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## OVERVIEW

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# Guidelines for the treatment of cleft lip and palate

About 120 babies are born with cleft lip and palate in Finland each year. Most cases consist of isolated cleft palate (60%), while less than one in four cases suffer from complete cleft lip and palate. In Finland, clefts are closed before the child's first birthday. Clefts affect appearance, occlusion and speech, and the final outcome of treatment can only be assessed at the end of the child's growth. Centralisation of services and a multidisciplinary team approach have a bigger influence on the final outcome than different treatment protocols, the comparative advantages of which remain unproven. Good documentation of results is important to assess the outcomes of treatment.

Cleft lip and palate is the most common congenital structural deformity. Cases can be divided into two main groups: cleft lip with possible cleft palate, and pure cleft palate (Figure 1). The former may be unilateral or (more rarely) bilateral. The severity of clefts varies widely. The most difficult cases involve considerable deformities in bone structure, soft tissues and nasal structures.

The prevalence of clefts in Europe is generally 15–20/10,000, but 25.3/10,000 in Finland. The larger incidence in Finland is explained by the large number of cleft palates. The prevalence of cleft palates in Finland is 14.3/10,000, whereas the European average is 6/10,000. The frequency of cleft palate in Finland is probably due to as-yet unknown genetic factors (Koillinen 2003). Cleft palate is more common in the area that is termed the new settlement, i.e. Eastern and Northern Finland, whereas the distribution of clefts in the older settled areas of Finland is closer to the general European distribution (WHO 2002 and 2003, Ritvanen and Sirkiä 2007). No less than 60% of all clefts in Finland are cleft palates, while 25% are a combination of cleft lip and palate and 15% cleft lips. Cleft palate is more common in girls, whereas combined cleft lip and palate is more prevalent in boys. Approximately 120 infants are treated for clefts in Finland each year, slightly more than a hundred of these by the Lip and Palate Centre (Husuke) of HUCH (Rautio and Hurmerinta 2000).

A cleft can have detrimental effects on appearance, eating, speech and occlusion, as well as causing ear and dental complications (Hukki et al. 1999). The disorder may also cause social impairment, particularly if treatment was not successful (Marcusson et al. 2001). The expert team at Husuke includes plastic surgeons, maxillofacial surgeons, an otorhinolaryngologist, speech therapists, orthodontists, a prostheticist and nurses trained in the treatment of clefts. The centre consults specialists from other areas, such as phoniatrics or medical genetics, where necessary. We seek to arrange all required examinations during a single outpatient clinic visit. Periodic examinations conducted every one or two years during the patient's childhood and youth document the outcome of the treatment. Any required further treatment is also planned and included in the overall treatment plan during these examinations. Most non-surgical treatment, such as orthodontics, speech therapy and ear treatments, are implemented in the patient's own hospital district.

