CLEFT LIP AND PALATE

Grand Rounds Presentation by
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Introduction

- Facial clefting is the second most common congenital deformity (after clubfoot).
- Affects 1 in 750 births
- Problems are cosmetic, dental, speech, swallowing, hearing, facial growth, emotional
- Otolaryngologist holds key role on CP team
Anatomy

- Hard Palate
  - Bones: Maxilla (Palatine Processes) + Palatine Bones (Horizontal Lamina)
  - Blood Supply: Greater Palatine Artery
  - Nerve Supply: Anterior Palatine Nerve
Anatomy

- Soft Palate
  - Fibromuscular shelf attached like a shelf to posterior portion of hard palate
  - Tenses, elevates, contacts Passavant’s Ridge
  - Muscles: Tensor Veli Palatini (CNV), Levator Veli Palatini (Primary Elevator), Musculus Uvulae, Palatoglossus, Palatopharyngeus (CN IX and X)
Embryology

- Primary Palate- Triangular area of hard palate anterior to incisive foramen to point just lateral to lateral incisor teeth
  - Includes that portion of alveolar ridge and four incisor teeth.
- Secondary Palate- Remaining hard palate and all of soft palate
Embryology

- **Primary Palate**
  - Forms during 4th to 7th week of Gestation
  - Two maxillary swellings merge
  - Two medial nasal swellings fuse
  - Intermaxillary Segment Forms:
    - Labial Component (Philtrum)
    - Maxilla Component (Alveolus + 4 Incisors)
    - Palatal Component (Triangular Primary Palate)
Embryology

- Secondary Palate
  - Forms in 6th to 9th weeks of gestation
  - Palatal shelves change from vertical to horizontal position and fuse
  - Tongue must migrate antero-inferiorly
Cleft Formation

- Cleft result in a deficiency of tissue
- Cleft lip occurs when an epithelial bridge fails
- Clefts of primary palate occur anterior to incisive foramen
- Clefts of secondary palate occur posterior to incisive foramen
Cleft Formation

- Secondary Palate closes 1 week later in females
- Cleft of lip increases likelihood of cleft of palate because tongue gets trapped.
Unilateral Cleft Lip

- Nasal floor communicates with oral cavity
- Maxilla on cleft side is hypoplastic
- Columella is displaced to normal side
- Nasal ala on cleft side is laterally, posteriorly, and inferiorly displaced
- Lower lat on cleft side - lower, more obtuse
- Lip muscles insert into ala and columella
Palatal Clefts

- Soft palate muscles insert on posterior margin of remaining hard palate rather than midline raphe.
- Associated Dental Abnormalities
  - Supernumery Teeth- 20%
  - Dystrophic Teeth- 30%
  - Missing Teeth- 50%
  - Malocclusion- 100%
Genetics

- Non-syndromic inheritance is multifactorial
  - Cleft Lip, With or Without Cleft Palate:
    - One Parent - 2%
    - One Sibling - 4% Two Siblings - 9%
    - One Parent + One Sibling - 15%
  - Cleft Palate:
    - One Parent - 7%
    - One Sibling - 2% Two Siblings - 1%
    - One Parent + One Sibling - 17%
Genetics

- Increased clefts with chromosome aberrations
- Clefts a part of a Syndrome 15-60% of time
- More than 200 syndromes include clefts
- Cleft Palate- Apert’s, Stickler’s, Treacher
- Cleft Lip +/- Palate- Van der Woude’s, Waardenberg’s
Epidemiology

- Cleft Lip +/- Palate - 2 Male: 1 Female
- Cleft Palate - 2 Female: 1 Male
- Cleft Lip +/- Palate - Native Americans > Oriental and Caucasians > Blacks
- Cleft Palate - Same among ethnic groups
- Environmental: Ethanol, Rubella virus, thalidomide, aminopterin
Epidemiology

- Increased Clefts with maternal diabetes mellitus and amniotic band syndrome
- Increased Clefts with increased paternal age
- Cleft Lip + Palate- 50%
- Cleft Palate- 30%
- Cleft Lip- 20%
- Cleft Lip + Alveolus- 5%
Management

- Team Approach
- Otolaryngologist has a pivotal role
- Initial Head and Neck Examination
- Speech Disorders
- Ear Disease
- Airway Problems
- Surgical Repair
Head and Neck Exam

- Head- facial symmetry
- Otologic- auricle and canal development and location, pneumatic otoscopy, forks
- Rhinoscopy- identifies clefting, septal anomalies, masses, choanal atresia
- Oral Exam- cleft, dental, tongue
- Upper airway- phonation, cough, swallow
Speech Disorders

- Errors in Articulation: Fricatives, Affricates
- Velopharyngeal Competence - Most important determinant of speech quality in cleft palate patients - 75% achieve competence after initial palate surgery
- Incompetence - nasal emission or snort
- Evaluation - Direct exam, Fiberoptic Exam
Ear Disease

- Cleft Lip- Incidence similar to normal pop.
- Cleft Palate- Almost all with ETD, CHL
- ETD- Due to abnormal insertion of levator veli palatini and tensor veli palatini into posterior hard palate
- ETD- Returns to normal by mid-adolescent
- Cleft Palate- Increased Cholesteatoma(7%)
Ear Disease

- **Otologic Goals For Cleft Palate Patients**
  - Adequate hearing
  - Ossicular chain continuity
  - Adequate middle ear space
  - Prevent TM deterioration

- **Indications for Myringotomy Tubes**
  - CHL, Persistent/Recurrent effusion, Retraction
  - Cleft palate: Multiple BMTs from 3mo. - 12 yrs
Airway Problems

- More common in Cleft Palate patients with concomitant structural or functional anomalies.
  - e.g. Pierre-Robin Sequence
    - Micrognathia, Cleft Palate, Glossoptosis
    - May develop airway distress from tongue becoming lodged in palatal defect
Surgical Repair- Cleft Lip

- Lip Adhesions-
  - 2 weeks of age
  - Converts complete cleft into incomplete cleft
  - Serves as temporizing measure for those with feeding problems
  - May interfere with definitive lip repair
  - Less often needed in recent years due to wider variety of specialty feeding nipples
Surgical Repair- Cleft Lip

- Cleft lip repaired at 10 weeks
- Rotation-advancement method- Most common in the U.S.
- Nine Landmarks
  - Rotation Flap cuts made first
  - Advancement cuts made next
  - Cleft side nasal ala cuts made last
Surgical Repair- Cleft Palate

- Several Techniques- Trend is towards less scarring and less tension on palate
- Scarring of palate may cause impaired mid-facial growth (alveolar arch collapse, midface retrusion, malocclusion)
- Facial growth may be less affected if surgery is delayed until 18-24 months, but feeding, speech, socialization may suffer.
Surgical Repair- Cleft Palate

- Bardach Method- Two Flap technique
  - Medial incisions made, which separate oral and nasal mucosa
  - Lateral incisions made at junction of palate and alveolar ridge
  - Elevate flaps, preserve greater palatine artery.
  - Detach velar muscles from posterior palate
  - Close in 3 layers
Non-Surgical Treatment

- Dental Obturator
  - For high-risk patients or those that refuse surgery.
  - Advantage: High rate of closure
  - Disadvantage: Need to wear a prosthesis, and need to modify prosthesis as child grows.
Conclusions

- Cleft Lip and Palate are common congenital deformities that often affect speech, hearing, and cosmesis; and may at times lead to airway compromise.
- The otolaryngologist is a key member of the cleft palate team, and is in a unique position to identify and manage many of these problems.