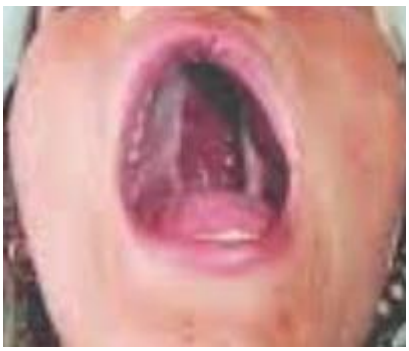
Cleft lip & Palate

Prof. Dr Balasubramanian Thiagarajan (drtbalu)

Children with cleft lip and palate have multiple ENT problems

Cleft lip and cleft palate are the commonest form of orofacial clefting



ENT Problems

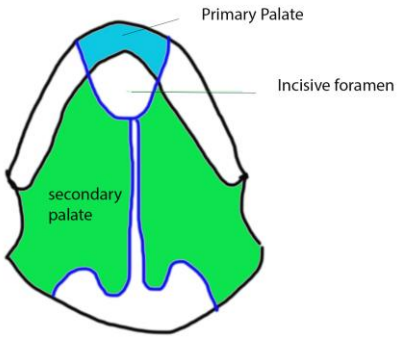
1. Airway problems
2. Hearing problems
3. Speech problems
4. Nasal problems

Cleft types & Classification

Cleft lip is defined as cleft of the structures anterior to the incisive foramen. This is also known as primary palate

Orofacial clefts are classified by laterality and extent of the defect

1950 Kernahan & stark proposed embryological classification of clefting in relation to the incisive foramen

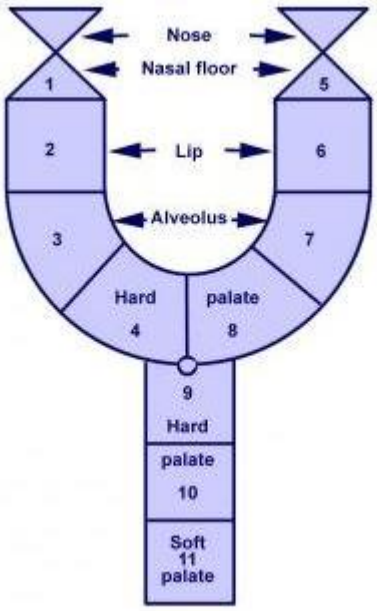


Veau's classification 1931 commonly utilized. It is incomplete since it makes no provision for isolated cleft lip

Veau's classification 1931

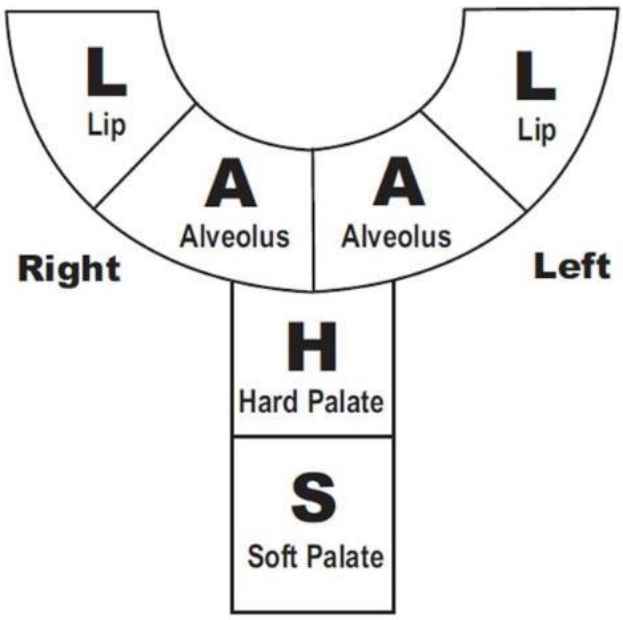
Group I – Defects of soft palate only
Group II – Defects involving the hard palate, and soft palate extending not further than the incisive foramen
Group III – Defects from the soft palate to the alveolus (usually involving the lip)
Group IV – Complete bilateral clefts

Modern Classification



Modified Kernahan classification 1971

This classification is based on intraoral view of the palate. A Y diagram is used for this purpose

