

Cleft lip and palate

Incidence and prevalence

-It's the most common craniofacial anomaly affecting still and live birth. affecting 1 in 600-1000 live births (incidence ~ 0.7%), which is considered very common.

- Clefts of the lip are more common in males, clefts of the palate are more common in females, and cleft lip and palate is equal between males and females, with some studies saying there is a male predominance.

Overall, Males>Females.

-Unilateral cleft lip is more common on the left side

-Incidence shows racial differences: Far east > Caucasian >African

#Incidence in Jordan:

- A retrospective study was made by Dr lyad and result were similar to that of the Caucasian (~ 1 in 770 live births)

- 30% cleft lip, 22% cleft palate and 48% cleft lip and palate.

- 55% Males and 45% Females (difference in gender is not statistically significant)

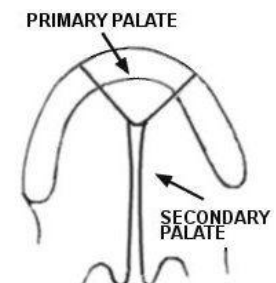
-18% of cases were associated with major anomalies or syndromes (that might be because there is a higher chance of recording major syndromes in a patients file than just recording a minor CLP)

*problems in retrospective studies: deficient data in patients file, leading to a chance of bias

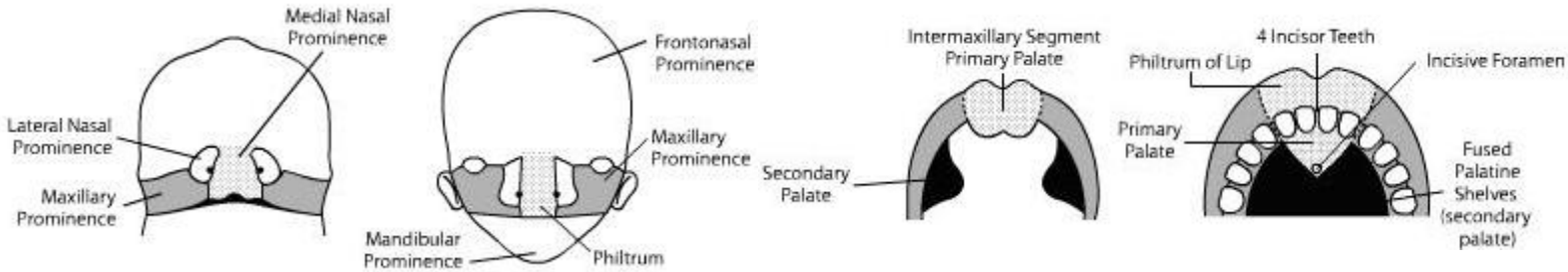
Development

-stage of primary palate development is between 4-6 weeks
(*problems during this time leads to cleft lip and/or anterior palate*)

-stage of secondary palate development is between 6-8 weeks
(*failure of fusion, or any problem in elevation of the palatal shelves during development leads to cleft palate*)

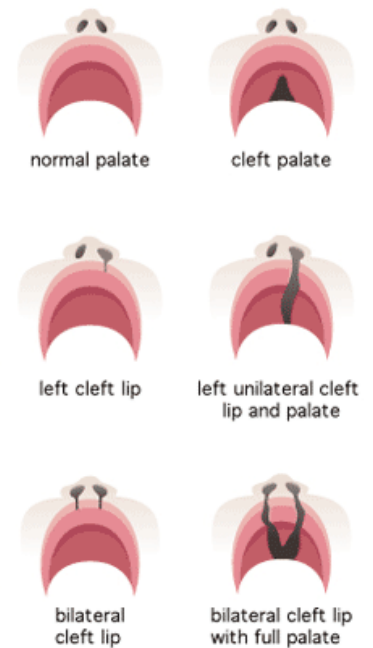


- upper lip forms by the fusion of medial nasal process, lateral nasal process and the maxillary process on both sides.
(failure of fusion leads to cleft lip)



- clinical presentation of CLP is wide, ranging from a unilateral cleft lip, to bilateral cleft lip and palate. Also, ranging in severity, a small part of the lip may be affected or the most posterior part of the soft palate, not necessarily the whole palate.

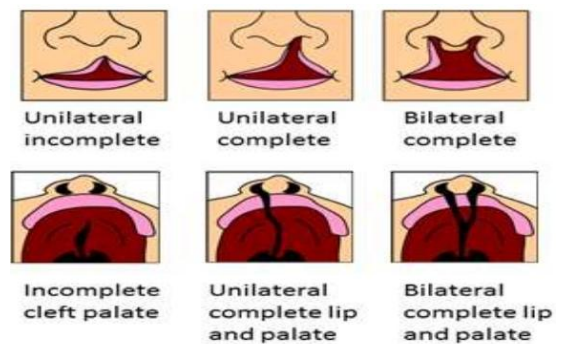
-submucous cleft is a variant of CLP not involving soft tissue, only the bone, which makes it difficult to diagnose.



