Congenital anomalies (development disturbances) of head and neck. Occurrence, etiology, pathogeny, classification, clinical symptoms. Surgical methods of treatment and rehabilitation at children and adolescence.

Pediatric Surgical Dentistry

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050-252-53-15

(Viber)

Plan and organizational structure of lecture

№ з/п	Basic stages of lecture and their maintenance	Type of lecture. Facilities of activation of students. Materials of the methodical providing	Distribution of time
1	Preparatory stage. Determination of actuality of theme, educational aims of lecture and motivation	См. of пп 1 and 2	5%
2	Basic stage of teaching of lecture material according to plan: 1.Статистика. 2.Эмбриогенез persons. 3.Этиология ununions of overhead lip and sky. 4.Классификация несра-щений of overhead lip and sky. 5.Анатомо-функциональные violations at несраще-ниях of overhead lip and sky. 6.Хирургические methods of treatment of ununions of overhead lip. 7.Основные stages of уранопластики on Лимбергу. 8.Наиболее frequent after-operating complications.	Introductory lecture. Thematic lecture. Clinical lecture. Lecture with the use of evidentness. Questions, problem situations, tasks. Facilities of evidentness : patient. Equipment.	85% - 90%
1 2 3	Final stage Resume of lecture, general conclusions. Answers for possible questions. Task for самоподготовки of students	Educational literature. Tasks, questions.	5%

Contents

Definition

Cleft lip (cheiloschisis) and cleft palate (palatoschisis) (colloquially known as harelip), which can also occur together as cleft lip and palate, are variations of a type of clefting congenital deformity caused by abnormal facial development during gestation. A cleft is a fissure or opening—a gap. It is the non-fusion of the body's natural structures that form before birth. 1 in 700 children born do have a cleft lip and/or a palate.

Clefts can also affect other parts of the face, such as the eyes, ears, nose, cheeks and forehead. In 1976, Dr. Paul Tessier described fifteen lines of cleft.

Cleft Lip & Palate

If the cleft does not affect the palate structure of the mouth it is referred to as cleft lip. Cleft lip is formed in the top of the lip as either a small gap or an indentation in the lip (partial or incomplete cleft) or it continues into the nose (complete cleft). Lip cleft can occur as a one sided (unilateral) or two sided (bilateral). It is due to the failure of fusion of the maxillary and medial nasal processes (formation of the primary palate).

Cleft Lip & Palate

A mild form of a cleft lip is a microform cleft.

A microform cleft can appear as small as a little dent in the red part of the lip or look like a scar from the lip up to the nostril. In some cases muscle tissue in the lip underneath the scar is affected and might require reconstructive surgery. It is advised to have newborn infants with a microform cleft checked with a craniofacial team as soon as possible to determine the severity of the cleft.

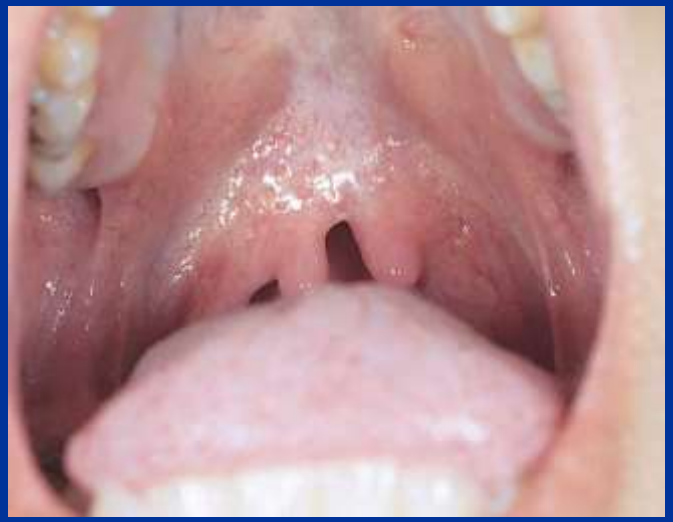
Cleft palate

- Cleft palate is a condition in which the two plates of the skull that form the hard palate (roof of the mouth) are not completely joined. The soft palate is in these cases cleft as well. In most cases, cleft lip is also present.

- Palate cleft can occur as complete (soft and hard palate, possibly including a gap in the jaw) or incomplete (a 'hole' in the roof of the mouth, usually as a cleft soft palate). When cleft palate occurs, the uvula is usually split. It occurs due to the failure of fusion of the lateral palatine processes, the nasal septum, and/or the median palatine processes (formation of the secondary palate).

- The hole in the roof of the mouth caused by a cleft connects the mouth directly to the nasal cavity.

Cleft palate



Submucous cleft palate (SMCP) can also occur, which is an occult cleft of the soft palate with a classic clinical triad of bifid uvula, a furrow along the midline of the soft palate, and a notch in the posterior margin of the hard palate.

