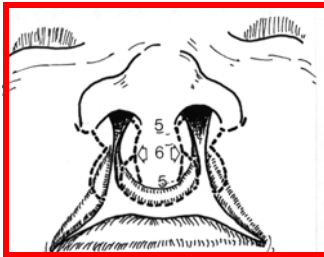


The lip-adhesion technique is only considered for babies born with a "median facial dysgenesis", in other words, with a cleft lip width of about 20 millimeters and more. These babies present with a complete cleft lip, alveolus and palate (LAP[u]) and the atrophic alveolar arches are separated by 20 millimeters or more. The columella is usually shortened.

Primary surgical repair of the bilateral cleft lip

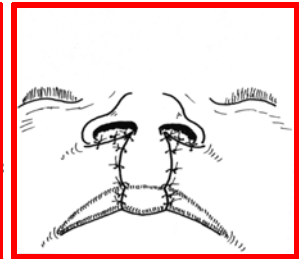
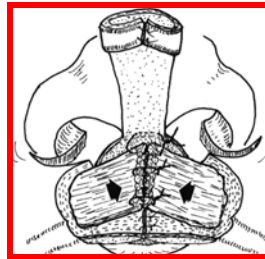
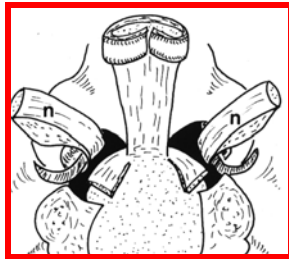
As in the case of the unilateral cleft lip, the repair of the bilateral cleft lip is done at seven months of age. Only one side of the bilateral cleft hard palate in cases of bilateral cleft lip, alveolus and palate (LAP[b]) must be closed at this second surgical intervention, as the other side of the cleft hard palate has been closed with the cleft soft palate repair at the age of five months. Bilateral anterior nasal floor reconstructions are indicated in cases of a cleft lip and complete cleft alveolus (LA[b]), as well as where there is a cleft lip, alveolus and palate (LAP[b]).

There is a protruding pro-labium-pre-maxilla component and the growth is always in the anterior direction. Mid-facial dysgnathial growth is therefore not of such importance, apart from where the surgeon should resect a part of, or even the whole, pre-maxilla. The latter surgical intervention is an absolute contra-indication. The type of closure chosen by the surgeon must be one which will produce a very good aesthetic result, directly post-operatively, as well as in the long-term. However, attention must also be given to a well-balanced closure of the underlying dento-alveolar arch with a deep enough alveo-labial sulcus so that future dental health in this region may be optimal.



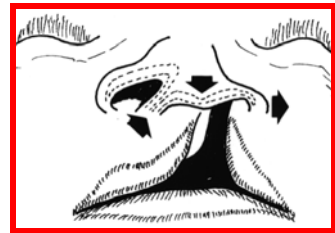
The scar caused by the vertical incision lines in the upper lip should resemble the philtral edges [Figure 21]. When designing these incision lines, the surgeon should accurately measure the intra-philtral distance for the proposed "philtral edges" [Figures 22 to 24]. There are only very few bilateral lip reconstructive procedures which allow full orbicularis oris muscle reconstruction.

A complete bilateral cleft lip always presents with an atrophic columella. After the surgical closure of the bilateral cleft lip the nasal appearance is flatter than normal in the antero-posterior dimension because the columella is atrophic. Last-mentioned is surgically corrected at twelve months of age.



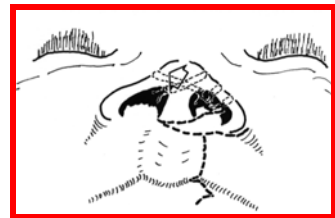
Nasal deformity, and its primary correction, including lengthening of columella

The severity of the nasal deformity in a patient with a cleft lip, with or without cleft alveolus and palate, depends on the width of the cleft in the lip, as well as on the completeness of the cleft lip. The very abnormal appearance of the nasal ala on the cleft side is a result of the rotated (anti-rotational) alar cartilage [Figure 25].