Classification

several classifications are present:

- Syndromic/non-syndromic (associated with syndromes or not)
- primary palate cleft/ secondary palate cleft/ both
- complete/ incomplete (involving the whole structure or not)
- unilateral/ bilateral



*simplest way to record the cleft lip and palate in a patients file is using a schematic diagram to draw or mark the clefted parts on it.

Etiology

most recent reviews shows that it's multi-factorial were genetics and environment interact.

genetics:

it has a strong family history but it's not a simple mendelian inheritance pattern; mode of inheritance is not completely understood.

mutations in chromosome number 6 are thought to cause CLP.

#environmental factors:

- nutritional deficiencies (most common)
mainly *folic acid* and *Iron*
(may explain its high incidence in far east and africa)

*It was found that improving the mothers nutrition during pregnancy and supplying her with supplementation reduced the occurrence of neural crest defects and CLP; specially **folic acid** supplements .*

a higher conc of folic acid is given for the pregnant mother in case there was a family history risk. (0.4mg if no family history and 4mg if there is family history)

- radiation
- drugs
- hypoxia
- viruses (EBV)
- vitamin deficiencies

Consequences of CLP

-Patients with untreated CLP usually have:

1-feeding problems

It's not usually a big problem, as people with unrepaired CLP, managed to live and adapt to it. Specialized craniofacial people may train the mother how to manage feeding the child at early age.

There are also special feeding bottles (haberman feeding bottle) that's got its opening from the bottom rather than the middle to prevent milk from going up to the nasal cavity



2-speech problem

the most difficult to fix, if treatment of CLP was late.

It will be affected if the defect was not treated at the right time; mainly nasal speech problems.

The right time to correct the defect is before 2 years old (the time which pronunciation and normal speech sounds are developing), if child reached 4 years old and CLP was not treated, it will be very difficult to treat the speech problems. Therefore intervention must be planned at the right time.

3-hearing problems

mostly due to ear infections and middle ear fluid accumulation. This will further complicate speech problems.

4-esthetic problem (most important complication)

Its the most concerning issue for the patient. Plastic surgeries now would achieve excellent results, but it's difficult to restore natural appearance by 100%.

5- psychological consequences

They are not severe, there's no depression or psychological diseases.

Most commonly they just have low self esteem. They might also have problem in interaction with people, mainly due to speech problems.

6-Dental problems: (least imp to worry about)

- *Congenital problems:*
 - >>hypodontia (most common, specially on the side of the cleft)
lateral incisors are the one most commonly missing, then the centrals
 - >>microdontia
 - >>impaction
mainly the permanent first molar,
also the canines get impacted due to absence of bone
 - >>hyperdontia (may also occur ex: mesiodens)
- *Iatrogenic* problems when fixing the CLP, like removing or injuring or stitching over the tooth bud may also lead to dental problems
- *occlusion*
there is a defect in soft tissue and hard tissue of the maxilla in case of cleft

palate leading to collapse in the size of the maxillary arch

The main cause of maxillary deficiency is early surgery to fix the CLP. It would lead to healing by secondary intention and scar tissue formation, also binding of the maxilla to the pterygoid processes, this will prevent the downward forward growth of the maxilla.

patients will have: >Anterior and posterior cross-bites (very common)

> class 3 incisal relationship (due to maxillary deficiency)

> also teeth will tend to tilt towards the area of the cleft (rotations)

Facial growth

The proof that maxillary deficiency in patients with CLP is due to early surgery is that people with *unrepaired* CLP had normal class 1 incisal relationship.

A research was conducted by Mars M, in the Far East (Thailand and India) in 1990, where people didn't repair the CLP for it is considered a fortunate event to have a cleft and a sign of power. They showed normal incisal relationship, compared with people with early repaired CLP, proving that surgery is what hinders the normal development of the maxilla. They also have succeeded to adapt to feeding in the presence of cleft.

Diagnostic considerations

>>Prenatal diagnosis of CLP

in the past, parents would be shocked after delivery of the child's appearance having Cleft lip and palate.

Now prenatal diagnosis before labor can be done using:

- *high resolution ultrasonography* (ultra-sound) at 3-5months of pregnancy
detection rate is more than 22%-33%
cleft lips are easier to detect on ultra-sound than cleft palate
- *3D ultrasonography* (has greater sensitivity and higher detection rate)

**advantage of prenatal Dx: -to psychologically prepare the parents (mainly)

-to prepare the hospital

-to prepare for fetal surgery if to be done

fetal surgery: there are new studies on prenatal repair of CLP (fetal surgery), for the child to be born without clefts (normal appearance).