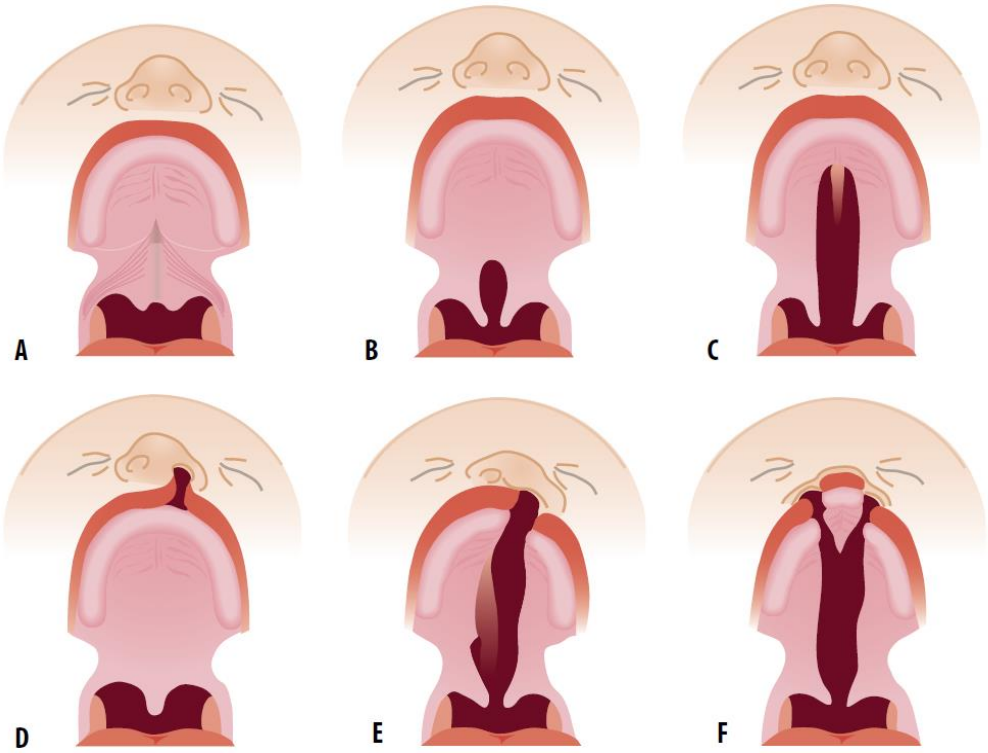


Figure 1. Cleft types: A) submucous cleft palate, B) cleft soft palate, C) complete cleft palate, D) cleft lip, E) unilateral cleft lip and palate and F) bilateral cleft lip and palate.

## Heredity and syndromes

In addition to environmental factors, several genes affect the creation of a cleft. Most of these genetic factors remain unknown. Maternal smoking and use of alcohol and some anticonvulsants – sodium valproate in particular – are known to contribute to clefting (Lorente et al. 2000, Artama et al. 2005). Clefts can also be divided into two categories with regard to heredity: 1) cleft lip or cleft lip and palate, and 2) pure palatal clefts. Pure palatal clefts are inherited as cleft palates, but inherited cleft lip can also involve a cleft palate. The hereditary risk of children of parents with a cleft or the siblings of children with a cleft is approximately 4%; the risk is significantly smaller (0.6%) for second-degree relatives (Harper 2004, Sivertsen et al. 2008). If more than one family member has a cleft, this increases the risk of inheritance.

Potential syndromes must be taken into consideration when determining the inheritance risk of a cleft since, according to certain studies, up to 30% of cleft lips and palates and 50% of pure palatal clefts may be related to a syndrome (Marazita 2002, Calzolari at al. 2007). More than 400 such syndromes caused by chromosome disorders, genetic defects and teratogens have been documented. Cleft-related abnormalities most often manifest in the central nervous system, heart, bone structure or limbs (Calzolari et al. 2007). If a child has other abnormalities in addition to the cleft, a consultation with a medical geneticist is recommended in order to identify any underlying syndrome through further examinations. The parents of a clefted child may also benefit from such a consultation, since it can eliminate unwarranted guilt related to the pregnancy.

## Nutrition after birth

The discovery of a cleft in an infant frequently caused feelings of anxiety and even guilt in the parents. The identification of cleft lip or palate is not an objective of the structural ultrasound examination performed after the 18th week of pregnancy (Reports of the Ministry of Social Affairs and Health 2009), but extensive clefts are sometimes discovered at this stage. In some countries, the foetus is actively examined for clefts in order to inform the parents during pregnancy. It is difficult to say whether this practice is better than that observed in Finland.

When a cleft is discovered after delivery, the family can immediately receive guidance and support through good communication in the maternity hospital and later in the cleft centre. Feeding difficulties are very common in the first weeks after childbirth. Cleft palates in particular prevent the infant from creating negative pressure in the oral cavity, which means that breast feeding is rarely successful. The infant can be fed from a bottle with the hole in the teat enlarged by a cross-cut, with sucking assisted by squeezing the plastic bottle or base of the teat according to the child's feeding rhythm. Most infants learn to feed in this manner, but various specialised teats are also available for feeding bottles. Learning to feed the baby requires patience and perseverance from the feeder. Resolving any feeding difficulties will reinforce the positive interaction and affection between the parents and child.