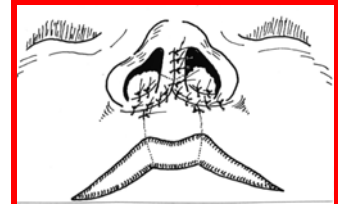
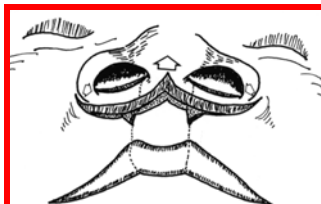
Unilateral cleft nasal deformity

The unilateral nasal deformity is corrected during the same procedure as the lip-plasty, therefore at the age of seven months. Long-term nasal growth deformity is avoided by not dissecting the nasal dome. The deformed nasal alar cartilage may be surgically repositioned. The deviated nasal dome must therefore be carefully sutured [Figure 26]. This allows uninhibited growth of the individual character of the nasal skeleton.



Columella lengthening

Lengthening of the columella is necessary when the cleft lip is bilateral and complete. This lengthening is done at the age of twelve months, as by this time the scar left by the repair of the bilateral cleft lip should be well healed. Projected



growth of the nasal dome in the antero-posterior dimension can take place after the correction of the columella [Figures 27 and 28].

Speech-language therapy and velopharyngeal incompetence

Speech-language therapy

The child with a cleft palate deformity (LAP, hPsP and sP) will nearly always require speech therapy, even if it is only for comprehensive speech and language assessment. The severity of the cleft is first evaluated and then the parents are informed about speech and language production development, the importance of speech and language stimulation and the contribution they themselves can make.

Age and speech development

Speech is produced with the first cry of the baby. The functional-orthognathic plate temporarily closes the cleft hard palate, so that the nasal cavity is separated from the oral cavity. The positioning of the tongue is therefore normalized as it is kept out of the cleft. Suction and drinking is then relatively functional, although not entirely normal and these functions stimulate the use of the soft palate muscles. Function of the soft palate muscle leads to normal speech production. Furthermore, this functional-orthognathic treatment plate leads to a more normal swallowing mechanism which in turn, results in less food particles being deposited in the area of the Eustachian orifice, and thus less chance of middle ear effusion with its sequella of serous otitis media. In this respect, the surgeon is also doing his bit by placing the tensor sling for improved Eustachian patency.

At the age of 12 months the baby may produce three short audible words with inconsistent use of the first word when referring to an object or person. At around this age, communication and hearing assessment are essential and the speech-language therapist has to plan and, if necessary, begin an active speech-language therapy programme as an early intervention. At about two years of age, the toddler should be using half of all the sounds/words of the home language, and more or less half of what he says should be intelligible to others. At the age of four, the child should be using the majority of sounds, and speech should be completely intelligible to others.

Structural and functional speech mechanism

The smooth development of speech depends on the structural and functional adequacy of the relevant anatomical structures. The build-up and release of air within the oral cavity produces the consonant sounds of the language. In the case of a cleft palate where no repair has been done or where air "leaks" under pressure into the nasal cavity, the child will communicate with a distorted, functionally compromised, speech pattern. "Velopharyngeal competence" is the term given to the sealing-off of the nasal cavity from the oral cavity, by means of contact between the soft palate and the pharyngeal wall. Hypernasality is therefore a typical sign of velopharyngeal incompetence, in other words, the seal between the soft palate (with its superior movement), and the pharyngeal wall (with its anterior and lateral movements) is not functioning adequately. Air is therefore escaping through the nose during speech, especially when the vowels are phonated.

Velopharyngeal insufficiency (incompetency)

The velopharyngeal valve is only one part of the vocal tract, and its degree of closure depends on the sounds which are to be produced. High vowel sounds combined with oral sibilant-fricative consonants require the most complete closure of the nasal cavity, and thus of the velopharyngeal valve. However, low vowels combined with nasal consonants require an open nasal cavity, and the velopharyngeal valve should therefore be at rest or be inactive.

As soon as there is incomplete instead of normal closure of the velopharyngeal valve, nasal air escapes and hypernasality occurs. This is called velopharyngeal insufficiency. One has to distinguish between the problem of a blocked nasal cavity (therefore a denasality will be noticed) and that of velopharyngeal insufficiency (with hypernasality), as abnormal speech production may sound similar to the layman. However, denasality mainly affects m, n, and g, where hypernasality mainly affects p, b, t, d, k, g, f, and s. Therefore both affect intelligibility.

Assessment of velopharyngeal function

The patient with velopharyngeal insufficiency should be assessed with regard to hearing, anatomical structures of the soft palate, pharynx (including the tonsils), hard palate, nasal cavity (especially air flow), teeth and lips. Furthermore, the neurological and psychological background of the child must also be assessed.