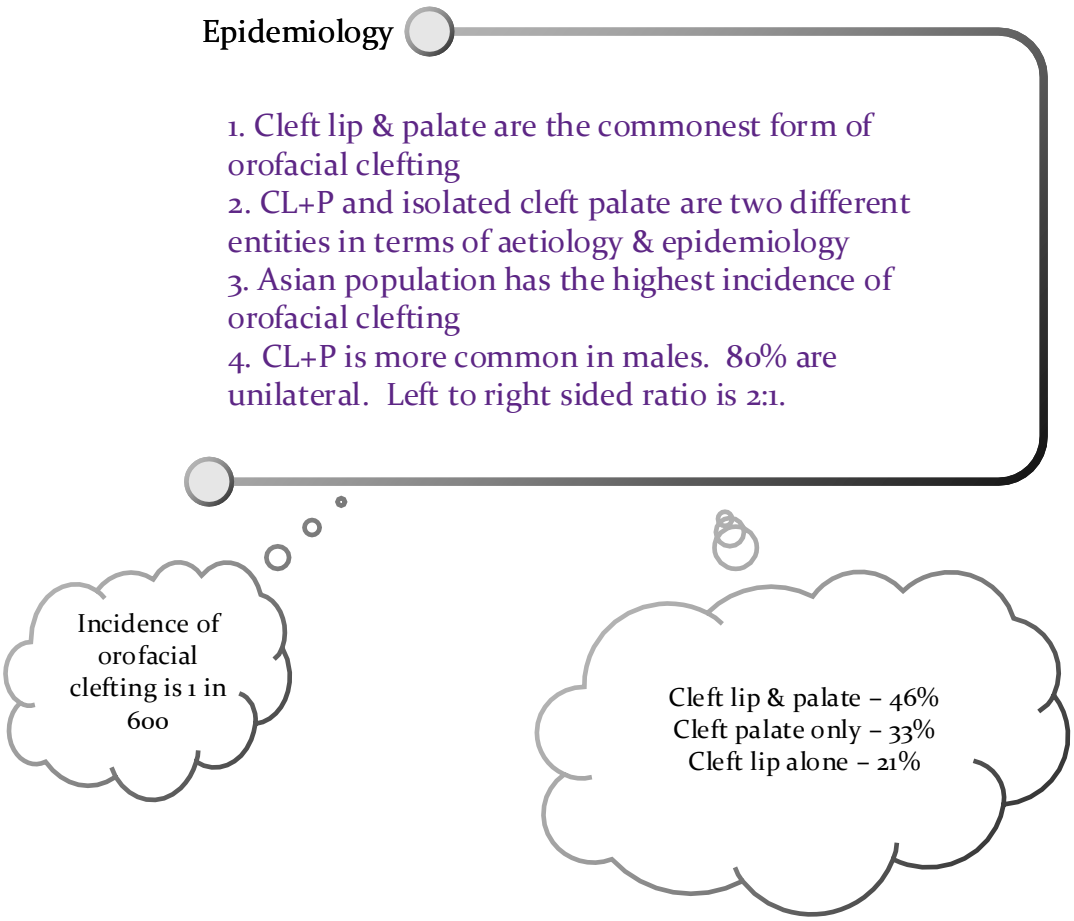


LAHSHAL system is used in UK.

Involves recording the cleft status starting on the right side and working from the lip back to the soft palate

The areas are subdivided into lip, alveolus, hard palate and soft palate

Epidemiology



Aetiology & Genetics

Aetiology

1. Orofacial clefts may be syndromic and non-syndromic and occur as an isolated anomaly
2. Cleft palate alone is more commonly associated with a syndrome than CL+P
3. Nearly 400 syndromes have been associated with CP alone
4. Most frequent interstitial deletion in humans is deletion of (22)(q11,2)
5. Approximately half of the cases of 22q deletion will have a palatal abnormality, the most common being submucous cleft palate / cleft palate. These children can also have non cleft velopharyngeal dysfunction due to deep post nasal space secondary to a wide skull base angle

If a primary relative has a facial cleft the chances of the child having a cleft in the absence of a defined syndrome is 3.5%