## History

The pioneer of Finnish surgery, Rikhard Faltin (1867–1952), expressed concern for the poor treatment of clefted children as early as the first years of Finland's independence. In 1935, he finally convinced his youngest assistant, Atso I. Soivio, to travel to Paris to study the treatment of clefts. The beginning of centralised treatment was delayed by the war, but the Finnish Red Cross Plastic Surgery Hospital was eventually opened in 1948. In 1975, the unit's name was changed to the Cleft Lip and Palate Centre (Husuke), and the unit was transferred to HUCH in 1984 (Rintala 1998). Oulu University Hospital (OYS) started primary surgery in its area in 1998. The hospital follows its own treatment programme, which is not discussed in this article. In 2001, the EU's Eurocleft working group issued a recommendation on the centralisation of cleft treatment in European countries. The Decree of the Ministry of Social Affairs and Health 767/2006 nevertheless divided the primary surgical treatment of clefts between HUS and OYS, with the exception of cleft lip operations.

## Operations during the first year of life

The principal outcome criteria for the surgical treatment of clefts are good appearance, speech capabilities and occlusion. The surgical treatment of clefts at various centres is highly varied, since very few controlled studies have been conducted on the matter (Shaw et al. 2000, WHO 2002). Despite good results achieved through various programmes, the optimal surgical procedure and timing remains unknown. It would indeed be very difficult to draw up Current Care Guidelines for the treatment of clefts.

***Cleft lips*** are closed at the age of 3–6 months. One in three infants with a cleft lip also have a cleft in the gums, usually in the area of the lateral incisor. Such clefts are repaired by a bone graft after the eruption of permanent teeth, at around 9–11 years of age (see bone graft).

***Cleft palates*** are closed at Husuke at roughly nine months of age, but the timing of the operation can vary between six months to up to two years of age at different centres. A cleft palate is closed by releasing the mucous flaps of the soft and hard palates and closing the cleft with sutures. In the past, the release cuts were always made on the sides of the palate to avoid stretching (von Langenbeck 1861) (Figure 2). The lateral wounds were left rather open and allowed to scar. Later practice involved lengthening the soft palate by lifting flaps from the hard palate and sewing them back on, which left an even larger scar on the palate. This technique has nevertheless been abandoned, since it resulted in growth defects. Current practice is to seek to repair the muscles of the soft palate as fully as possible. This is believed to improve the development of speech, although there is no reliable evidence.

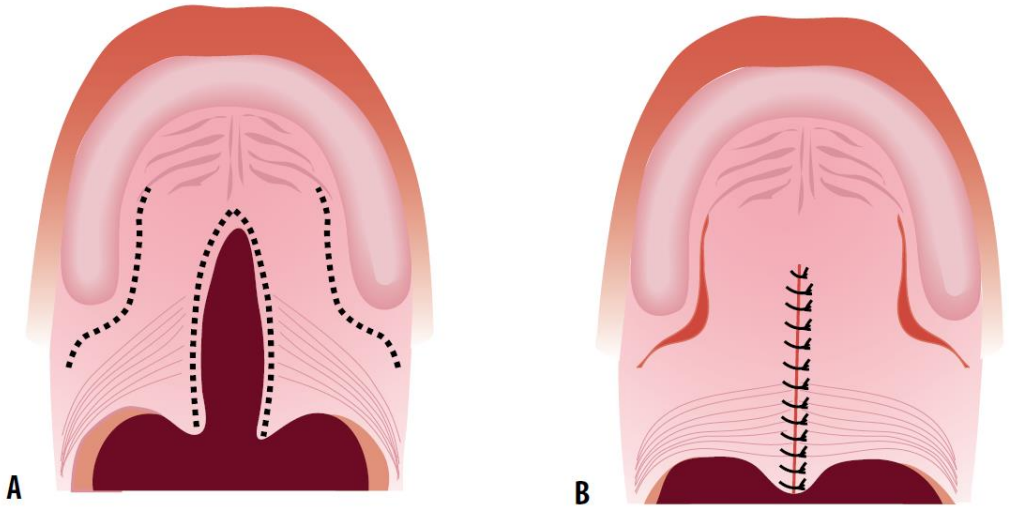


Figure 2. A cleft palate can often still be closed according to the basic principle of von Langenbeck, in use since 1861. The muscles of the soft palate are repaired. Lateral cuts are rarely necessary.