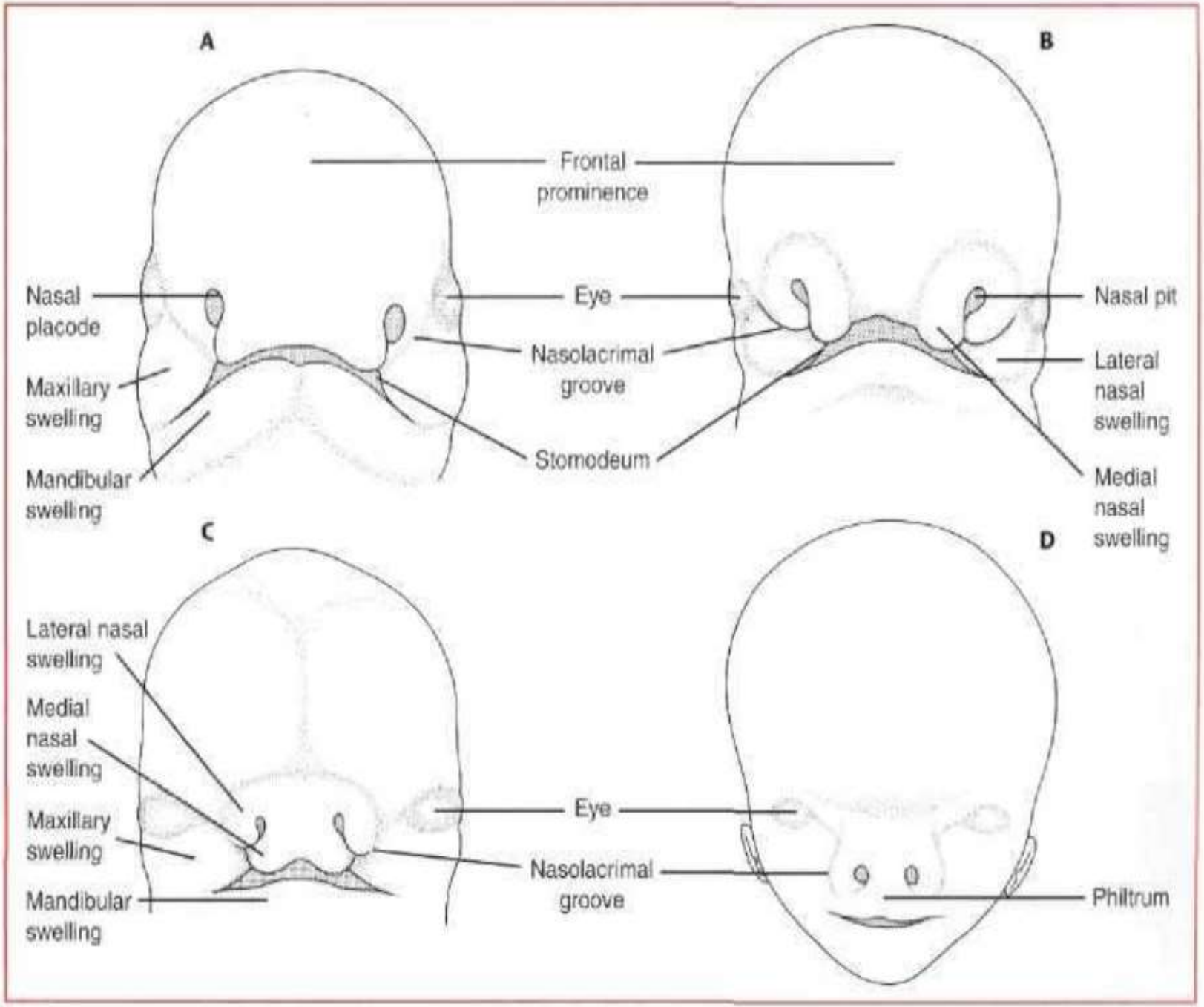
Causes of clefts

The development of the face is coordinated by complex morphogenetic events and rapid proliferative expansion, and is thus highly susceptible to environmental and genetic factors, rationalising the high incidence of facial malformations. During the first six to eight weeks of pregnancy, the shape of the embryo's head is formed. Five primitive tissue lobes grow:

a) one from the top of the head down towards the future upper lip; (Frontonasal Prominence)

b-c) two from the cheeks, which meet the first lobe to form the upper lip; (Maxillar Prominence)

d-e) and just below, two additional lobes grow from each side, which form the chin and lower lip; (Mandibular Prominence)


A
B
Frontal prominence
Nasal placode
Eye
Nasal pit
Nasolacrimal groove
Lateral nasal swelling
Maxillary swelling
Stomodeum
Mandibular swelling
Medial nasal swelling
C
D
Lateral nasal swelling
Medial nasal swelling
Eye
Maxillary swelling
Nasolacrimal groove
Philtrum
Mandibular swelling

Causes of clefts

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

- The upper lip is formed earlier than the palate, from the first three lobes named a to c above. Formation of the palate is the last step in joining the five embryonic facial lobes, and involves the back portions of the lobes b and c. These back portions are called palatal shelves, which grow towards each other until they fuse in the middle. This process is very vulnerable to multiple toxic substances, environmental pollutants, and nutritional imbalance. The biologic mechanisms of mutual recognition of the two cabinets, and the way they are glued together, are quite complex and obscure despite intensive scientific research.

Causes of clefts

Genetic factors contributing to cleft lip and cleft palate formation have been identified for some syndromic cases, but knowledge about genetic factors that contribute to the more common isolated cases of cleft lip/palate is still patchy.

Syndromic cases: The **Van der Woude Syndrome** is caused by a specific variation in the gene IRF6 that increases the occurrence of these deformities threefold. Another syndrome, Siderius X-linked mental retardation, is caused by mutations in the PHF8 gene (OMIM 300263); in addition to cleft lip and/or palate, symptoms include facial dysmorphism and mild mental retardation. In some cases, cleft palate is caused by syndromes which also cause other problems. **Stickler's Syndrome** can cause cleft lip and palate, joint pain, and myopia. **Loeys-Dietz syndrome** can cause cleft palate or bifid uvula, hypertelorism, and aortic aneurysm. Cleft lip/palate may be present in many different chromosome disorders including **Patau Syndrome** (trisomy 13).

Causes of clefts

Causes of clefts

- **Non-syndromic cases**: Many genes associated with syndromic cases of cleft lip/palate (see above) have been identified to contribute to the incidence of isolated cases of cleft lip/palate. This includes in particular sequence variants in the genes IRF6, PVRL1 and MSX1. The understanding of the genetic complexities involved in the morphogenesis of the midface, including molecular and cellular processes, has been greatly aided by research on animal models, including of the genes BMP4, SHH, SHOX2, FGF10 and MSX1.