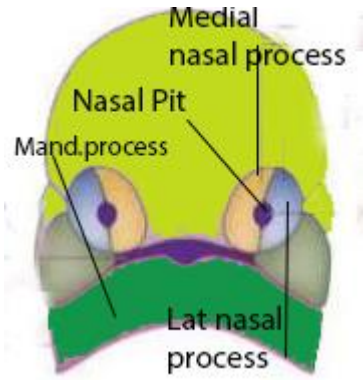
Risk factors associated with CL/P

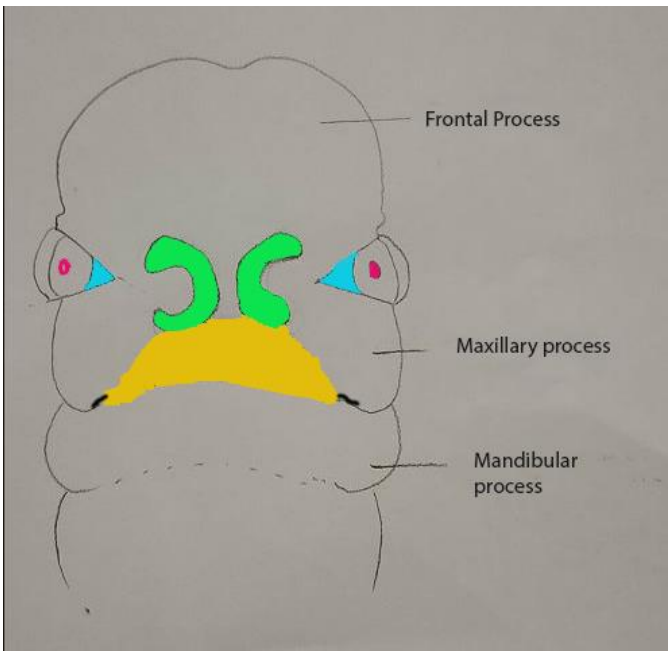
1. Maternal age
2. Maternal smoking
3. Maternal obesity
4. Prepregnancy diabetes
5. First trimestor heavy alcohol consumption
6. Medications (anticonvulsants, folate antogonists, retinoic acid)
7. White non-Hispanic race
8. Sex

Any pt with a palatal defect and any other manifestation of 22q11 (cardiac malformation, neurodevelopment delay, immunodeficiency) should be screened

For parents who already have a child with an orofacial cleft, the risk of subsequent child having a cleft is around 5%

Embryology





Embryology

1. 5 Main prominences are responsible for the development of cranium and face
2. These prominences include: frontonasal prominence, the paired right and left maxillary prominences and the right and left mandibular prominences
3. Incomplete / aberrant timing of fusion of the frontonasal and maxillary prominences results in cleft lip and / or cleft palate
4. The frontonasal prominence is split into medial and lateral nasal prominences by the development of nasal pit on the ventrolateral aspect of the frontonasal prominence
5. Formation of lip occurs between the fourth and sixth weeks of gestation when the bilateral maxillary prominences fuse with the medial nasal prominence to form the lip and alveolus.
6. The secondary palate begins to form during the 6th week of development as the palatine shelves which are outgrowths from the maxillary prominences advance obliquely downward to lie horizontally over the tongue. The palatine shelves fuse with the previously formed primary palate and then from anterior to posterior the palatine shelves fuse in the midline so that by week 12 the plate is intact. Failure of these processes can result in clefting

Functional Anatomy

Lip

1. Integrity of the lip and oral sphincter is important for normal function of mouth
2. A defect in the lip results in abnormal insertion of orbicularis oris and loss of continuity of the vermilion border
3. Both of these defects need to be addressed in cleft lip repair
4. Mucocutaneous area of the lip is divided into three regions: The cutaneous skin of the upper lip and philtrum, an intermediate area of dry mucosa known as the vermilion and an intermediate area of moist mucosa
5. Orbicularis oris normally forms a full sling under the mucosal covering. Aberrant muscle due to the cleft results in insertion of the orbicularis oris into the dermis and nasal ala on the cleft side and insertion into the columella on the non cleft side

Nose

1. Abnormality of insertion of orbicularis oris contributes to nasal deformity seen as part of cleft lip
2. Insertion of orbicularis oris into the nasal alar base there is outward splaying of the lower lateral cartilage causing the alar base on the cleft side to sit more lateral and inferior than it should
3. Columella is shortened

Functional anatomy (contd)

