CLEFT LIP AND PALATE

SAMIR JABAITI, MD, FRCS.
PLASTIC SURGEON
SCHOOL OF MEDICINE
UNIVERSITY OF JORDAN

EMBRYOLOGY

Frontonasal process which is proliferation of mesenchyme from the ventral surface of the developing brain, forms the, **nose, the central part of the upper lip (the philtrum), and the central part of the alveolar process (the part which carries the central and lateral incisors)**. The first pharyngeal arch gives rise to the **maxillary and the mandibular processes**, from the former, develop the lateral parts of the upper lip, the lateral parts of the alveolus, it also gives rise to the palatine shelves which fuse in the midline to form the palate. The mandibular processes from each side fuse to form the lower lip and the mandible.
So the cleft lip and cleft alveolus result from non fusion of the frontonasal process with either one or both maxillary processes resulting in left, right, or bilateral cleft. Non fusion of the palatine shelves, result in cleft palate.

INCIDENCE

The incidence of Cleft Lip or Cleft Palate is 1:750. It constitutes 2/3 of all craniofacial anomalies; the incidence of cleft lip is two times that of cleft palate.
In cleft lip, 60% of the cases affect the left side, 30% the right side, and 10% bilateral.
Cleft lip is more common in males, while cleft palate is more common in females.
Isolated cleft palate is associated with other syndromes in 30% of cases.

ETIOLOGY

Not known. While hereditary plays important role, other factors include:

1. Vitamin deficiency in pregnancy (folic acid).
2. Drugs as steroids.
3. Gestational viral infections or irradiation.
4. Loss of amniotic fluids.

CLEFT LIP

Cleft lip, which is usually associated with nasal deformity, is purely aesthetic problem, although it can be corrected at birth or later at any age. Most surgeons prefer to repair it at 3 months of age.

CLEFT ALVEOLUS

Cleft alveolus may lead to abnormal teething especially of the lateral incisors and canines. Orthodontic treatment may be needed to correct the alignment of the alveolar arch, and those children need alveolar bone graft at the age of 8-9 years to allow the eruption of the permanent canine.

CLEFT PALATE

Cleft palate is not an aesthetic problem as cleft lip but it is associated with many functional problems: **feeding**, **speech, regurgitation of food from nose and may lead to hearing loss due to recurrent ear infections**. Normally Eustachian tube is patent to equalize pressure. Patients with cleft palate have Eustachian tube dysfunction due to abnormal insertion of muscles, so the tube is not patent (obstructed) so fluids will accumulate in the tube leading to **secretory otitis media** which is evident by accumulation of fluids behind the ear drum, this is treated by the ENT specialist by drugs as anti-histamines, or by drainage of the fluids surgically by puncturing the ear drum and putting tubes (Gromet tubes) for continuous drainage. If secretory otitis media is not properly treated it would be complicated by bacterial acute otitis media (recurrent ear infections) that may lead to with and hearing loss. So remember that hearing loss in cleft palate patients is not congenital by acquired due to repeated ear infections.

Function of the soft palate:

The soft palate (velum) is formed of muscles that elevate the soft palate and push it backward to meet the posterior pharyngeal wall so as to close and separate the nasopharynx from the oropharynx. So the **velopharyngeal competence** is defined as the ability of the soft palate (velum) and the pharynx to act as a valve between the mouth and the nose. This valve should be open in **breathing** to allow air to get into the airways. Also it should be closed in **swallowing** to prevent nasal regurgitation of food.

Normal speech also requires closure of the port between the mouth and the nose so as to create a positive pressure inside the oral cavity to pronounce most of the consonants. **In suckling** the **port between the mouth and the nose should be closed to create negative pressure for suckling.**

Failure of this valve mechanism is called **VELOPHARYNGEAL INCOMPETENCE.** Among many causes, **cleft palate is the most common cause of this incompetence**, which is attributed to 3 abnormalities in the patient with cleft palate:

1. The mechanical defect of the cleft.
2. Hypoplasia of the palate.
3. Abnormal insertion of the palatal muscles

Surgical correction of cleft palate, aims at closure of the cleft palate to restore the velopharyngeal competence.

FAMILY COUNSLING

The parents of the cleft baby, should be counseled, this means reassuring them, relieving their anxiety, discussing with them the problems associated with cleft palate, especially the feeding and its management. Also they should be informed about the other functional deficits: nasal regurgitation, speech abnormalities (nasal speech), and the importance of follow up by the ENT specialist to manage the ear infections. The family should be introduced to the Cleft Palate Team which consists of: Plastic surgeon, Pediatrician, ENT surgeon, Dentist, Orthodontic surgeon, Speech therapist, Cleft palate nurse, and Social worker.

1. FEEDING

For normal feeding the baby should suckle the milk and then swallow it. It is so important to educate the mother that babies with cleft palate have **defective sucking**, simply because they can not create negative pressure inside their oral cavity to suckle (the mouth is communicating with the atmosphere through the nose). This makes breast feeding difficult. **Although suckling is defective, swallowing is normal.**
With these fruitless efforts the baby will get exhausted and fall asleep with insufficient feeding that leads to weight loss. Although there are special nipples available in the market, that are designed to close the cleft while feeding to help suckling. This special nipple is not mandatory as a simple solution of the problem of ineffective suckling is simply to passively introduce the milk to the mouth by **widening the opening of the bottle nipple,** so as the milk is getting to the mouth passively.
The following roles should be applied:

1. The mother is the best nurse; she is the person who should be involved from the very beginning in the feeding and care of her baby.
2. Nasogastric feeding should not be used for permanent feeding.
3. Babies should be nursed in semi-sitting position, and should be burped well, to get rid of the swallowed air.
4. It should be realized that feeding of cleft babies – at least in the beginning- takes more time than normal babies, the mother must be patient!!!
5. Although breast feeding is difficult some babies can be breast fed as the mother would help this by pressing her breast. So breast feeding although difficult is not contra-indicated.

2. SPEECH.

Normal speech requires that air coming from the lungs, passing through the vocal cords, is collected in the oral cavity to create positive pressure, before passing through the lips to pronounce most consonants.
Patients with cleft palate, with **velopharyngeal incompetence** are unable to create this positive intra-oral pressure as air leaks through the nose, leading to nasal escape, or abnormal nasal speech. Again surgical correction of the cleft helps to restore normal speech, in addition they need the help of speech therapist to achieve good speech.

3. RECURRENT OTITIS MEDIA AND HEARING LOSS

Normally the Eustachian tubes should be patent and aerated to balance the pressure on the two sides of the tympanic membrane.
In cleft palate patients the tubes are not patent due to abnormal insertion of pharyngeal muscles, so fluid accumulates behind the eardrums leading to recurrent otitis media, and if not managed may result in hearing loss.
The ENT surgeon, is an important member of the Cleft Palate Team deals with these problems by treating secretory otitis media (by drugs or Gromet tubes), and treating acute otitis media by suitable antibiotics.

4. NASAL REGURGITATION;

Children with cleft palate have embarrassing nasal regurgitation as swallowed food will escape from nose.

TIMING OF SURGICAL REPAIR OF CLEFT PALATE

Speech therapists believe that, the earlier the cleft palate repair is, the better the outcome of speech would be, so they encourage early repair, but the facial surgeons think that early surgical repair would interfere with the facial bony growth leading to retardation of maxillary growth (dish face). So the compromise between these two opinions is to operate at 1 year of age.