CLEFT LIP and PALATE

Sahlgrenska University Hospital
Göteborg, Sweden

Information about Cleft Lip and Palate

English version
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What are Cleft Lip and Palate (CLP)?

Common malformation
Cleft lip and palate are common malformations. Treatment has continuously been improved, and now a complete team of specialists is available for patients with cleft lip and palate.

Fatal growth week 4-12
The lips, jaws and palate are formed in the fetus during the second and third months of pregnancy. They are made from parts of the face that gradually grow together and merge. Sometimes there is an incomplete union and a cleft is formed. Clefts can be formed in different parts of the face, but the most common place of the cleft is the lip, the alveolar ridge (jaw) and the palate. Sometimes the cleft is complete, going through all these structures, and sometimes only part of the lip or palate is affected. The cleft can also be unilateral (one-sided) or bilateral (two-sided), thus the clefts can appear in many different forms.

The treatment consists of surgery, jaw orthopedic-orthodontic treatment and speech therapy.
There are well-established surgical methods for all types of clefts. In most cases, it means multiple surgical procedures on different occasions. Between the surgical interventions, other kinds of treatment can be performed such as speech therapy and jaw orthopedic- and orthodontic treatment.

Which children can get a cleft lip and palate?
There is no way to accurately predict which child is going to be born with a cleft malformation. However, it is known that:
- One child out of 500 will be born with a cleft.
- Parents who themselves have a cleft malformation; have a higher risk than others to have a child with a cleft.
- For parents that have one child with a cleft malformation, the risk that the next child will also have a cleft is somewhat higher. Nevertheless, that risk is still rather small.
Why has the child been affected by this malformation?

The reason is not yet quite clear
It is difficult to explain why a child has a cleft lip and palate. We know that there are hereditary factors contributing, but we do not know the exact causes. Heredity does not follow simple patterns. Hereditary traits can be hidden and may not appear in every generation. In order for a cleft to form, hereditary factors have to coincide with other factors. These relationships are difficult to understand.

General care

Feeding of children with clefts
Feeding of children with cleft lip and palate should be performed in a way that works best for the child. The same kind of food as other children of their age should be given. The cleft child has the same reflexes for sucking and swallowing as other children but because of the cleft, they will have difficulties in sucking. If the child has a cleft in the lip and jaw it might be possible to breast-feed. This can also be possible if there is a small palatal cleft, only affecting part of the soft palate. In patients with larger clefts, bottle-feeding is necessary. Sucking is the base for the development of movements of the lip and mouth. Cleft children may need extra help to start sucking and to develop a normal pattern of sucking and swallowing.

Stimulating sucking and swallowing will promote the development of movements of the lip and mouth.

Bottle-feeding
The type of the nipple is important. In order to make it similar to breast function during breastfeeding, a nipple with broad base and 5 to 6 small holes is recommended. Start with a soft bottle that can be lightly squeezed when the child makes sucking movements. The food to be given in suitable amounts each time. Using a breast pump, the milk from the mother can be collected and used. Feeding is best facilitated if the child is kept in a more vertical position, in order not to fill the nose with food. The broad base of the nipple is pressed towards the lip/alveolus of the child and directed slightly downwards. In children with bilateral clefts, the nipple must be positioned in the midline and unilateral clefts, from the cleft side towards the non-cleft side in the mouth. Feeding usually takes longer than with non-cleft children. When the nipple has been used for a while the holes may become too large and should be replaced.

Most often feeding with a soft bottle is the best alternative.

After feeding, the nose should be carefully cleaned. The easiest way is with water and cotton swabs.
How will the treatment be performed?

Information about the treatment plan will be given at the consultation with the Plastic Surgeon.
The paediatric doctor will refer the child to the Department of Plastic Surgery. At the Department of Plastic Surgery, they have much experience with cleft patients. Each child will have an individual treatment plan, depending on the type of cleft. Patients with a cleft only in the lip or palate will come for a consultation at the out patient clinic. The plastic surgeon will then inform them about the malformation and the treatment. Patients with complete clefts of the lip jaw and palate will have an appointment at the Department of Plastic Surgery during the third week of life. They will stay at the hospital for a day and after the clinical examination, the parents will get information about the malformation, the general care and the treatment plan. During this day, there is also a consultation with the orthodontist, who will inform them about future treatment. In some cases, an impression for plaster models is taken. This model is used for evaluation of the complexity of the cleft and also for help in the planning of the treatment. This first appointment also includes a visit to the speech pathologist who will give information about speech, feeding and future follow-ups.
Surgery

Surgery is performed in several steps and is somewhat different depending on the type of cleft.