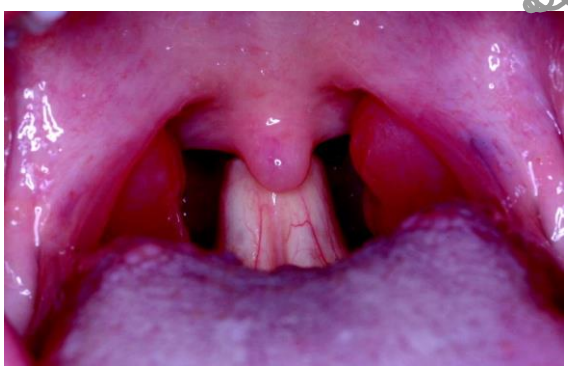
Palate

1. Clefts of palate are associated with bony and soft tissue abnormalities
2. Adequate closure of velopharyngeal port for speech and swallowing is the aim of palatal repair
3. Primary velar muscles are the levator veli palatini, palatopharyngeus and palatoglossus.
4. Levator veli palatini is the primary elevator of the palate. It originates from the medial portion of ET and from the petrous temporal bone and runs anteromedially to enter the middle third of velum between the two heads of palatopharyngeus to join with its partner from the opposite side, thereby forming a levator sling.
5. In patients with cleft palate the levator veli palatini no longer has this transverse orientation, but instead has longitudinal orientation getting inserted into the bony cleft margin and posterior palatine bones.
6. The palatoglossus & palatopharyngeus arise from the back of palatal aponeurosis and maxillary tuberosity. The palatoglossus muscle is a thin sheet of muscle that extends to form the anterior pillar of tonsil
7. Palatopharyngeus is a more substantial muscle that is split into two heads by the insertion of the levator veli palatini and runs down to form the posterior tonsillar pillar, and gets inserted into the thyroid cartilage and pharyngeal aponeurosis.
8. The palato pharyngeus and palatoglossus muscles act as depressor and along with levator veli palatini these muscles lengthen the velum
9. Tensor veli palatini, its primary function is to open the ET. Its fibers originate from the spine of the sphenoid, scaphoid fossa and lateral lamina of ET cartilage, forms a tendon which winds around the pterygoid Hamulus and spreads into a fibrous aponeurosis in the anterior third of the soft palate.

ENT Issues

1. All children are obligate nasal breathers for the first few months of life.
2. This is due to the high position of the infantile larynx that gradually descends as the infant ages.
3. In the neonate the anterior epiglottis can often be seen in the oral cavity behind the soft palate. The superior position of larynx allows the infant to suckle.
4. There are many congenital anomalies associated with cleft lip and palate and one of the most frequent is Pierre Robin sequence. These children have micrognathia and glossoptosis may result in upper airway obstruction

Upper airway obstruction can happen immediately following birth / develop progressively during weeks or months



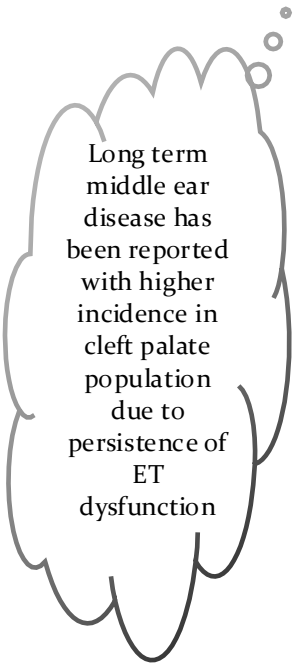
All children with craniofacial disorders should undergo overnight oximetry

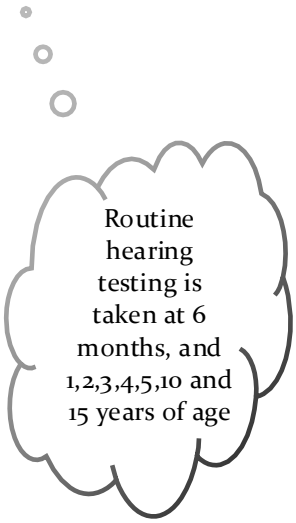
Upper airway obstruction management

Non surgical : Nasopharyngeal airway, CPAP
Surgical : Tongue lip adhesion, tracheostomy, mandibular distraction

Hearing Loss

Cleft lip & cleft palate children have hearing loss. It could be conductive, sensorineural or mixed.
OME is seen commonly in children with cleft lip and palate. About 97% of these children are thought to have had an episode of OME by the age of 2.
ET dysfunction has been attributed as a cause for increased incidence of OME in these children. This is due to malposition of the tensor veli palatini muscle.
Insertion of ventilation tubes at an early age, ideally during the time of palatal repair.
Long term middle ear disease has been reported to be of higher incidence in cleft palate children





Management

Nearly 75% of children with cleft lip and palate can be picked up as early as 13-16 weeks of intrauterine life

Issues to be addressed

Speech
Hearing
Appearance
Dental growth & Hygiene
Psychosocial health

Surgery

3-6 months – Primary lip + Nose repair + hard palate repair
9-12 months – Palate repair + /- ventilation tubes
4-5 years – Secondary speech surgery
8-10 years – Alveolar bone graft if required
15-18 years – Secondary rhinoplasty
16 years + - Orthognathic surgery if required

Nasoalveolar moulding

This is a form of presurgical infant orthopaedics designed to reduce the severity of the cleft deformity & move the alar cartilages into a more favorable position

Involves using an oral plate which is individually moulded with nasal stents attached