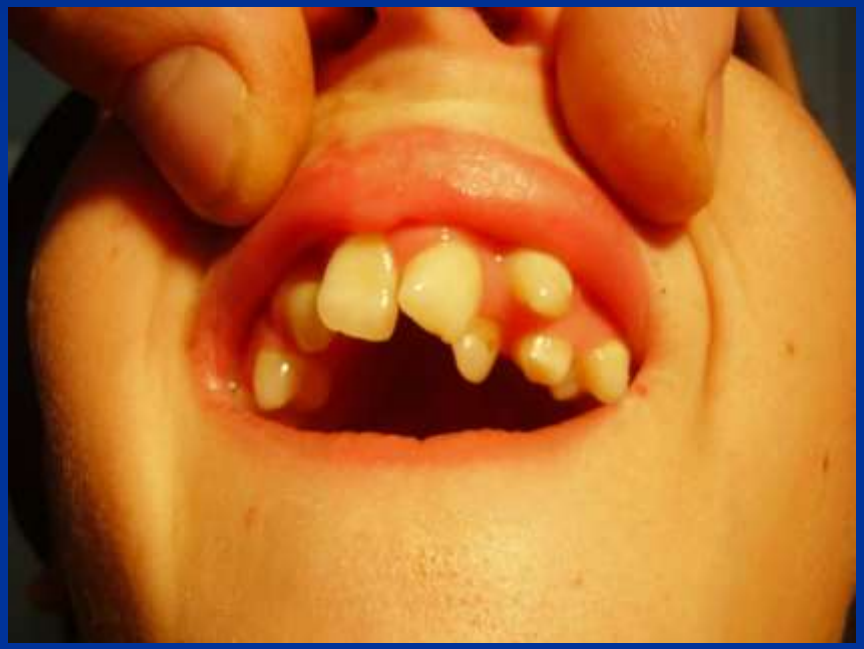
- If a person is born with a cleft, the chances of that person having a child with a cleft, given no other obvious factor, rises to 1 in 14. Many clefts run in families, even though in some cases there does not seem to be an identifiable syndrome present, possibly because of the current incomplete genetic understanding of midfacial development.

Causes of clefts

Environmental influences may also cause, or interact with genetics to produce, orofacial clefting. An example for how environmental factors might be linked to genetics comes from research on mutations in the gene PHF8 that cause cleft lip/palate (see above). It was found that PHF8 encodes for a histone lysine demethylase, and is involved in epigenetic regulation. The catalytic activity of PHF8 depends on molecular oxygen, a fact considered important with respect to reports on increased incidence of cleft lip/palate in mice that have been exposed to hypoxia early during pregnancy. In humans, fetal cleft lip and other congenital abnormalities have also been linked to maternal hypoxia, as caused by e.g. maternal smoking, maternal alcohol abuse or some forms of maternal hypertension treatment. Other environmental factors that have been studied include: seasonal causes (such as pesticide exposure); maternal diet and vitamin intake; retinoids - which are members of the vitamin A family; anticonvulsant drugs; alcohol; cigarette use; nitrate compounds; organic solvents; parental exposure to lead; and illegal drugs (cocaine, crack cocaine, heroin, etc).

Current research continues to investigate the extent to which Folic acid can reduce the incidence of clefting.

Diagnosis



History

- –Prenatal and birth history
- –History of prenatal drug/medication use
- –Family history of cleft lip or palate, or other birth defects
- –Ethnic background

Physical exam

- –Location and extent of the defect
- –Presence or absence of submucosal cleft palate (must be palpated, because it may not be visible beneath the oral mucosa)
- –Dental examination
- –Cardiac examination

Diagnosis

Studies

- –Syndromic cleft lip with or without cleft palate is detected at fetal ultrasound in 38% of cases
- –Nonsyndromic clefting is difficult to see on fetal ultrasound
- –Screening Echo for cardiac anomalies and
- –Chest X-ray for presence of thymus

Screenings

- –Neonatal and periodic hearing screening
- –Calcium level and immunoglobulins for DiGeorge

Dental and speech evaluations

Signs and symptoms

- Orofacial cleft defects are divided into two major groups: cleft lip with or without cleft palate or cleft palate only. Cleft of the lip may involve the alveolus (premaxilla) and may extend through the palate (hard and soft). Congenital clefts of the face occur most commonly in the upper lip. They can range from a simple notch to a complete cleft from the lip edge, through the floor of the nostril and through the alveolus. Cleft lip can occur on either or both sides of the midline but rarely along the midline itself. A cleft lip involving only one side is a unilateral cleft lip, and a cleft on both sides of the midline is a bilateral cleft lip. When a bilateral cleft lip involves clefting of the alveolus on both sides of the premaxilla, the premaxilla is separated from the maxilla into a freely moving segment.

Signs and symptoms

- A cleft of the palate only may be partial or complete, involving only the soft palate or extending from the soft palate completely through the hard palate. A cleft palate can occur alone or with a cleft lip. Isolated cleft palate is more commonly associated with congenital defects other than isolated cleft lip with or without cleft palate. (See Variations of cleft lip and cleft palate.) The constellation of U-shaped cleft palate, mandibular hypoplasia, and glossoptosis is known as Pierre Robin syndrome, or Robin syndrome. Robin syndrome can occur as an isolated defect or one feature of many different syndromes; therefore, a comprehensive genetic evaluation is suggested for infants with Robin syndrome. Because of the mandibular hypoplasia and glossoptosis, careful evaluation and management of the airway are mandatory for infants with Robin syndrome.

Pre-Surgical Devices for Cleft Lip Treatment

In some cases of a severe bi-lateral complete cleft, the premaxillary segment will be protruded far outside the mouth.