1. Cleft lip:
   (Clefts in the lip only)

Lip-nose repair
In patients with clefts in the lip only, the surgery is performed as early as 6 months of age. Sometimes secondary corrections may be needed. Decisions about secondary corrections are made at one of the regular follow-ups that take place up to adulthood. It can be performed at five years of age, or later.

Lip-nose repair

The result at three years of age
2. Clefts in the lip and alveolus (jaw)

**Lip-nose repair**
In patients with clefts in the lip only, the surgery is performed as early as 6 months of age. Sometimes secondary corrections may be needed. Decisions about secondary corrections are made at one of the regular follow-ups that take place up to adulthood. It can be performed at five years of age, or later.

**Bone grafting to the alveolus**
At around 8-9 years of age bone grafting is performed from the tibia to the cleft, in the alveolus. At this age the lateral incisor or the canine erupts into position in the cleft region. If bone is missing, the tooth cannot erupt properly. After bone grafting to the cleft, the tooth can grow and erupt into the grafted bone and be brought into position by orthodontic means. **Further corrections can be performed in the late teens**

The patients are followed regularly, up to adulthood, and if secondary corrections are needed they can be discussed, the patients are informed and can participate in the decision about suitable surgical procedures.

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**Lip-nose repair**

*Result at three years of age*
Bone transplant to the alveolus

Result at ten years of age

Child with a cleft in the lip and alveolus

The same child at age 15
3. Cleft palate (Clefts in the palate only)

Soft palate closure
Hard palate closure performed in a separate operation

In patients with isolated palatal clefts, the soft palate is operated on at 6 months of age. In those patients who have a cleft that also involves the hard palate, a residual cleft will persist in the hard palate. As the patient grows, the cleft in the hard palate will decrease in size. Around 3 years of age, the size of the hard palate cleft is often very small and can be closed by a minor operation.

If a complete cleft in the hard and soft palate is closed in the first stage, several studies have shown that growth of the maxilla may be affected. This may result in crowding of the teeth, malocclusion and possibly restricted midfacial growth.

Operation to improve the speech:

An operation to improve the speech is done occasionally, most often a velopharyngeal flap.

Apart from the operations previously mentioned, an operation to improve the speech is sometimes necessary. To elongate the soft palate, a velopharyngeal flap can make it easier for the child to pronounce certain sounds correctly. Most often, it is done before the child starts school.

Soft palate closure
Hard palate closure

**Picture 1 **):

Incision

*A mucosal flap is elevated*

*An incision on the edge of the cleft to create a pouch between the mucosal layer and the bone*

**Picture 2 **):

Bone

*The backside of the mucosal flap*

*The mucosal flap is tucked into the pouch between the bone and mucosal layer.*

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4. **Unilateral clefts in the lip alveolus (jaw) and palate (UCLP)**

*Soft palate closure and lip-nose repair is done in one operating session.*

**Closure of the hard palate is performed in a separate operation**

In unilateral complete cleft lip and palate, a repair of the soft palate, the lip and nose is performed in the first operation at 4-6 months of age.

After surgery, there will be a residual cleft remaining in the hard palate. Studies have shown that early closure of the cleft in the hard palate may restrict the growth of the upper jaw resulting in undesired facial appearance and poor occlusion. Therefore, closure of the cleft in the hard palate will not be performed until 3 years of age. At that age, the hard palate cleft will have reduced its size by growth of the palatal edges and can be closed by a minor surgical procedure.
Lip-nose repair

The result at age 3

Soft palate closure

The result at age 3
Hard palate closure

*Picture 1 *)

Incision

A mucosal flap is elevated

An incision on the edge of the cleft to create a pouch between the mucosal layer and the bone

*Picture 2 **):

Bone

The backside of the mucosal flap

The mucosal flap is tucked into the pouch between the bone and mucosal layer.
Bone transplant to the alveolus
The last planned operation in patients with UCLP will take place at 8-9 years of age, when bone grafting is performed. In that operation, bone is taken from the lower leg and transplanted to the cleft, in the alveolar ridge. The lateral incisor or the canine is, at that time, on its way to erupt into its position in the cleft area. If bone is missing, the tooth cannot erupt properly. After bone grafting it is possible for the tooth to erupt through the grafted bone and can be brought into position by orthodontic means.