The golden rule is that the speech-language therapist treats the child intensively for six months. The speech production and velopharyngeal function are then re-assessed. Should there still be a velopharyngeal insufficiency, then imaging techniques should be used for examination of the velopharyngeal function.

Fluoroscopic examination

This radiologic examination may be done by means of any of the following techniques: normal radiographs, cine or video. The most modern method is multiview video-fluoroscopy which allows the evaluation of the dynamics of the velopharyngeal valve during production of speech. The most important planes are the lateral facial view [Figure 29] and the frontal or postero-anterior view. The lateral facial dimension shows movement of the soft palate and posterior pharynx.



Before surgery is contemplated for correction of the velopharyngeal inadequacy, the child, usually in the age group of 3½ to 5½ years, is referred to the prosthodontist of the facial cleft deformity clinic for assessing whether a speech bulb may be possible. In certain instances, especially where the muscles of the soft palate are used ineffectively, the velopharyngeal insufficiency may be controlled prosthetically by means of a pharyngeal bulb or palatal lift.

Surgical repair of velo-pharyngeal inadequacy

The velopharyngeal insufficiency and its symptom of hypernasality may be corrected by placing a relatively non-physiological pharyngeal flap between the posterior pharyngeal wall and the soft palate [Figure 30]. Intensive speech-language therapy is necessary after this surgical reconstruction so that the previous abnormal compensatory speech habits may be re-addressed, and the correct production of speech sounds taught.



Oro-nasal fistula

Oro-nasal fistula - a complication after primary repair and its locations

An oro-nasal fistula, that is a small opening between the oral and nasal cavities, may occur in some cases after primary closure. Such a fistula may vary in size, and may be present in various locations.

As much care as possible should be taken to lessen the chances of an oro-nasal fistula occurring after the primary reconstruction of the soft palate, the hard palate and the anterior nasal floor, where it may especially be found at the junctions of the hard and soft palate as well as at the junction between the anterior nasal floor and the hard palate. The size of the oro-nasal fistula will determine the type of closure and the timing of the closure.

The hairline oro-nasal fistula which is not visible, will not have any negative effect on speech development. Such an opening should only be probed and then surgically closed once midfacial growth is almost completed. Temporary closure of a larger oro-nasal fistula may be achieved by means of a prosthetic device, such as an obturator, but a secondary surgical repair is necessary for permanent closure. Major dissection should never be done on a child, as midfacial growth disturbances (dysgnathia) will most certainly follow.

Pre-operative or directly post-operative viral and/or bacterial infections may have a negative effect on the healing process, particularly in those flaps which have been sutured under tension during the primary repair of the cleft.

The size and the location of the oro-nasal fistula is of real importance:

- A hairline oro-nasal fistula may be ignored as it does not interfere with speech. The patient may force air through it, resulting in a type of whistling sound.
- A small oro-nasal fistula, one to two millimeters at its broadest, is at first ignored and the development of speech controlled by speech therapy. Should the oro-nasal fistula constantly interfere with speech production in a child between 3 to 3½ years of age, then an obturator may be used. Should the child be 4½ years or older, then a surgical closure may be done.
- A bigger oro-nasal fistula of three millimeters and more at its widest dimension, requires an obturator. An obturator cannot be used for an oro-nasal fistula situated adjacent to the muscles in the soft palate, neither for one which is next to the uvula.

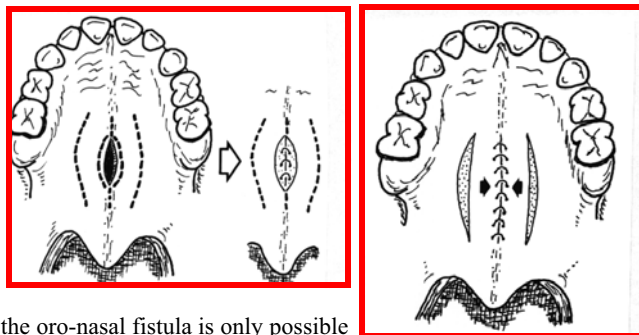
Surgical repair of the oro-nasal fistula in the child

Surgical closure of the oro-nasal fistula should only be done after the age of 4½ years. Where there is also velo-pharyngeal inadequacy (established by fluoroscopic and/or naso-endoscopic examinations, as well as by the treatment of a speech-bulb), both surgical reconstructions should be executed at the same time.