Nasoalveolar molding followed by surgery can improve long-term nasal symmetry among patients with complete unilateral cleft lip-cleft palate patients compared to surgery alone, according to a retrospective cohort study. Significant improvements in nasal symmetry were observed in the measurements of the projected length of the nasal ala, position of the superoinferior alar groove, position of the mediolateral nasal dome, and nasal bridge deviation. "The nasal ala projection length demonstrated an average ratio of 93.0 percent in the surgery-alone group and 96.5 percent in the nasoalveolar molding group" this study concluded.

Treatment

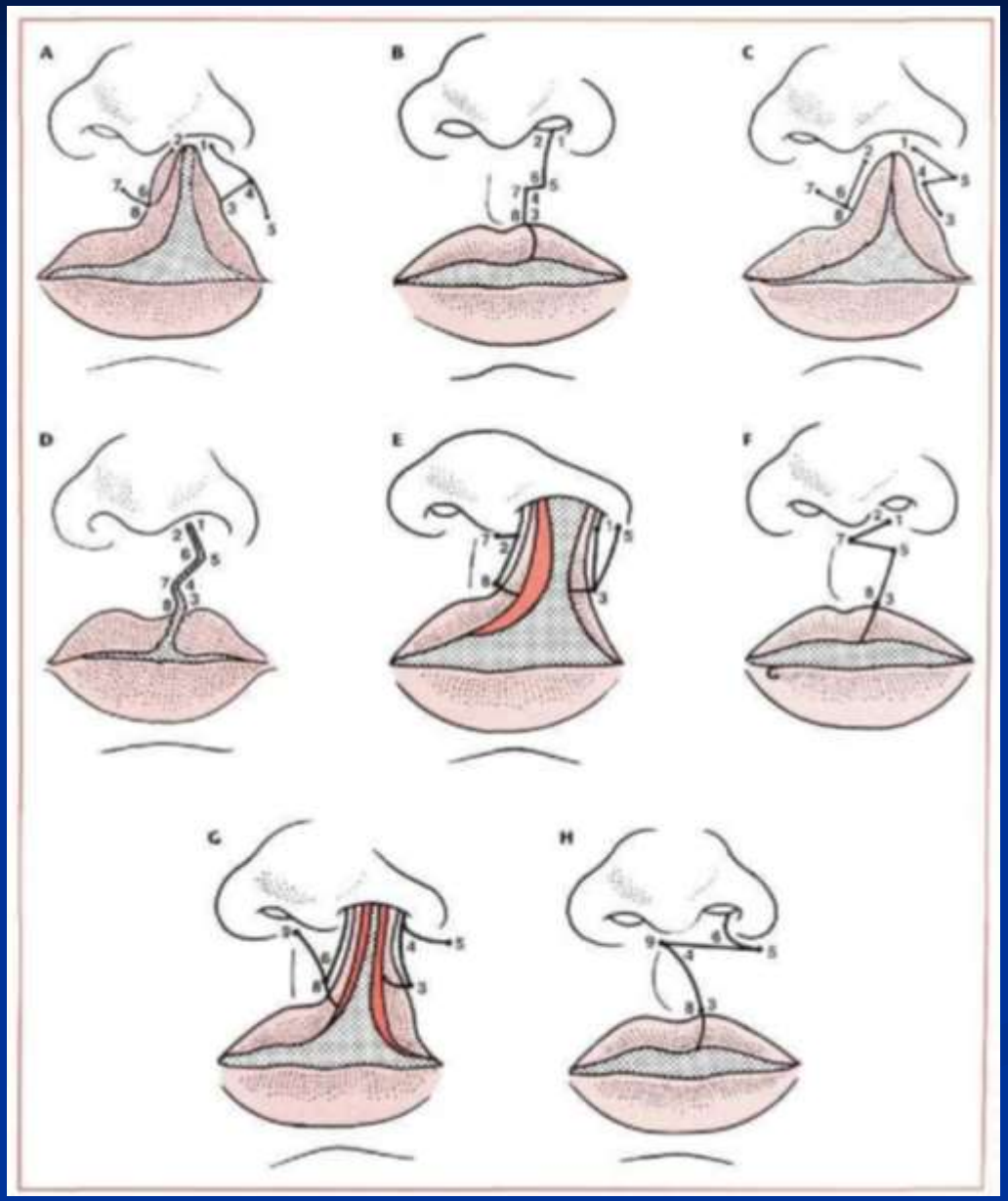
- Treatment consists of surgical correction, but the timing of surgery varies. Some plastic surgeons repair cleft lips within the first few days of life to make feeding the baby easier. However, many surgeons delay lip repairs for 8 to 10 weeks (sometimes as long as 6 to 8 months) to allow the infant to grow and mature, thereby minimizing surgical and anesthesia risks, ruling out associated congenital anomalies, and allowing time for parental bonding. Cleft palate repair is usually completed by the 12th to 18th month. Still other surgeons repair cleft palates in two steps, repairing the soft palate between ages 6 and 18 months and the hard palate as late as age 5 years. In any case, surgery is performed only after the infant is gaining weight and infection-free.

- Surgery must be coupled with speech therapy. Because the palate is essential to speech formation, structural changes, even in a repaired cleft, can permanently affect speech patterns. To compound the problem, children with cleft palates commonly have hearing difficulties because of middle ear damage or infections.

