Clefts of the Lip, Alveolus and Palate
Overview

- Introduction
- Basic Science
- Timetable of Events
  - neonatal
  - toddler
  - gradeschool
  - teenage
- Surgical Procedures
- Conclusion/Future Directions
Introduction

A TEAM APPROACH IS REQUIRED

- pediatrician
- surgeon
- OMFS
- dentist
- ENT
- psychiatrist
- speech
- nurse coordinator
Introduction

- Most common congenital malformation of H and N (1:1000 in US; 1:600 in UK)
- Second most common overall (behind club foot)
Epidemiology

 Syndromic CLAP

 - associated with more than 300 malformations
  - Pierre Robin Sequence; Treacher-Collins, Trisomies 13,18,21, Apert’s, Stickler’s, Waardenburg’s

 Nonsyndromic CLAP

 - diagnosis of exclusion
Syndromic CLAP

- Single Gene Transmission
  - trisomies 21, 13, 18
- Teratogenesis
  - fetal alcohol syndrome
  - Thalidomide
- Environmental factors
  - maternal diabetes
  - amniotic band syndrome
Epidemiology: continued

- Isolated cleft palate genetically distinct from isolated cleft lip or CLAP
  - Same among all ethnic groups (1:2000, M:F 1:2)

- Isolated CL or CLAP
  - Different among ethnic groups
    - American Indians: 3.6:1000 (m:f 2:1)
    - Asians 3:1000 (m:f 2:1)
    - African American 0.3:1000 (m:f 2:1)
Embryology

- Primary versus secondary palate
  - Divided by incisive foramen
    - Primary palate develops 4-5 wks
    - Secondary palate develops 8-9 wks

- Primary palate
  - Mesodermal proliferation of frontonasal and maxillary processes
  - Never a cleft in normal development
Embryology: continued

- Secondary palate
  - medial ingrowth of lateral maxillae with midline fusion
  - always a cleft in normal development
    - macroglossia, micrognathia may provide anatomical barriers to fusion
Classification

Veau Classification - 1931

- Veau Class I: isolated soft palate cleft
- Veau Class II: isolated hard and soft palate
- Veau Class III: unilateral CLAP
- Veau Class IV: bilateral CLAP

Iowa Classification - a variation of Veau Classification
Complete Clefts
- absence of any connection with extension into nose
- vomer exposed

Incomplete Clefts
- midline attachment (may be only mucosal)
  - ex: submucous cleft (midline diastasis, hard palatal notch, bifid uvula)
Anatomy - Normal

- **Lip:** “Cupid’s Bow”
- **Maxilla**
  - primary/secondary palates
  - soft palate
  - alveolus
  - maxillary tuberosity
  - hamulus
Anatomy: palatal muscles

- Superior constrictor
  - primary sphincter
- Tensor veli palatini
  - tenses palate
- Levator Veli palatini
  - elevates palate
  - dilates ET
- Salpingopharyngeus, palatopharyngeous, palatoglossus: minor contribution
Cleft Anatomy

- Unilateral Cleft Lip and alveolus
  - Lack of mesodermal proliferation
    - Cleft of orbicularis
      - Medial portion to columella
      - Lateral portion to nasal ala
    - Cleft of alveolus
      - Alveolar bone graft
Cleft Anatomy - The Nose

- Ipsilateral LLC
  - flattened
  - rotated downward
- Short columella
- Bifid tip
Cleft Antatomy: continued

- Bilateral Cleft Lip/Alveolus/nose
  - Duplication of unilateral defect
    - Premaxilla
    - Orbicularis to alar cartilages bilaterally
    - Bifid tip
    - Extremely short columella
Cleft Anatomy: continued

- Clefts of the primary hard palate/alveolus
  - Cleft alveolus always associated with cleft lip
  - Cleft lip not necessarily associated with cleft alveolus
  - By definition there is opening into nose
Clefts of secondary palate

- Failure of medial growth maxillae
  - fusion at incisive foramen
  - macroglossia
- Submucous vs. complete
- Vomer
Multidisciplinary Approach

- These are not merely surgical problems
  - Requires team approach throughout life
    - neonatal period
    - toddler
    - grade school
    - adolescence
    - young adulthood
The Neonatal Period