Operation to improve the speech:
An operation to improve the speech is done occasionally, most often a velopharyngeal flap.
Apart from the operations previously mentioned, an operation to improve the speech is sometimes necessary. To elongate the soft palate, a velopharyngeal flap, can make it easier for the child to pronounce certain sounds correctly. Most often it is done before the child starts school.

More corrections might be necessary in the late teens
After bone grafting, the planned surgical treatment is finished. However, the patients will be followed regularly at 10, 13, 16 and 19 years of age. On these occasions, the results from earlier operations are discussed and the patient is informed about possible future secondary corrections.

Bone transplant to the alveolus

The result at age 9 and 16
5. Bilateral clefts in the lip alveolus (jaw) and palate (BCLP):

**Lip adhesion is the first closure of the lip**
In complete bilateral cleft lip and palate the first operation takes place at 6 – 8 weeks of age. At that time a lip adhesion is performed. The patients must weigh over 4 kilos and be otherwise healthy. The aim of this first surgical procedure is to create a normal looking lip. Corrections of the nose will also be the result of this first operation. If the patient’s premaxilla is protruding, the lip adhesion has to be performed in two stages; first on one side then the other. The second time it will be done in conjunction with the soft palate closure.

**Soft palate closure**
The next operation is the soft palate closure and it is performed at 6 months of age. In this procedure, only the posterior part of the palate is closed. Thus, after surgery, there will be residual clefts remaining in the hard palate. Final lip-nose repair in patients with bilateral cleft lip and palate should be performed at one year of age. In order to support symmetrical growth of the nose during the early years of childhood, sometimes a nose conformer is used one week after the operation. This stage is important in assuring an aesthetic result of the nose. Immediately after surgery, the scar in the lip is often red and swollen. There might also be a contraction in the scar. This is a result of scar tissue and will gradually be resolved. However, it may take one to two years before the scar is soft and it is possible to see the outcome.

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**Operation 1**

*Lip adhesion of a child with bilateral cleft lip and palate at 2 months of age*

(Soft palate closure is performed at 6 months of age)
Soft palate closure

The result at age three
Hard palate closure

**Picture 1 **):

Incision

A mucosal flap is elevated

An incision on the edge of the cleft to create a pouch between the mucosal layer and the bone

**Picture 2 **):

Bone

The backside of the mucosal flap

The mucosal flap is tucked into the pouch between the bone and mucosal layer.
**Hard palate closure**
When the patient is 3 years old and the residual clefts in the hard palate are visible as two small openings, they can be closed by one or two minor operations. Studies have shown that early closure of the cleft in the hard palate may restrict the growth of the upper jaw. This can cause undesirable facial appearance and bad occlusion of the teeth. Therefore, closure of the clefts in the hard palate should not be performed until 3 years of age. By this age, the hard palate clefts are smaller, due to growth of the palatal edges. This delay in surgery allows closure of the hard palate to be a minor surgical procedure.

**Lip-nose repair**
In patients with a bilateral cleft lip and palate, corrections of the lip and nose, are performed at one year of age to create symmetry and balance. The nostrils are often allowed to be a little wider than normal, if the nostrils are made too small, it is difficult to perform secondary corrections later on in life. To further support a symmetrical growth and postoperative healing, a nasal conformer is fitted. Postoperatively the scarring of the upper lip is red and bulky. As the scarring of the upper lip heals, the bulk and reddishness will go away. It can take up to two years before the final outcome is visible.
Bone transplant to the alveolus
The last planned operations in patients with BCLP, will take place at 8-9 years of age, when bone grafting to the alveolar ridge is performed. This is done in two operations; first to one side of the upper jaw, then to the other. Bone is taken from the lower leg and transplanted to the cleft in the alveolar ridge. The lateral incisor or the canine is, at that time, erupts into its position in the cleft area. If bone is missing, the tooth cannot erupt properly. After bone grafting it is possible for the tooth to erupt through the grafted bone and can be brought into position by orthodontic means.