Treatment

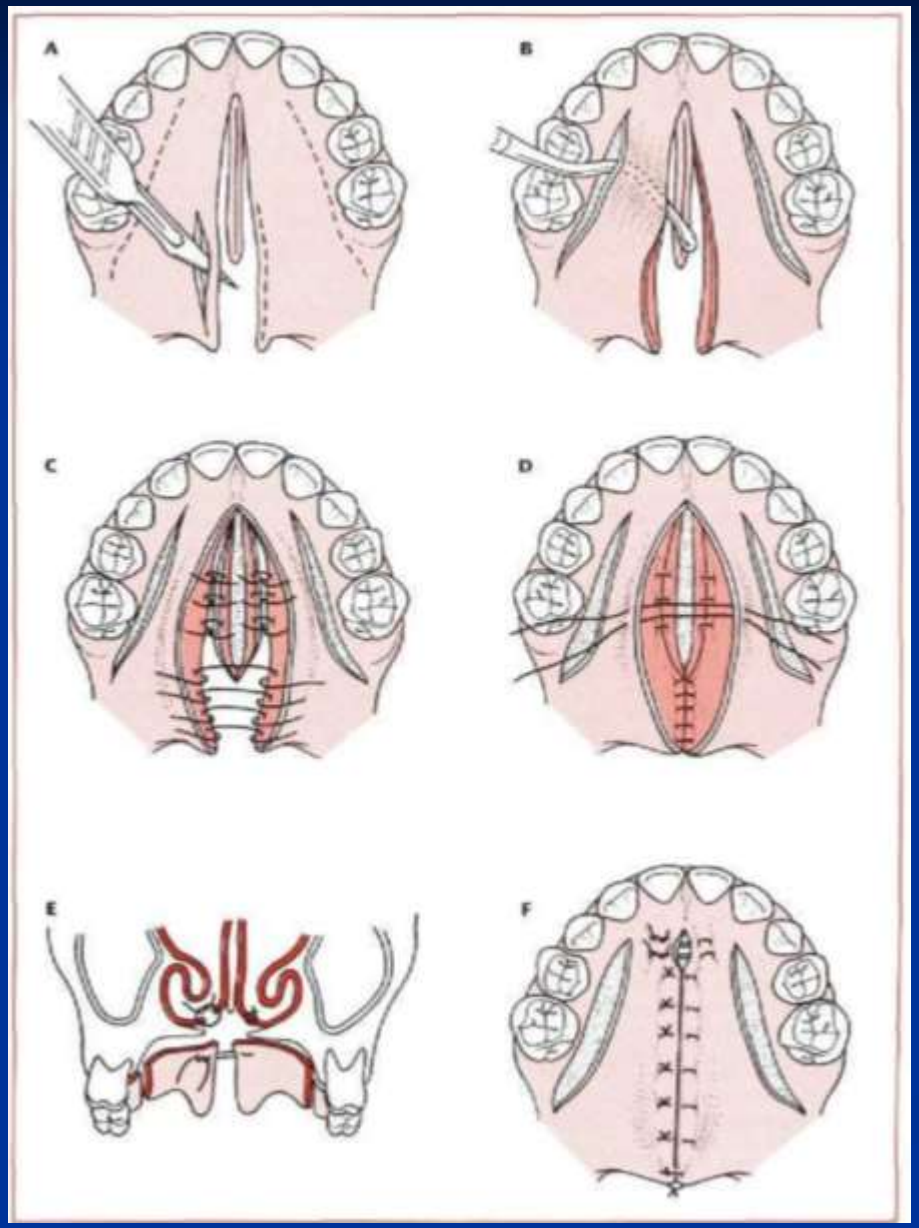
year	0 m	3 m	6 m	9 m	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18
Palatal obturator																						
Repair cleft lip																						
Repair soft palate																						
Repair hard palate																						
Tympanostomy tube																						
Speech therapy/Pharyngoplasty																						
Bone grafting jaw																						
Orthodontics																						
Further cosmetic corrections (Including jawbone surgery)																						

Several cheilorrhaphy techniques



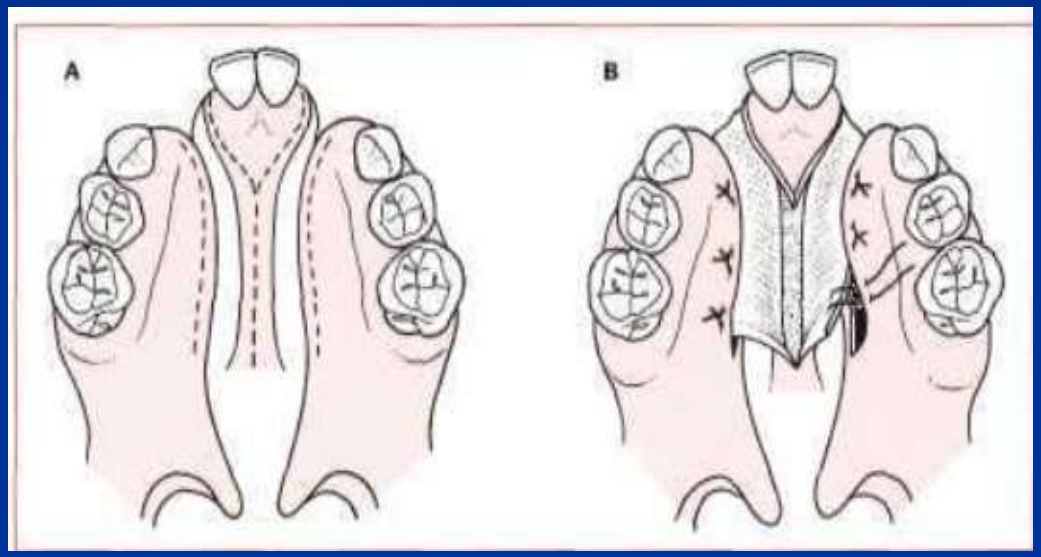
- **A and B, *Ie.* Mesurier** technique tor incomplete unilateral cleft.
- **C and D. Tennison** operation.
- **E and F, Wynr operation**.
- **G and H, Millardoperation** (i.e.., rotation advancement technique).