Studies were done on animals, with limited human trials. Thoughts that it will be used in the near future as a treatment modality after prenatal diagnosis of CLP.

**disadvantages of prenatal Dx: - emotional disturbance
-maternal anxiety

Management

These patients must be managed by a trained *cleft lip and palate team (craniofacial team)*. For example, surgeons, orthodontists and speech therapists who are trained to deal with CLP.

They require many stages of intervention from birth to adulthood (~15 years old).

#History of management:

- ✓ in the early 19th century, they used to use prosthodontic devices (obturators) to cover the palatal clefts
- ✓ in late 19th century, surgical correction of the cleft became the treatment of choice
- ✓ in 20th century (1940's), cephalometry was introduced and was used in evaluation of growth in CLP patients and orthodontists became involved in cleft repairing.
- ✓ in 1970's, alveolar bone grafting was introduced to restore the integrity of the arch and provide bone for teeth to erupt
- ✓ in 1980's, osteotomies of the maxilla were introduced to restore size and shape
- ✓ in 1990's, distraction osteogenesis to increase size of the maxilla was used to restore defect with *minimal scar tissue* formation. It also has an advantage that it's done early (*during growth*) between 8-10 years old, unlike surgical osteotomies - orthognathic surgery which needs to be done after 18 years old (when growth is completed). Yet, results were not as good as they were expected to be, therefore it didn't replace conventional surgery which gave more predictable and solid outcomes.

Many studies were done to help us know the best way to manage patients with CLP including: **1) Euro Cleft study 2) Dutch Study 3) Americleft study**

Euro Cleft study(1999):

was made in Europe by orthodontists, it studied the outcome of different management approaches that were used in 6 centers in different European countries (Norway, Netherlands, Germany, UK, France)

the outcomes that they looked at were the final appearance, maxillary growth, dental features and also speech

Results showed that there was no consensus on one treatment protocol, each centre used a different approach and Results were variable.

Mostly speech and appearance outcome were poor compared to the ideal outcome.

Norway team was the best team and their management protocol showed the best results. Therefore their protocol was adopted (**Oslo protocol**)

Oslo Team protocol

Cleft team has a *team coordinator* which is the maxillofacial/plastic surgeon OR the orthodontist. Other team members include the speech therapist, the psychologist, the social worker, trained nurse, pediatrician, pediatric dentist, ENT.. etc

#Management sequence:

1- at birth > records are taken, social counseling and trained nurse to treat the mother how to feed her child

2- from 2 weeks - 3 months > closure of cleft lip (lip repair)

its usually not done at birth due to high risk of bleeding, since liver function is not complete. Yet, we can give medications to decrease bleeding and do the surgery at birth. Its recommended to be done as early as possible to assure the parent, mostly before 6 months.

3- 18-24 months > closure of palatal cleft

as late as possible to decrease impact on maxillary growth but before 2 yrs old for speech not to be affected

4- 7-8 years > slight orthodontic alignment of rotated incisors and incisal crossbites can be done before the placement of bone grafts, but mainly comprehensive orthodontic treatment is done after grafting.

5- 8-11 years old > Secondary operations like *Alveolar bone graft* are done, if needed. They must be done before canine eruption to provide them with bone to erupt. So, ABG help retain the integrity of dental arch, they are important for teeth eruption and they help supporting the ala-nasal process

6- Early teenage years >> comprehensive orthodontic treatment
must asses if orthodontic treatment is enough to correct the crossbites- if mild- or future surgery is needed-if severe.

7- After 18 years old >> orthognathic surgery, if needed, to regain correct arches relation. (maxillary osteotomy, or setting back the mandible)

Record taking

mainly done by orthodontist:

- *at birth- before lip surgery
- *3 years- after palatal surgery
- *5-6 years -before mixed dentition
- *10 years-at time of ABG and before ortho
- *18 years-orthoganthic surgery
(satisfaction of patient is recorded after surgery, when all treatment is done)

Timing of the minimum records						
timing	models	ceph	photos	speech	audio	Satis- faction
1st surgery	X		X			
3 years				X	X	
5-6 years	X		X	X	X	
10 years	X	X	X	X	X	
18 years	X	X	X	X		X

Advances in technology:

- CAD/CAM produced devices and splints that help in restoring the maxillary arch in surgery
- fetal surgery before delivery
- tissue engineering and stem cells to promote normal healing
 (example: BMPs and other growth factors)

P.S: the reference for this lecture is the sheet, as the slides were prepared for post-graduate students. GOOD LUCK ^_^

Done by Jumana Qussad