Where speech has developed normally, the surgical closure of the oro-nasal fistula may be delayed up to the age 11 ± 2 years when

the secondary osteoplasty procedure is done. Mid-facial growth at this age is nearly completed, and the surgical effect on dysgnathia would be negligible.

The success of the surgical repair of an oro-nasal fistula is much less predictable than that of primary reconstruction of the cleft palate [Figures 31 and 32]. Where a single layer of muco-periosteum [or muco-perichondrium] is extensively used in the primary surgery of the cleft hard palate, successful closure of the oro-nasal fistula is only possible by means of a double-layered technique.



Cleft Alveolus and Osteoplasty

The cleft alveolus should never be treated during the primary reconstructive procedure, as a bony fusion between the dento-alveolar segments will lead to mid-facial growth disturbances. Facial growth must be completed by at least two-thirds before any type of alveolar cleft reconstruction may be considered.

Well-aligned dento-alveolar arch segments requiring no mixed dentition orthodontic treatment (apart from the initial functional-orthognathial treatment), may be treated initial by surgery only.

In those cases where there is a slight to severely collapsed dento-alveolar arch segment (usually also with a cross-bite in that particular segment), an osteoplasty procedure must be done at the same time as the main orthodontic treatment.

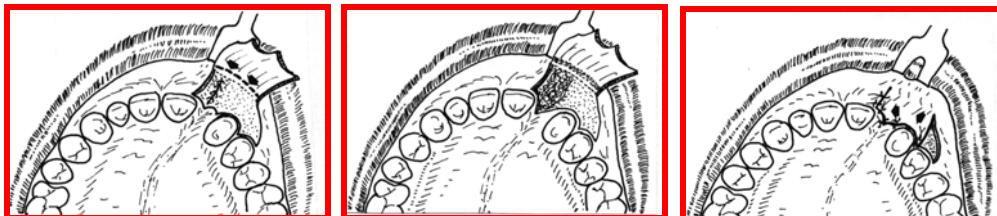
Osteoplasty (Osteofusion)

Osteoplasty or osteofusion describes the surgical technique whereby bone is transplanted into the cleft alveolus. The bone may be harvested from the crista iliaca, mandible, rib, calvaria or from the tibia. Spongiosa or a combination of spongiosa with cortex bone may be placed in the alveolar cleft.

This procedure is done at different ages, namely: the late secondary osteoplasty (executed in the age group 11 ± 2 years). The tertiary osteoplasty describes the stage where this procedure is done in the late adolescent years, usually at the same time as orthognathic reconstruction of the face. The ideal timing for an osteoplasty procedure is therefore when the root formation of the canine adjacent to the alveolar cleft is between one-third and two-thirds of its projected length. The child is then usually 11±2 years old.

The anterior nasal floor (area superior to the cleft alveolus), should be closed tightly by surgery, during the primary reconstruction of the nasal floor. Flaps adjacent to the alveolar cleft are used for a tight closure towards the palate, as well as in the buccal and sulcus

areas. Bone is transplanted into the pocket between the alveolar segments. The narrower the alveolar cleft, the easier it is to close the buccal and palatal area by means of attached (keratinised) epithelium [Figures 33 to 34].



Nasal alar base support

The nasal alar base on the cleft side is usually more posteriorly displaced than that on the non-cleft side. The reason for this is that the lesser alveolar segment is relatively atrophic with buccal bone loss in the alveolar cleft. During the osteoplasty procedure a sub-

periosteal tunnel is dissected beneath the nasal alar base, so that this part of the nasal structure may be lifted more anteriorly. The defect remaining between the alveolar segment and the nasal alar base is then filled with harvested.

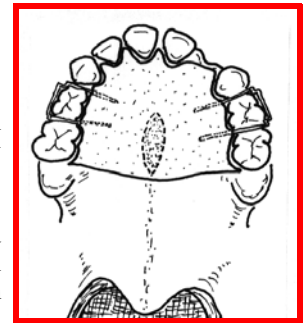
Prosthetic treatment

Obturator and speech bulb

Obturator

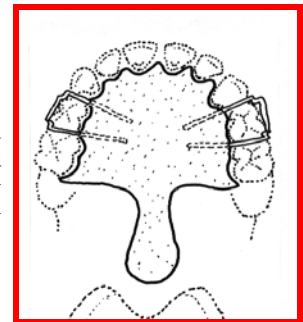
The obturator, a prosthetic appliance, is an artificial plate which closes an opening in the palate. In a facial cleft deformity this opening is the acquired oro-nasal fistula which develops after the primary reconstruction of the cleft alveolus, hard and soft palates.