***Unilateral cleft lips and palates*** are currently usually closed at Husuke by first closing the clefts in the lip and hard palate at four months of age. The soft palate is then closed at nine months. Other methods for treating these clefts exist as well. Some physicians prefer to close the soft palate together with the cleft lip, postponing the closure of the hard palate by several years in some cases. It is also very common to close only the cleft lip at the first stage and the entire palate at the second. Closing the entire cleft in a single operation is also a possibility. We do not know what sequence is best with regard to upper mandible growth and the development of speech. Similarly to cleft lips and gums, cleft gums are corrected by bone grafts at the age of 9–11 (see bone graft).

***Bilateral cleft lips and palates*** are closed according to the same general principles as unilateral clefts. Reconstructing the nose can be difficult in both unilateral and bilateral clefts, and additional operations are frequently required. Rhinoplasty is nearly always required for these patients in order to correct the low and wide nasal profile. Nasal support structures are repaired with cartilage grafts at six years of age or later.

## Later operations

Further operations are seldom required after primary surgery in cases of cleft lip or pure cleft palate. Cleft lips and palates, on the other hand, require additional operations more often. Good primary surgery can nevertheless significantly reduce the need for secondary corrective surgery. The surgical burden for patients has decreased considerably in comparison to previous decades. Those born with unilateral cleft lips and palates as recently as the 1960s and 70s required an average of eight operations by adulthood. Today, the number of operations required has been halved. The minimisation of the treatment burden is indeed one of the key goals of modern cleft treatment. This saves public funds and spares the young patient from unnecessary hospital experiences.

## Speech surgery

Monitoring the development of speech is important after primary surgery. The speech of children with corrected cleft palates may initially be less clear and more nasal than that of their peers. However, the speech of many patients is corrected autonomously or with the help of speech therapy. Approximately one in five of such children later require pharyngeal flap surgery to improve speech. Husuke previously employed a surgical procedure whereby the nasopharynx was tightened by sewing a wide flap from the laryngopharynx into the soft palate. However, the procedure frequently caused snoring and sometimes even sleep apnea. If adult cleft patients develop such complaints, a consultation is in order. Our primary method is currently the extension of the soft palate using the Furlow procedure (Figure 3). This normalises the anatomy of the nasopharynx and reduces congestion.

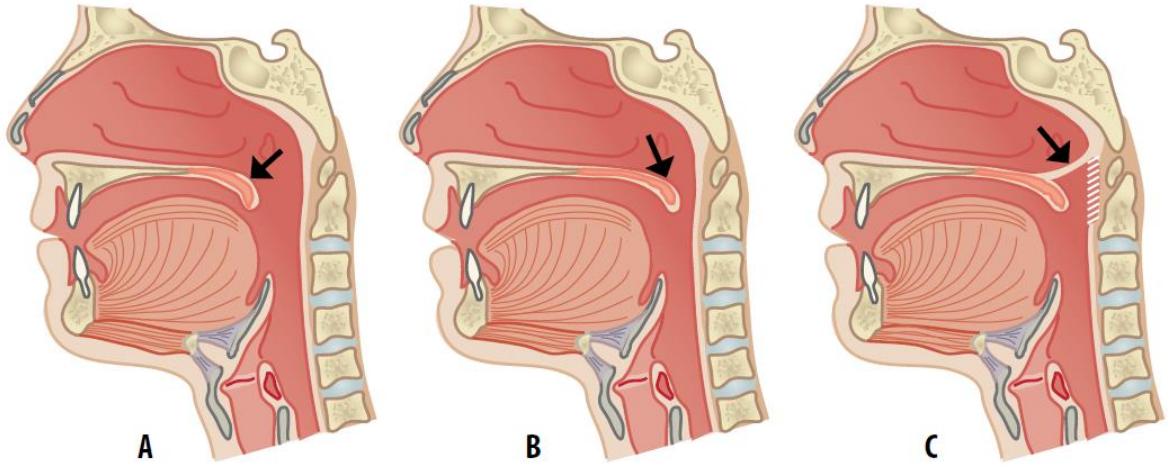


Figure 3. A) After the palate has been closed, the soft palate does not extend far enough to close the nasopharynx. B) The soft palate has been extended using the Furlow procedure. C) The nasopharynx has been tightened with a flap raised from the laryngopharynx.

