

Facial clefts

Facial clefts significantly affect the appearance, they require complex and long-term treatment (clefts + conductive hearing loss , speech disorder, dentition disorders, etc.). Treatment of patients with cleft lip and palate is complex and multidisciplinary in the Czech Republic. A genetic registry was established in 1964, which currently contains more than 4,500 families.

Embryology

The upper lip develops through the fusion of the medial nasal folds and the maxillary folds. A cleft lip occurs when these bumps fail to fuse at 5 weeks of gestation. The floor develops from two foundations - from the primary floor and from the secondary floor. The secondary palate is formed by the fusion of the lateral palatal processes (maxillary cusps) between the 8th and 12th weeks of gestation and forms the basis of the hard and soft palate in the area behind the foramen caecum. The primary palate forms the base of the premaxilla. Cleft palate includes cleft lip and cleft alveolus. ^[1]

Etiopathogenesis

- Among the most common congenital malformations ,
- conditioned multifactorially with a significant share of heredity,
- *exogenous factors*: viruses (rubella , influenza), toxoplasmosis , CMV , hypervitaminosis A + D , some ATBs (tetracyclines, erythromycin), some antidiabetics + antiepileptics , corticoids , X- ray radiation, drugs , organic solvents and other teratogens .

Proportion of causes of origin:

- **INTRINSIC HEREDITARY DISPOSITION** - about 20% of patients
- **EXTERNAL CAUSES** - around 10-15% of patients
- **COMBINATION OF CAUSES** - about 60-70% of patients, when the hereditary disposition manifests itself only through the action of a combination of external causes
- *racial differences*: most common among Japanese and whites, least among American blacks,
- **cleft lip + jaw**: fusion disorder of the maxillary + nasomedial process,
- **cleft palate**: disc fusion deficit (hypoplasia / horizontalization disorder).

Classification

Typical facial clefts

- *Lip* (**cheiloschisis**),
- **lip + jaws** (*cheilognathoschisis*),
- **isolated palatal** (*palatoschisis*),
- **overall** (*cheilognathopalatoschisis*).

Atypical facial clefts

- **Transverse** ,
- **upper middle** (of the nose, upper lip, upper lip with intermaxillary defect),
- **lower middle** (lower lip, lower lip + jaw),
- **oblique** (lip + cheeks, cheeks + lower lid, with cleft palate typical + atypical).



Unilateral cleft lip in a 19-month-old boy

Prenatal diagnosis

Cleft lip can be easily visualized with the appropriate position of the fetal face during prenatal ultrasound examination in the 2nd trimester. Cleft palate is usually not diagnosed by ultrasound (sometimes an atypical range of tongue movement can be detected in the lateral view).^[2]

Therapy

The definitive solution must be postponed until the end of jaw growth . The team of experts providing therapy for a patient with cleft lip and palate consists of a plastic surgeon, an orthodontist, a speech therapist, a psychologist , a maxillofacial surgeon , an otolaryngologist, a geneticist, a pediatrician, a dentist and a social worker.

Orthodontic treatment ideally achieves correction. Unfortunately, this is often not successful and the patient needs to be rehabilitated prosthetically (especially in patients with total clefts). Prosthetic treatment requires a favorable shape, position and dimension of the alveolar arches and can only be started after the patient has finished growing.

Non-invasive 3D analysis is an integral part of prosthetic treatment planning regarding its stability. It gives us a new perspective on the growth and development of the jaw at specific stages of treatment and helps to decide on the sequence and timing of therapy.

Surgical treatment

Basic timing of operations	
Lip and nose reconstruction	from the 2nd day of life
Floor reconstruction	from 6 months
Adding bone to the jaw	when changing teeth
Other operations as necessary	

Neonatal lip surgeries have been common practice in the Czech Republic since 2005 at the Children's Hospital Brno. Another center that operates on cleft lip after birth is FNKV Prague.