The obturator [Figure 35] should not be confused with the suction and drinking plate, which is placed prior to the primary repair of the cleft palate and which extends partially into the soft palate cleft. An obturator may only be placed at the age of $4\frac{1}{2} \pm 1$ years when there is an oro-nasal fistula and when it interferes with speech development.



Speech bulb

The specialised field of prosthodontics plays an important role in the management of palatal lifts and pharyngeal speech aids in the patient with a facial cleft deformity. The palatal lift [Figure 36] is used in the young child so that velopharyngeal incompetence may be overcome and so that speech may develop normally. This palatal lift-prosthesis is used until the child is able to speak without any hypernasality.



Revision surgery

The decision to perform any revision surgery on the lip and/or nose, depends on whether there is adequate growth of the lip/nose or whether skeletal reconstructive surgery [orthognathic surgery] has provided adequate tissue to the facial skeleton. The soft tissue can therefore only be adjusted once the hard tissue is in the correct position. Furthermore, it is highly recommended that the orthodontic as well as the prosthetic treatment be completed before the revision surgery, as normal lip support and posture may be incorrect while these two treatment modules are not yet finalized.

The revision surgery is therefore done at the age of 16 ± 2 years, by which time most major cleft treatment has been completed and growth disturbances to the mid-face no longer play a role.

Lip scar and muscle revision

Unilateral cleft lip

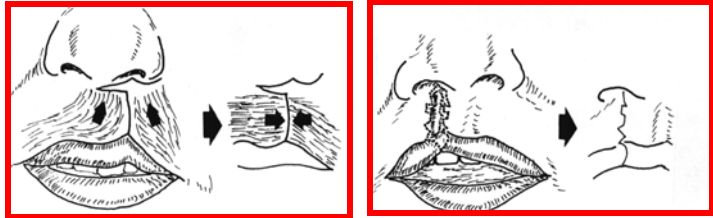
Where the primary lip reconstruction has been as ideal as possible, less revision surgery is necessary and the better the revision surgical result will be. However, this also depends on the initial width of the cleft in the lip, as well as the way in which the body will form scar tissue, which is an inherited factor. Where there has been previous revision surgery, it is more difficult to obtain the "perfect" end result.

During assessment of the lip and its scar, the following aspects are of importance:

- the aesthetic appearance of the scar,
- the type of cleft lip repair used,
- the shape of the cupid's bow,
- the length of the lip on the repaired side,
- the orbicularis muscle reconstruction and function,

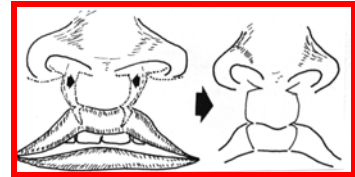
- the scar at the vermillion border,
- the indentation of the vermillion in the region of the lip red dry-wet junction,
- the concavity at the lip-nasal floor junction.

For each and every small deformity there is usually more than one procedure which may be used to correct that particular problem. A wide scar always requires some correction. However, if the scar is rotated inwardly, then the muscle also has to be re-aligned surgically [Figure 37]. A fine Z-plasty or Webster-plasty may be used to interrupt the even scar line for a better aesthetic appearance [Figure 38].



Bilateral cleft lip

The aesthetic aspects are similar to those of a unilateral cleft lip. However, symmetry of the lip scar is very important so that the scar may resemble the philtral edges, thus mimicking a balanced philtrum [Figure 39].



Lip volume adjustment

The patient with the bilateral cleft often has a relatively tight upper lip as the lip may be very thin or there may be a very atrophic prolabium. Fortunately, the latter is seldom seen where the repaired lip has been well-balanced. Once again, the skeletal base, that is, the maxilla with its intermaxillary part and the dento-alveolar arch, should first be placed in the correct position either by means of orthodontic rehabilitation and/or of orthognathic surgery and an osteoplasty before the lip volume is adjusted.

Revision of the nasal deformity

The nasal deformities associated with a facial cleft deformity differ entirely according to whether the deformity is unilateral or bilateral. The reconstruction of the residual deformity of the nose in a patient with a unilateral cleft is a more difficult procedure as it is not so easy to achieve a perfectly balanced nose.