Palatorrhaphy



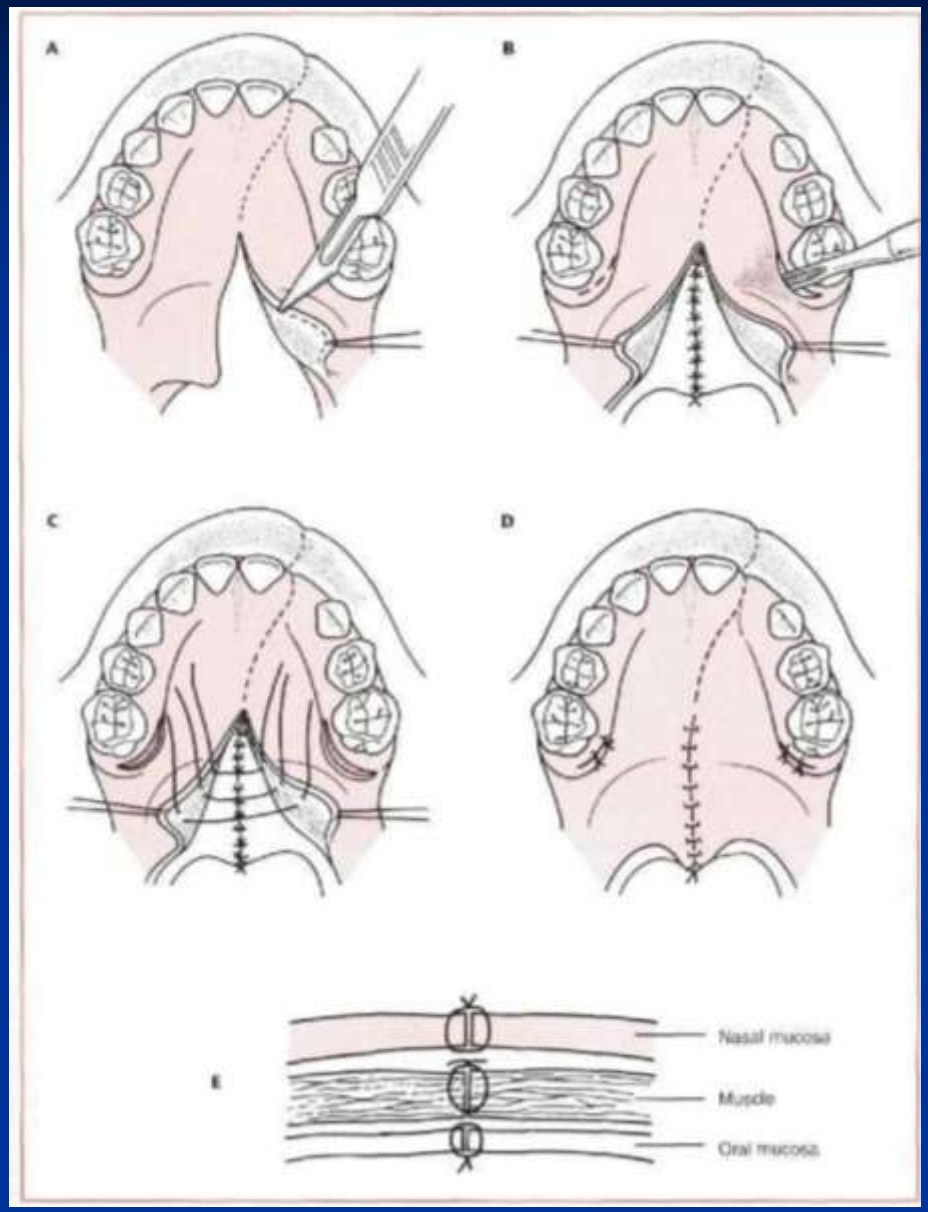
- **Variation of von Langenbeck operation** for concomitant hard and soft palate closure. It yeses three-layer closure for soft palate (i.e., nasal mucosa, muscle, oral mucosa) and two-layer closure for hard palate (i.e., flap from vomer and nasal door to produce nasal closure, palatal flaps for oral clo- sure).
- **A**, Removing mucosa from margin of cleft.
- **B**, Mucoperiosteal flaps on hard palate are developed; note lateral releasing incisions.
- **C**, Sutures placed into nasal mucosa after development of nasal flaps from vomer and nasal floor. Sutures are placed so that knots will be on nasal side.
- **D**, Nasal mucosa has been closed.
- **E**, Frontal section showing repair of nasal mucosa.
- **F**, Closure of oral mucoperiosteum,