## Bone graft

In unilateral and bilateral cleft lips and palates, as well as in cases of cleft lip where the cleft extends into the gums, a bone graft procedure is performed in the area of the cleft gum, usually at 9–11 years of age (Figure 4). The purpose of the procedure is to repair the bone deficiency in the alveolar ridge and nasal base and to secure the eruption of

permanent teeth in the area of the cleft. The graft usually consists of spongy bone taken from the ridge of the iliac bone. Additional corrective surgery of the lip and nose is combined with the operation if necessary. Orthodontics are nearly always required after a bone graft.

## Surgical treatment of maxillary growth defects

Roughly one in four patients with a cleft lip and palate develop defects in the growth of the upper mandible and midface. The resulting severe malocclusion can have a major detrimental impact on mandible function and facial appearance, which can be psychologically difficult for teenagers. A growth defect in the upper mandible cannot be corrected through orthodontics alone, but orthognathic surgery is required to correct the alignment of the mandibles.

An upper mandible growth defect is most often treated by Le Fort l osteotomy (Figure 5). Distraction of the upper mandible, whereby the upper mandible is gradually pulled to the desired position, is also a possibility. Severe growth defects may require both procedures: distraction during the growth stage and osteotomy towards the end of growth. A lower mandible osteotomy is sometimes also required to correct the facial structures. In addition to correcting occlusion, particular attention is paid to nose and lip profile and the nasolabial fold in the treatment of clefted patients. The nasal deformity typical of cleft patients may become more pronounced as a result of the Le Fort 1 osteotomy. A thorough rhinoplasty operation is thus often performed at this stage.

## Ear and hearing problems

Nearly all children with cleft palates develop middle-ear aeration problems. The abnormality of the palatal muscles interferes with the operation of the Eustachian tube, which typically results in glue ear. The effusion in the middle ear causes conductive hearing loss. There is disagreement on the treatment of glue ear, including for children with clefts (Ponduri et al. 2009). The usual recommendation is to monitor glue ear in children for a few months before undertaking any operation. With clefted children, however, glue ear is often caused by structural and functional abnormalities, and the situation is unlikely to change over a short monitoring period. For this reason we recommend early myringotomy and tube insertion, usually in connection with the cleft lip or palate operation. Tube insertion aerates the middle ear and normalises hearing, which facilitates the development of speech. The child's growth and surgical treatment of the cleft usually improve the aeration of the middle ear by school age (Valtonen et al. 2005).

Hearing defects related to the inner ear or ossicles are rare in clefted children, but nevertheless more common than in other children (Chen et al. 2008). If a child with a cleft is suspected to be hard of hearing and the middle ear is aerated normally, he or she should be referred to audiometric examinations. The ears of a child with a cleft palate are monitored at Husuke, by specialist medical care in the child's municipality of residence or at the health centre. Follow-up appointments after tube insertion can be made as for other children, but routine follow-up examinations of the ears are recommended until the age of eight, and beyond if any problems arise.

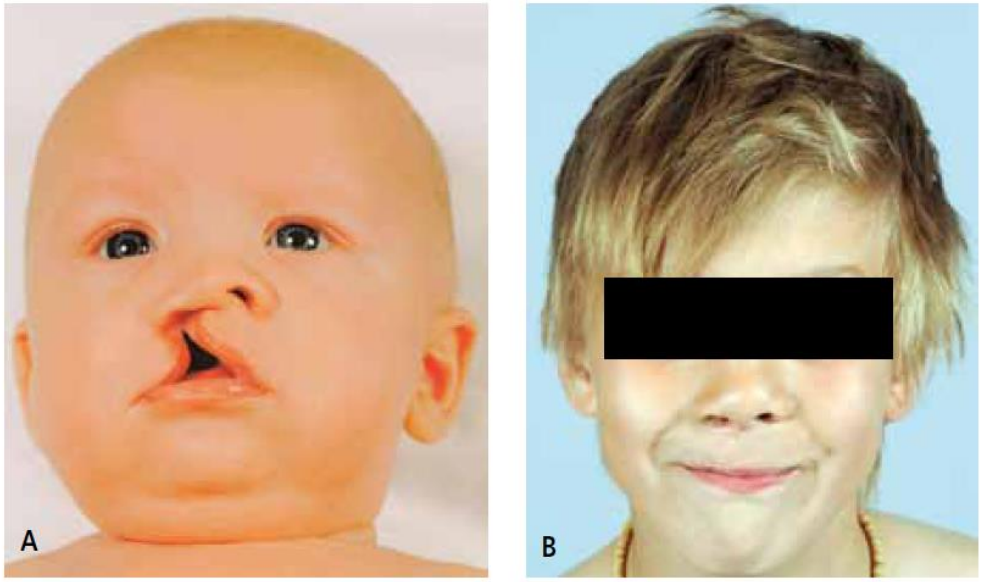


Figure 4. A) Complete cleft lip and palate. B) The same boy at bone graft age.