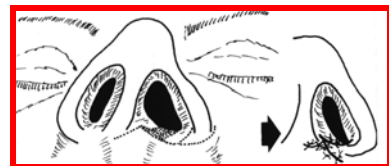
Unilateral nasal deformity

The severity of the unilateral nasal deformity in the adolescent depends on the initial width of the cleft lip deformity. Therefore, the wider the cleft in the lip at birth, the more severe the nasal deformity.

It must always first be determined whether there is any nasal tip deviation and/or deficient alar base support, and if this is so, it must be corrected so that there is sufficient bony support for the nasal alar base.

Basic variations of composite deformities of the nose on the cleft side are:

- nasal tip deviation,
- a posteriorly displaced alar cartilage,
- lateral part of alar cartilage (crura) buckled or bent into an anti-convexity,
- obtuse angle between the lateral and medial crura,
- right-angular alar sill - labial connection,
- widened or narrowed nostril floor, quite frequently depressed [Figure 40],
- deviated columella.



The major surgical corrections are best attempted by means of external excisions, preferably through the columella. The rotation and repositioning of the cartilages at the dome and on the lateral sides are less difficult and it should therefore be possible to perform these procedures accurately.

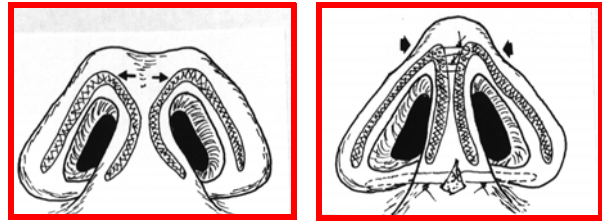
Bilateral nasal deformity

The nasal deformity, in cases of a bilateral facial cleft deformity, is usually relatively symmetrical, and may include the following problems:

- a short columella,
- a wide columella, which may be inferiorly or superiorly displaced,

- deviation of the alar cartilages with a midline dome retraction,
- widened nasal sills with wide nostril floors.

The columella lengthening procedure needs to be planned according to the tissue available and the scar tissue present [Figures 41 and 42].



Compromised adolescent / adult

The "compromised adolescent / adult", also known as the "end-stage" of a cleft lip and palate deformity, may either have a severe deformity or other problems following maturation of the facial skeleton. The patient may often, in fact, be described as a "functional and aesthetic cripple".

Severe deformities of varying degrees:

- a large residual oro-nasal fistula,
- mid-facial retrusion, such as a severe brevi-retrognathia with a retronasal appearance, either with or without other skeletal and/or dento-alveolar abnormalities,
- a severe speech defect,
- unsightly cosmetic appearance.

The features of these facial cleft deformity patients usually appeared quite acceptable after the primary repair when they were babies and children. However, as they grew older and reached the adolescent stage, the midfacial deformity became more and more pronounced. The reason for this mal-development lies in two main problem areas:

- the **multiple surgical attempts** to which such a poor baby or child has been subjected for improvement of the cleft deformity in the growing facial skeleton [the surgeon or cleft team members never adhered to any type of "therapy or treatment protocol"],
- the use of certain **primary surgical procedures** which lead to **growth disturbances** of the mid-facial region in the long-term.

One might argue, that every primary and intermediary surgical involvement may cause growth disturbances. This is true. However, the surgical procedures chosen for the baby, should be those which lead to the least dysgnathial and functional problems, including speech problems. Furthermore, the primary surgical techniques should also end in a good aesthetic result. It is not acceptable that the repair of the facial cleft deformity should be delayed until the age of 14 years, as this adolescent would then have been exposed to severe socio-psychological trauma and he/she would not be capable of normal speech.

The compromised adolescent / adult requires a comprehensive assessment of the present facial appearance as well as of all the different functional aspects. The easiest method of doing this is to take the therapy protocol and to compare the patient with the stage at which he should be.