Palatorrhaphy



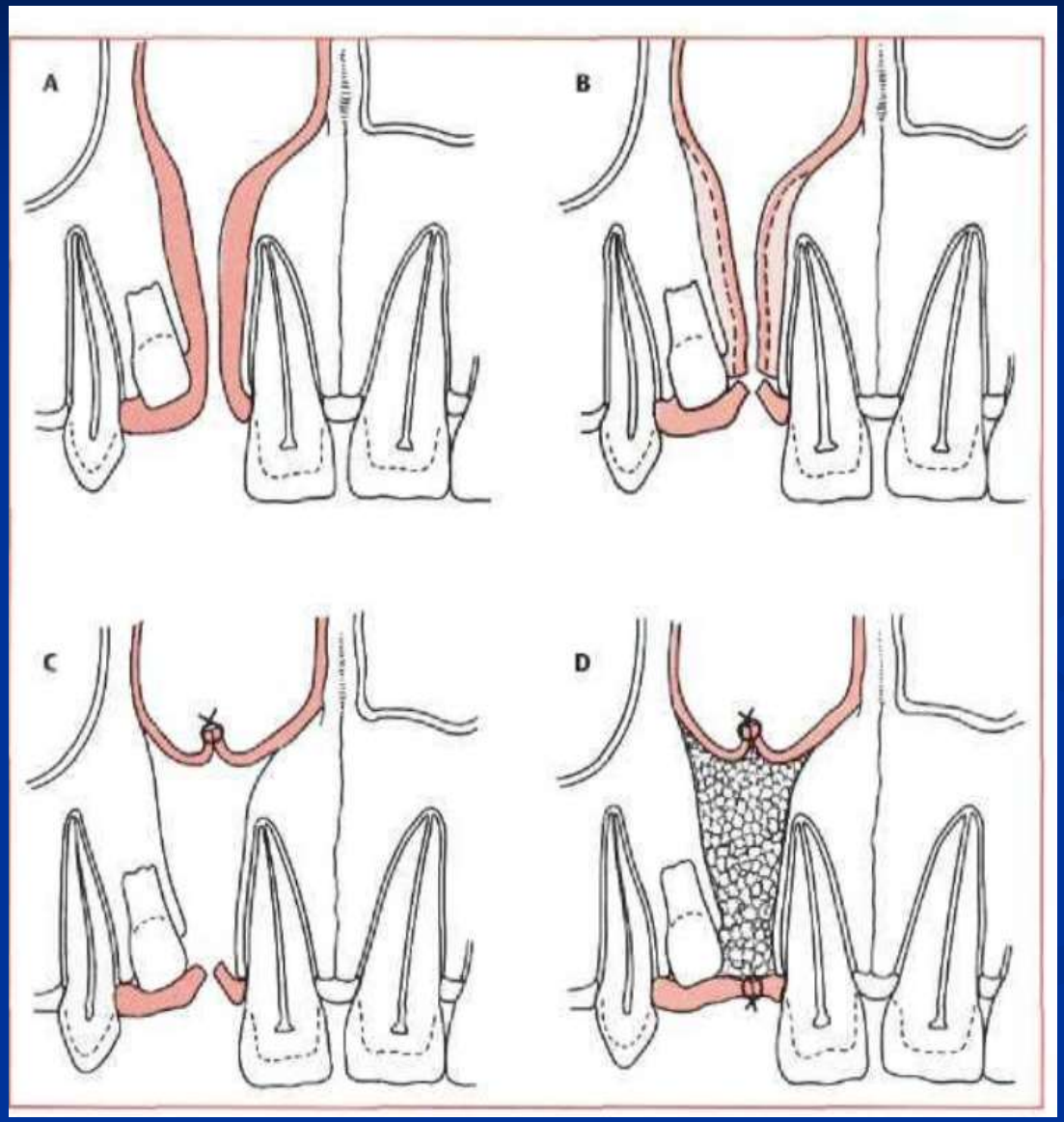
- **Vomer flap technique** for closure of hard palate cleft (bilateral in this case).
- **A**, Incisions through nasal mucosa on underside of nasal septum (i.e., vomer) and mucosa of cleft margins.
- **B,** Mucosa of nasal septum is dissected off nasal septum and inserted under palatal mucosa at margins of cleft. This is one-layer nasal closure only. Connective tissue undersurface of nasal mucosa will epithelialize. This technique, because it does not require extensive elevation of palatal mucoperiosteum, producer less starring with attendant growth restriction.

Palatorrhaphy (triple-layered soft palate closure)



- **A**, Excisfon of mucosa at cleft margin.
- **B**, Dissection of nasal mjcosa from soft palate to facilitate closure. Nasal mucosa is sutured together with knots tied or nasal (i.e., superior) surface. Note small incision made to insert instrument for hamular process fracture. This maneuver refeases tensor veli palatini and facilitates approximation in midline.
- **C**, Muscle *h* dissected from insertion into hard palate, and sutures are placed to approximate muscle in midline.
- **D**, Closure of oral mucosa is accomplished last.
- **E**, Layered closure of soft palate

Alveolar cleft bone grafting



- **A**, Preoperative defect viewed from labial aspect. Fistula extends, into nasal cavity.
- **B**, Incision divides mucosa fistula, which allows development of nasal and oral flaps.
- **C**, Mucosal flap developed from lining of fistula is turned inward, up into nasal cavity, and sutured in watertight manner
- **D**, Bone graft material is packed into cleft, and oral mucosa is closed in watertight manner.

Speech and hearing treatments

- A tympanostomy tube is often inserted into the eardrum to aerate the middle ear. This is often beneficial for the hearing ability of the child.

- Speech problems are usually treated by a speech-language pathologist. In some cases pharyngeal flap surgery or augmentation pharyngoplasty is performed to reduce the escape of nasal airflow in speech sounds requiring oral air pressure, to improve the pronunciation of those sounds, and reduce nasality in those parts of speech that are not normally nasalized. The speech-language pathologist may also be called on to correct incorrect speaking habits that the child developed before the cleft was corrected surgically.

Craniofacial team

- A craniofacial team is routinely used to treat this condition. The majority of hospitals still use craniofacial teams; yet others are making a shift towards dedicated cleft lip and palate programs. While craniofacial teams are widely knowledgeable about all aspects of craniofacial conditions, dedicated cleft lip and palate teams are able to dedicate many of their efforts to being on the cutting edge of new advances in cleft lip and palate care.

Craniofacial team

- Many of the top pediatric hospitals are developing their own CLP clinics in order to provide patients with comprehensive multi-disciplinary care from birth through adolescence. Allowing an entire team to care for a child throughout their cleft lip and palate treatment (which is ongoing) allows for the best outcomes in every aspect of a child's care. While the individual approach can yield significant results, current trends indicate that team based care leads to better outcomes for CLP patients.
- The members of the craniofacial team at a minimum include a plastic or facial plastic surgeon trained in craniofacial surgery, otolaryngologist, geneticist, orthodontist, and social worker.