Further corrections can be performed during the teenage years.
After bone grafting, the planned surgical treatment is finished. However, the patients will be followed regularly at 10, 13, 16 and 19 years of age. On these occasions, the results from earlier operations are discussed and the patient is informed about possible future secondary corrections.
After completing the planned surgeries, the patients will be followed regularly at 10, 13, 16 and 19 years of age. On these occasions, the results from earlier operations are discussed and the patient is informed about possible future secondary corrections.

Sometimes operations will be performed to improve speech.
If speech is impaired, after the primary operations, a lengthening of the palate may be indicated. This can be done by a local procedure within the palate itself or by a so-called palatal-pharyngeal flap, where the palate is connected to the posterior wall of the pharynx by a flap in the midline. If necessary, this procedure is performed, before the child starts school.

Bone transplant to the alveolus is done to one side at the time

The result at age 10
### Surgical treatment of children with Cleft Lip and Palate

<table>
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<tr>
<th>Type of cleft</th>
<th>Information</th>
<th>Operation 1</th>
<th>Operation 2</th>
<th>Operation 3</th>
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<th>Operation 5</th>
<th>Operation 6</th>
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<td>Lip (L)</td>
<td>Within one month in an out patient visit</td>
<td>6 months Lip-nose repair</td>
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<td>Lip-alveolus (L-A)</td>
<td>Within one month in an out patient visit</td>
<td>6 months Lip-nose repair</td>
<td>8-9 years Bone-transplant</td>
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<td>Palate (P)</td>
<td>Within one month in an out patient visit</td>
<td>4-6 months Soft palate closure</td>
<td>3 years Hard palate closure</td>
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<td>Uni lateral cleft lip and palate (UCLP)</td>
<td>1-3 weeks Admitted to the hospital</td>
<td>3-5 months Lip-nose repair and soft palate closure</td>
<td>1-3 years Hard palate closure</td>
<td>8-9 years Bone-transplant</td>
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<td>Bilateral cleft lip and palate (BCLP)</td>
<td>1-3 weeks Admitted to the hospital</td>
<td>1,5-2 months Lip adhesion one side</td>
<td>5-6 months Soft palate closure and lip adhesion</td>
<td>1 year Lip-nose repair</td>
<td>3 years Hard palate closure</td>
<td>8 years Bone-transplant</td>
<td>9 years Bone-transplant</td>
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Ears and hearing

**Hearing is important for the development of speech and language and should be tested early and regularly.**
Children with cleft lip and palate have a high risk for infection of the middle ear. Therefore, hearing has to be tested early and immediate treatment implemented, if hearing is affected.

**Children with cleft lip and palate have a high risk for infection of the middle ear.**
In children with cleft lip and palate, the muscles in the cleft palate are not working properly and the function of the Eustachian tube may be affected. The Eustachian tube connects the pharynx and the middle ear, regulating the pressure of the middle ear. An equal and normal pressure is a prerequisite for normal hearing.

**Chronic inflammation of the middle ear can develop if the Eustachian tube is not functioning properly.**
If the Eustachian tube is not functioning properly, chronic inflammation can arise. If this is the case a negative pressure can develop in the middle ear and liquid will be produced. On examination, the liquid can be seen behind the eardrum and the negative pressure can be measured with a simple method. The chronic inflammation will cause impaired hearing, varying in severity. It is common in children with cleft lip and palate but can also be present in children without any malformation.
Often the chronic inflammation will disappear spontaneously, but sometimes use of drugs that reduce swelling have to be used.
If chronic inflammation remains for a long time, the eardrum has to be punctured, the liquid removed by suction and the pressure normalized. Sometimes the hole in the eardrum has to be kept open for a longer period time, to prevent build up of new fluid.

**Treatment with grommets**
In order to maintain the hole in the eardrum, a plastic tube (grommet), is placed and has to remain for some months. This treatment has to be performed under general anaesthesia.

**Impaired hearing due to chronic inflammation can be difficult to detect**
It is sometimes difficult to identify chronic inflammation of the middle ear. The child may not complain of pain. Since this is common problem, it is important that an ENT doctor check the ears and hearing regularly, this should be done even if the child does not appear to have problems. Ear problems most often disappear after palate surgery or as the child grow.