Cleft lip

- Stitching of the skin, mucous membrane and muscles of the cleft lip + reconstruction of the nasal floor + closure of any cleft jaw,
- goal: reconstruction of all tk. cleft lip with Cupid's bow preserved,
- lip surgery before 6 months of age,
- for bilateral clefts, restore the orbicularis oris muscle as soon as possible,
- the problem of solving nose deformities.

Cleft palate

- The goal of closing the soft + hard palate against the nasal cavity,
- early closure of the soft palate → speech training,
- the operation consists of suture of the palate in 3 layers (stitching of the nasal mucoperiosteum, muscles of the soft palate and oral mucoperiosteum) + lengthening of the reconstructed palate by its retroposition (favorably affects the quality of speech),
- secondary operations: lengthening of the nasal septum in case of bilateral clefts, bone implantation in the jaw, correction of the nose, lengthening of the palate by the so-called pharyngofixation, etc.



Cleft lip and palate surgery scar - 5-year-old girl

Case report

Patient JV was born on August 3, 1990, from a physiological pregnancy.

Before birth, a diagnosis of **cheilognathopalatoschisis I. sin** was established, after birth then a combination of congenital heart defects called tetralogy of Fallot, during the later course of treatment, an allergy to penicillin was also diagnosed.

At 14 months, the patient underwent surgical closure of the cleft lip. A congenital heart defect, complicating the possibilities of treatment at an earlier age, contributed to the relatively late resolution of the defect.

At the age of 5, orthodontic therapy was started with a removable appliance - modification of the palate plate according to the course of the therapy. At the same age, cleft palate plastic surgery was performed. Vestibulonasal communication, which in pac. discovered after the closure of the cleft palate was surgically closed at the age of 10. All surgical procedures were performed in an antibiotic drape for routine endocarditis prevention. In 2003, when the patient was 13 years old, treatment with a removable orthodontic appliance was completed.

Since the orthodontic treatment did not bring the desired results, a surgical solution of the lateral crossbite was approached and in January 2005, at the age of 14 and a half years, a maxillotomy was performed in the Le Fort I line, the Hyrax appliance was fixed intraoperatively and subsequently achieved by turning the appliance screw in the morning and evening by 1/4 revolution of the displacement of the jaw in the transverse direction by 8 mm. From the age of 15 to 18, the patient underwent therapy with a fixed orthodontic appliance (Roth ring appliance). In 2005 and 2008, bone grafts from the hip or retromolar space and augmented into the cleft defect.

Neither orthodontic nor surgical therapy led to a final result, but provided the conditions for making stable prosthetic work. The patient's parents were advised to close the loco 22 gap using an implant and a prosthetic crown or by making a fixed bridge in the range of 11-24. Rehabilitation using a fixed all-ceramic bridge was chosen.

At the beginning of 2009, the prosthetic rehabilitation of the patient began - teeth 11, 21, 23, 24 were sharpened for a fixed bridge under the supervision of an anesthetist, a temporary bridge was handed over, models were made, and finally an all-ceramic fixed bridge in the range 11-24 was handed over. Since the patient also exposed the gingiva when smiling, the modeled gingival part is also part of the fixed bridge.