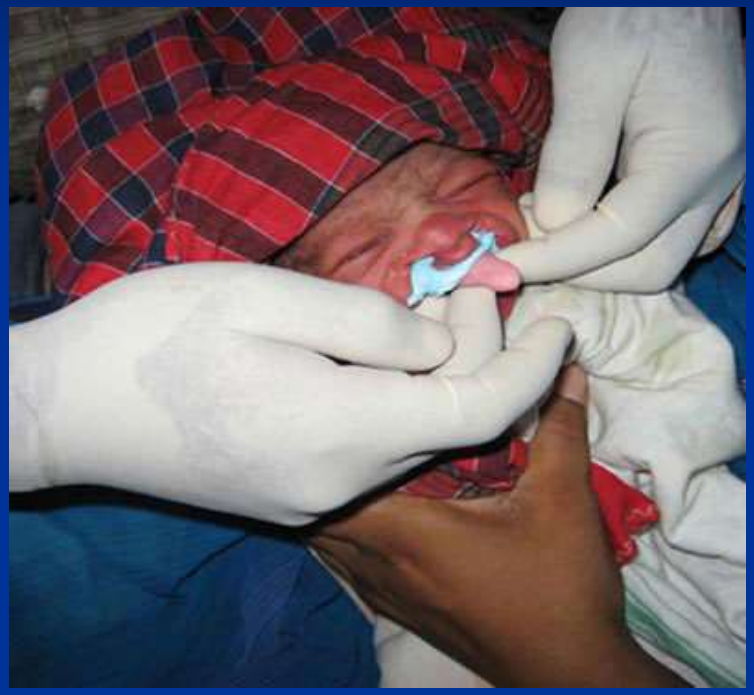
Complications

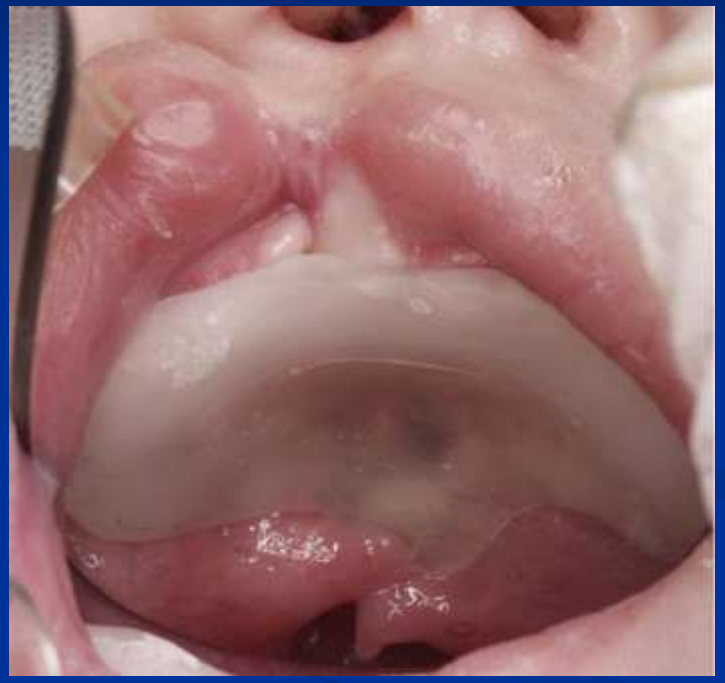
- Cleft may cause problems with feeding, ear disease, speech and socialization.

Complications

- Due to lack of suction, an infant with a cleft may have trouble feeding. An infant with a cleft palate will have greater success feeding in a more upright position. Gravity will help prevent milk from coming through the baby's nose if he/she has cleft palate. Gravity feeding can be accomplished by using specialized equipment, such as the Haberman Feeder, or by using a combination of nipples and bottle inserts like the one shown, is commonly used with other infants. A large hole, crosscut, or slit in the nipple, a protruding nipple and rhythmically squeezing the bottle insert can result in controllable flow to the infant without the stigma caused by specialized equipment.

Infant palatal obturator





- Palatal obturators are typically short-term prosthetics used to close defects of the hard/soft palate that may affect speech production or cause during feeding.

Infant palatal obturator

Complications

- Individuals with cleft also face many middle ear infections which can eventually lead to total hearing loss. The Eustachian tubes and external ear canals may be angled or tortuous, leading to food or other contamination of a part of the body that is normally self cleaning.
- Speech is both receptive and expressive. We hear and understand spoken language (receptive) We learn to manipulate our mouth, tongue, oral cavity, to express ourselves (expressive).

Complications

- Hearing is related to learning to speak. Babies with palatal clefts may have compromised hearing and therefore, if the baby cannot hear, it cannot try to mimic the sounds of speech. Thus, even before expressive language acquisition, the baby with the cleft palate is at risk for receptive language acquisition. Because the lips and palate are both used in pronunciation, individuals with cleft usually need the aid of a speech therapist.

- Bonding with the infant, socializing with family and community may be interrupted by the unexpected appearance, unusual speech and the surgical interventions necessary. Support for the parents as well as for the child can be pivotal.

Psychosocial issues

- Having a cleft palate/lip does not inevitably lead to a psychosocial problem. Most children who have their clefts repaired early enough are able to have a happy youth and a healthy social life. However, it is important to remember that adolescents with cleft palate/lip are at an elevated risk for developing psychosocial problems especially those relating to self concept, peer relationships, and appearance. It is important for parents to be aware of the psychosocial challenges their adolescents may face and to know where to find professional help if problems arise.

Psychosocial issues

- A cleft palate/lip may impact an individual's self-esteem, social skills, and behavior. There is a large amount of research dedicated to the psychosocial development of individuals with cleft palate. Self-concept may be adversely affected by the presence of a cleft lip and or cleft palate. Research has shown that during the early preschool years (ages 3–5), children with cleft lip and or cleft palate tend to have a self-concept that is similar to their peers without a cleft. However, as they grow older and their social interactions with other children increase, children with clefts tend to report more dissatisfaction with peer relationships and higher levels of social anxiety. Experts conclude that this is probably due to the associated stigma of visible deformities and speech abnormalities, if present. Children who are judged as attractive tend to be perceived as more intelligent, exhibit more positive social behaviors, and are treated more positively than children with cleft lip and or cleft palate. Children with clefts tend to report feelings of anger, sadness, fear, and alienation from their peers. Yet these children were similar to their peers in regard to "how well they liked themselves."