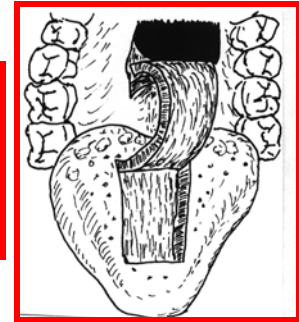
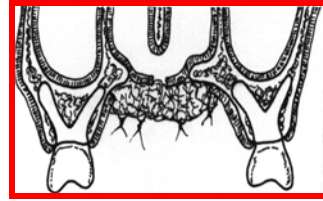
The main step is to **evaluate the facial-oral-pharyngeal structures** comprehensively from extra-orally to intra-orally:

- **nose:** midline deviation, width, retro-nasality, columella, alar cartilage, alar base, alar sill, nostril floor and scar tissue,
- **lip:** scar position and symmetry in a bilateral cleft lip, aesthetic appearance of the scar, orbicularis oris muscle function and approximation, lip skin-vermilion [red-white] junction, form of the cupid's bow, lip red dry-wet junction in the vermilion and the amount of vermilion in the cleft region, and the vermilion bulk tissue of the cleft lip segments,
- **orthognathic deformity:** maxillary retro-brevignathism or retrognathism, maxillary longignathism, mandibular prognathism, retro-brevigenion or progenion, and any type of laterognathism,
- **dento-alveolar:** anterior cross bite due to the premaxilla position, lateral cross bite due to collapsed lateral alveolar segment; superiorly rotated alveolar segment, general malocclusion, anterior nasal floor oro-nasal fistula, fusion of the alveolus cleft by means of a previous osteoplasty-procedure, width of the alveolus cleft and loss of teeth,
- **hard palate:** residual oro-nasal fistula and its size, scar tissue in the hard palate,
- **soft palate and velopharyngeal sphincter closure:** oro-nasal fistula in the soft palate, speech muscle realignment, adequate velopharyngeal competence,
- **other aspects:** dental and oral health, teeth present, bone resorption of part of, or of the total alveolar ridge and prosthodontic problems.

Treatment must be considered on the basis that the hard tissue repair, for example, fusion of the dento-alveolar segments and adjustment of the facial skeleton position, should be done before the soft tissue is repaired. Many of the reconstructions do not differ from those already described.

Large oro-nasal fistula repair

The large oro-nasal fistula results from various attempts at closure of a small oro-nasal fistula. After each surgical attempt, the oro-nasal fistula usually becomes wider and mid-facial dysgnathia may develop. The more scar tissue present, the less successful a closure by means of local flaps. In such cases, distance flaps, such as a tongue [Figure 43] or buccal fat pad [Figure 44] or temporal muscle flap, should be used.



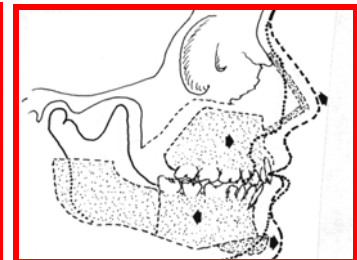
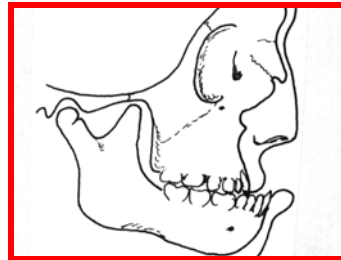
Severe orthognathic deformities

Severe mid-facial retrusion is always attributed to the type of surgical technique used for the primary reconstruction, as well as to the multiple surgical involvements attempted at achieving palatal closure and/or improving the appearance of the lip in the growing child. The primary surgical techniques which cause dysgnathial growth of the mid-face are:

- large areas of palatal mucoperiosteal lifting,
- undermining of the periosteum at the anterior dento-alveolar ridge and in the infraorbital area for cleft lip closure,
- partial or total removal of the premaxilla in a bilateral cleft facial deformity, and
- the type of technique used in the unilateral cleft lip repair.

Combination of orthognathic deformities, oro-nasal fistula and other

A comprehensive orthodontic and orthognathic surgical approach is necessary for the treatment of the recently-matured facial skeleton where there is a maxillary hypoplasia, with or without mandibular prognathism [and/or retrogenion or progenion] [Figures 45 and 46]. There might also be a small oro-nasal fistula. An osteoplasty procedure may or may not be indicated. All these problems may be corrected during one surgical involvement.



Other surgical combinations are also possible, such as an orthognathic correction with a nasal bridge and/or columella augmentation.

In these cases a mid-facial degloving procedure is usually indicated. Another combination would be an orthognathic correction with a velopharyngeal flap. However, one has to take into account the effect this might have on the patency of the airway directly post-operatively.