The course of the entire therapy was carefully documented by taking x-ray images and plaster models. A total of 5 plaster models of the upper jaw and palate were made in 1991, 1998, 2006, 2008, 2009. The models were evaluated by classical measurement of the transverse growth of the jaw as well as by computer digitization after scanning the models with a scanner. For this measurement, the tips of temporary and permanent canines (dimension 3-3) were used as reference points. To visualize growth changes and changes in the position of individual teeth at important time points of treatment, an overlay of images of scanned models made in the indicated years was used. One of these important treatment points is the condition before and after palatoplasty, when there is both a change in the transverse dimension (closure of the cleft) of the hard palate and a change in the position of the teeth (here the position of tooth 55, who had a mesial shift after surgery). The individual development of the patient was further compared with a control group of 147 healthy boys aged 3-19 years and with a group of 89 patients from the FNKV plastic surgery clinic. In CLP patients at 3 years of age, the 3-3 dimension is practically the same as in the healthy population. However, after plastic surgery of the hard palate at the age of 4-5 years, a significant narrowing occurs due to surgery. By the age of 16, the 3-3 dimension increases due to intensive orthodontic therapy. The final prosthetic treatment increases the transverse dimension by another approx. 2 mm. In order to be able to evaluate surface changes on the palate and jaw, not just selected transverse dimensions, 3D morphometric analysis was used. The individual development of the patient was further compared with a control group of 147 healthy boys aged 3-19 years and with a group of 89 patients from the FNKV plastic surgery clinic. In CLP patients at 3 years of age, the 3-3 dimension is practically the same as in the healthy population. However, after plastic surgery of the hard palate at the age of 4-5 years, a significant narrowing occurs due to surgery. By the age of 16, the 3-3 dimension increases thanks to intensive orthodontic therapy. The final prosthetic treatment increases the transverse dimension by another approx. 2 mm. In order to be able to evaluate surface changes on the palate and jaw, not just selected transverse dimensions, 3D morphometric analysis was used. The individual development of the patient was further compared with a control group of 147 healthy boys aged 3-19 years and with a group of 89 patients from the FNKV plastic surgery clinic. In CLP patients at 3 years of age, the 3-3 dimension is practically the same as in the healthy population. However, after plastic surgery of the hard palate at the age of 4-5 years, a significant narrowing occurs due to surgery. By the age of 16, the 3-3 dimension increases due to intensive orthodontic therapy. The final prosthetic treatment increases the transverse dimension by another approx. 2 mm. In order to be able to evaluate surface changes on the palate and jaw, not just selected transverse dimensions, 3D morphometric analysis was used. The individual development of the patient was further compared with a control group of 147 healthy boys aged 3-19 years and with a group of 89 patients from the FNKV plastic surgery clinic. In CLP patients at 3 years of age, the 3-3 dimension is practically the same as in the healthy population. However, after plastic surgery of the hard palate at the age of 4-5 years, a significant narrowing occurs due to surgery. By the age of 16, the 3-3 dimension increases due to intensive orthodontic therapy. The final prosthetic treatment increases the transverse dimension by another approx. 2 mm. In order to be able to evaluate surface changes on the palate and jaw, not just selected transverse dimensions, 3D morphometric analysis was used. FESA . Log-linear interpolation of areal changes leads to the creation of a color map that graphically and in color shows the growth changes. The areas with the greatest growth potential are shown in pink and those with the least potential in blue. The FESA method was used in patient JV in the age range of 7-17 years. During this period, not only the width of the floor changed, but also its height.

Links

Related Articles

- Typical facial clefts
- Atypical facial clefts
- Oral cavity
- Nasal cavity
- Facial development

External links

- Cleft surgery (cz) (<http://stastny-usmev.cz/operace/>)
- Orthodontics of clefts (cz) (<http://stastny-usmev.cz/ortodoncie-2/>)
- ENT Treatment (cz) (<http://stastny-usmev.cz/orl-2/>)
- Development of the Face and Palate (video) (<https://www.youtube.com/watch?v=iLbqzTIZ6yA>)
- Face Development in the Womb - Inside the Human Body: Creation - BBC One (video) (https://www.youtube.com/watch?v=wFY_KPFS3LA)
- DVOŘÁK, Zdeněk. *Rozštěpové centrum při klinice plastické a estetické chirurgie Fakultní nemocnice U Sv. Anny v Brně* [online]. Přehled chirurgických metod u rozštěpových vad, [cit. 2016-07-07]. <<http://www.rozstep.cz/prehled-chirurgicky-metod-a-nove-trendy-lecby-rozstepu-patra/>>.

References

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1. <https://emedicine.medscape.com/article/995535-overview#a2>
2. <https://emedicine.medscape.com/article/995535-workup>