## Orthodontic treatment

Cleft lips and palates frequently involve dental abnormalities, malocclusion and maxillary growth defects. In order to correct these defects, nearly all clefted children require orthodontic treatment at school age. Prosthetics and orthognathic surgery may also be required (see Surgical treatment of maxillary growth defects). Orthodontics will also be requiredin connection with a bone graft operation.

Typical dental abnormalities related to clefts include deviations in the number, structure and form of teeth, as well as issues related to teeth eruption. Disorders are more common in permanent than primary teeth. The likelihood of abnormalities in permanent teeth increases in proportion to the original extent of the cleft. Approximately one in five patients with cleft lips and more than half of those with cleft lips and palates lack some permanent teeth (Ranta 1986). Teeth deficiencies can be corrected either by closing the gaps through orthodontic treatment or replacing the missing teeth prosthetically. As a rule, the severity of malocclusion also increases in proportion to the original extent of the cleft.

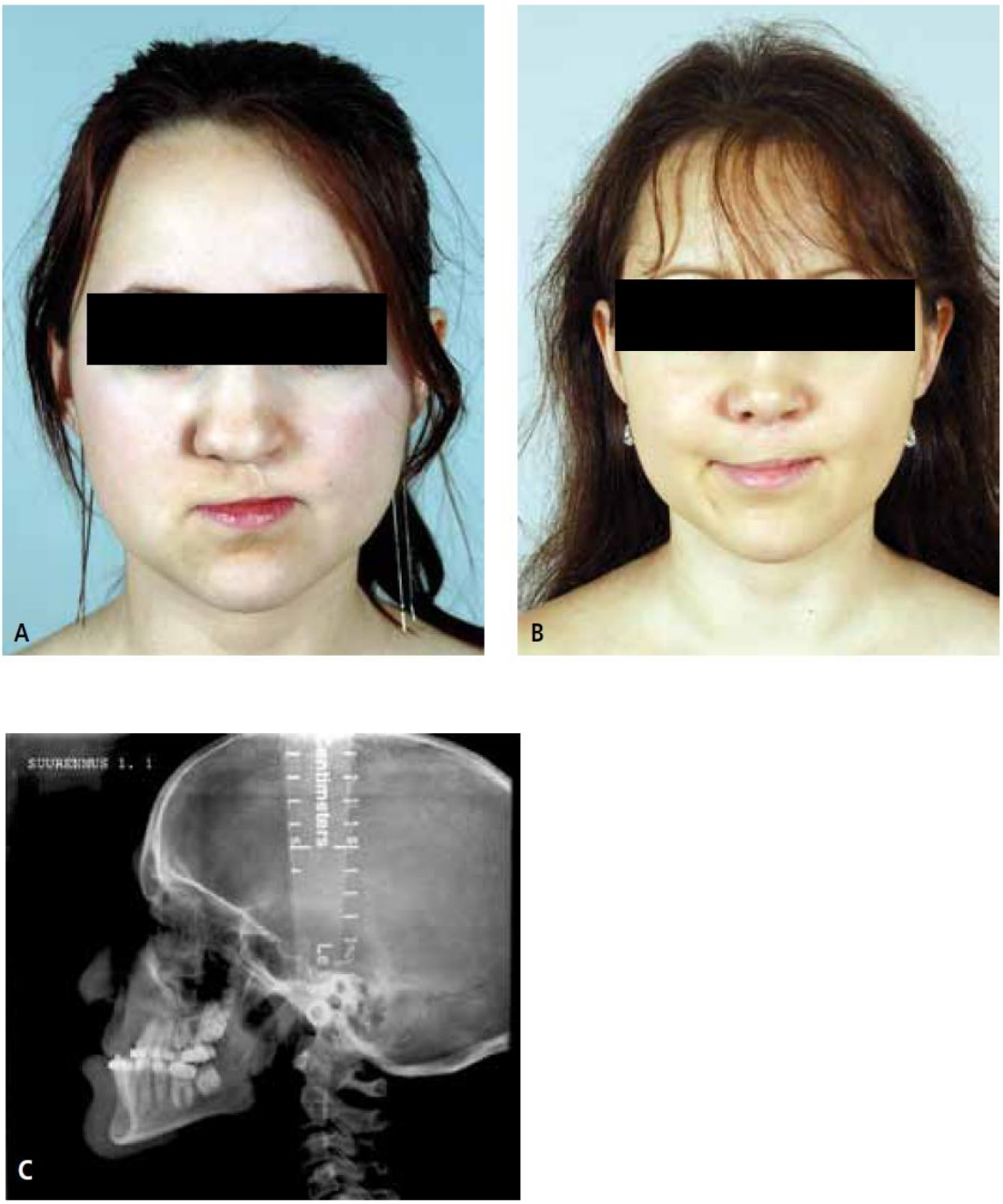


Figure 5. A patient with a cleft lip and palate who has developed a growth defect of the upper mandible. A rhinoplasty with cartilage grafting has been performed in connection with the osteotomy. A) Before the Le Fort 1 osteotomy. B) Six months after the Le Fort 1 osteotomy. C) Lateral radiographic image of the skull before the operation.

Typical types of malocclusion include incorrect tooth positions in the area of the cleft, crowding and defective development of the upper mandible, which can lead to anterior and lateral overlapping (Semb and Shaw 1996).

# In conclusion

Modern treatment methods nearly always achieve a good functional and aesthetic outcome, provided that the treatments are administered by experienced personnel. In recent years, treatment results have shown marked improvement, treatment periods have shortened and the cost of treatment has decreased.

Different cleft centres follow different treatment methods and timings. No single best treatment protocol exists. Since the mutual comparison of treatment outcomes has proven problematic, a group of North European centres decided to compare their results using harmonised indicators. The survey showed that the best results were achieved in Norway and Denmark, where treatment had been concentrated in the hands of a few surgeons who operated on large numbers of patients. In the UK, where treatment results were the worst, surgeons sometimes