**Impaired hearing due to a middle or inner ear malformation**
Some of the children with clefts of the palate, have malformations of the middle and/or inner ear. This should be evaluated early with auditory testing and in some cases also with a computerized x-ray (CT).
If it is discovered that the hearing is impair due to a malformation, there are different hearing aids or operations to facilitate the hearing.
If the Eustachian tube is blocked, a liquid is formed in the middle ear. The liquid is removed when the grommets are placed.

Speech

The speech pathologist can provide, from an early age, information about feeding, oral motor function and speech.

The local speech pathologist will see the newborn child at the hospital and give the first information about feeding and further follow-ups. The family then will get more information about:

- Feeding and development of oral motor function
- Speech and language development
- How a cleft can affect speech

The local speech pathologist will see the patients on regular follow-ups until adulthood.

If the cleft includes the palate, a speech pathologist on the cleft team, will see the child around the age of one, to provide information and evaluate babbling.

The lips, tongue and the soft palate have important functions in speech. The soft palate closes the passage between the oral and nasal space, and is used in pronunciation of most speech sounds. It is necessary to be able to move the lips, tongue and palate fast and with precision in order to have correct and clear articulation.

The child will improve speech development if:

- a normal pattern of sucking and swallowing is established.
- extra stimulus for the tip of the tongue and the lips
- the lips and the soft palate can be operated upon early.

A remaining cleft in the hard palate (residual cleft) can make it more difficult to develop normal articulation and can result in incorrect patterns of speech. The child has a good chance of spontaneously correcting these anomalies when the residual cleft is closed.
A normal pattern for suction and swallowing, extra stimulus of anterior mouth movements and early operation of the soft palate, improves the development of the pronunciation of the speech sounds.

The child will be seen for further follow-ups by the local speech pathologist. Some children with cleft lip and palate will need speech therapy to correct articulation and also develop speech and language. Such speech therapy can start at around 3 years of age, sometimes earlier. Speech and language is also improved by playing and by interacting with people around the child. Speech pathologists will provide information to the parents, teachers and staff in day-care centres.

Patients with cleft lip and palate will also be checked regularly at the Department of Speech Pathology at Sahlgrenska University Hospital. These follow-ups will be performed in conjunction with visits for surgery or other appointments with surgeons or orthodontists. The speech pathologist will record the speech and evaluate it and the function of the palate.

**Recordings and evaluation of the speech and the function of the palate are performed by a speech pathologist at the University Hospital on several occasions in conjunction with follow-ups by the plastic surgeon and the orthodontist.**

Sometimes a surgical procedure, lengthening the palate, is necessary due to speech problems. Before this operation can be performed, the functions of palatal movements and swallowing have to be investigated. Speech pathologist, dental radiologists and ENT doctors specialized in speech are all involved in this examination.

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<th>Normal sound production</th>
<th>Altered sound production in a cleft patient</th>
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Teeth

Orthodontic treatment will start at 7-8 years of age. The final phase of orthodontic treatment is usually performed when the patient is 12-14 years of age.

From early childhood, an orthodontist is engaged in evaluating growth and development of the jaws, teeth and occlusion. Children, in need of a bone transplant often require active orthodontic treatment prior to the operation. The next phase of orthodontic treatment is usually implemented when all the permanent teeth have erupted (12-14 year). Orthodontic treatment is usually done with fixed orthodontic appliances. If there are teeth missing in the cleft region, this can be treated by prosthesis or the space can be closed by using orthodontics. The prosthodontic treatment will not take place until the patient is 18–20 years old. A titanium implant or a conventional crown and bridge can be used.

The final orthodontic treatment is performed when all the permanent teeth have erupted.
Quality assurance

All the cleft centres in Sweden participate in a common quality assurance program, the National Register for Quality Assurance for Cleft-Lip and Palate Treatment. All cleft children treated or evaluated at the Swedish cleft centres are registered. The immediate outcome and late complications of surgery are registered as well as mid-facial growth, occlusion and speech. All data are stored in a centralized register.

GÖTEBORG TEAM

The cleft team in Göteborg consists of:

PLASTIC SURGERY: Jan Lilja, Anna Elander, Pelle Sahlin

ORTHODONTICS: Lars Enocson, Catharina Hagberg

SPEECH: Anette Lohmander, Christina Persson, Christina Havstam

HEARING: Birgitta Wahlström, Ann-Charlotte Axelsson