**Pediatrician:**
- directs care
- establishes feeding
  - complete clefts preclude feeding
    - breast feeding not possible
    - a soft, large bottle with large hole is required
    - a palatal prosthesis may be required
The Neonatal Period

• Presurgical Orthodontics (Baby Plates)
  - Molds palate into more anatomically correct position
  - decreases tension
  - may improve facial growth
  - Grayson, presurgical nasal alveolar molding (PSNAM)
The Neonatal Period

- Surgical Repair
  - Cleft Lip
    - In US - “the rule of tens” - 10 wks, 10 lbs, Hgb 10
    - Lip adhesion vs baby plates
  - Cleft Palate
    - Varies from 6-18 months - most around 10 mo
    - Early repair may lead to midface retrusion
    - Early repair improves speech
The Toddler Years

Priority: Speech

“Cleft errors of speech” in 30%
- primary defects - due to VPI (hypernasality)
  - consonants are most difficult sounds (plosives)
- secondary defects - due to attempted correction
  - glottic stops, nasal grimace

Velopharyngeal insufficiency
- diagnosed by fiberoptic laryngoscopy or BaSw
- surgical repair after failed speech therapy - usually around age 4
The Toddler Years

- Growth hormone deficiency
  - 40 times more common in CLAP
  - suspects when below 5% on growth chart
The Grade School Years

Three primary issues

- Orthodontics
  - poor occlusion
  - congenitally absent teeth
- Alveolar bone grafting
  - fills alveolar defect - around age 12
- Psychological growth
  - considered standard of care
The Teenage Years

- Midface retraction
  - etiology - ?early palatal repair
  - surgical correction around age 18

- Psychological development
  - counseling standard of care

- Rhinoplasty
  - usually last procedure performed, around age 20
Surgical Techniques

**Cleft Lip Repair**
- **unilateral**
  - rotation-advancement flap developed by Millard
- **complications**
  - dehiscence
    - infection
  - thin white roll
    - excess tension
Surgical Techniques

Cleft Lip Repair

- bilateral
  - bilateral rotation advancement with attachment to premaxilla mucosa
- complications
  - dehiscence
  - thin white roll
Surgical Techniques

- Velopharyngeal Incompetence
  - Superior based pharyngeal flap
  - Sphincter pharyngoplasty
    - Palatopharyngeus
  - Complications
    - Continued VPI
    - Stenotic side ports
Surgical Techniques

- Alveolar Bone Grafting
  - iliac crest bone graft
  - complications
    - infected donor site
      - hematoma
    - failed graft
      - dehiscence
      - palatal prosthesis
Surgical Techniques

- Midfacial Advancement
  - LeFort osteotomies
    - Leave vascular pedicle attached in back of maxilla - prevents necrosis
  - Complications
    - Malocclusion
    - Infection
    - Necrosis
Surgical Techniques

* Rhinoplasty
  - standard techniques
    - tip projection
    - alar rotation
    - columnellar length
  - complications
    - alar stenosis
Controversies: Otologic Disease

>90% have COME

- Robinson, et al
  - prospective, 150 patients - 92%
- Muntz, et al.
  - retrospective, 96%

Pathology: ETD (controversial)

- abnormal muscular attachment
- Huang, et al. - Cadaveric study
  - palatal repair restores ET function. ?Midface growth?
Controversies: Timing of Repair

Early repair

- **Advantage:** improved speech
  - Rohrich, et. al; retrospective study. The earlier the repair, the better speech.

- **Disadvantage:** worsening midface retrusion
  - Rohrich, et. al; people with unrepaired palates have less midface retrusion
Controversies: VPI

*Surgical Repair*

- **Reserved for failure of speech pathology**
- **Pharyngeal Flap - superiorly based**
  - **Advantage:** time tested, severe cases
  - **Disadvantage:** passive obturator
- **Sphincter Pharyngoplasty (palatopharyngeus rotation flap)**
  - **Advantage:** active sphincter
  - **Disadvantage:** new technique
Controversies

- Presurgical Nasal Alveolar Molding
  - Molds palate, alveolus and nose
    - Advantage: excellent early results
    - Disadvantage: no long term results
  - Grayson, et al.
Conclusion and Future Directions

- Multidisciplinary approach
- Not merely a “surgical problem”
- Alveolar bone grafting
- PSNAM
- Pharyngoplasty vs. pharyngeal flap