only operated on one or two complete cleft lips and palates per year. The key factor for the success of treatment was therefore found to be the experience and skill of the surgeon, not the method or timing of the treatment (Shaw et al. 1992). Based on the results of this survey and an additional study conducted by the Department of Health, most of the 57 centres then operating in the United Kingdom were closed (Bearn et al. 2001). Today, only 12 centres remain in operation, each serving a population base of roughly five million residents. The European Union's Eurocleft programme recommends that each member of a cleft treatment team should treat at least 40 new patients each year (Shaw et al. 2000). This ensures the high quality of surgery results, the clinical experience of the team and a sufficiently extensive long-term monitoring of patient data, which facilitates scientific research and the development of the field. In Finland, the treatment of clefts was centralised at an exceptionally early stage, in 1948.

The Eurocleft cooperation has also led to the first extensive, international, randomised and prospective multicentre studies on the surgical treatment of cleft lips and palates. Husuke participates in this (Scandcleft) and other international research. The comparison and development of the methods used requires systematic and centralised evaluation of the results of surgery, the development of speech and hearing, maxillary growth and occlusion, as well as cooperation with foreign cleft centres. The final outcome of treatment can only be evaluated at the end of the child's growth.

KEY FACTS

- Approximately 120 new cases of cleft lip and palate occur in Finland each year.
- The incidence of cleft palate in Finland is twice that of other countries.
- The centralisation of treatment is vital in order to ensure good treatment results.


## Summary

**Treatment of cleft lip and palate in Finland**

In Finland about 120 babies are born with cleft lip and palate per year. The largest group is those with isolated cleft palate (60%) and only one fourth have complete cleft lip and palate. The clefts are closed under one year of age. Clefts affect appearance, occlusion and speech and the final outcome can only be assessed at the end of the growth. Centralisation of services and a multidisciplinary team approach has a bigger influence on the final outcome than different treatment protocols, the comparative advantages of which remain unproven. Good documentation is important to assess the level of treatment outcomes.

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