Diagnosis, Care and Management of The Pediatric and Craniofacial Patient

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Dr. Stone has no relevant financial relationships to disclose.

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Embryology

- Face develops from 5 prominences
  - Frontonasal
  - Bilat maxillary prominences
  - Bilat mandibular prominence
- Lip development 4-7 wks gestation
- Primary palate is formed from 4-7 wks (fusion of MNP)
- Secondary palate devel 8-12wks
  - Fusion of palatal shelves (medial maxillary process).

Embryology

- Cleft lips occur from failure of fusion of medial nasal prominence and maxillary prominence
- Cleft palate occurs from failure of fusion of medial nasal with the lateral palatine shelves (maxillary process)
**Incidence**

1 affected child
- 4% for second child
- 2nd child – risk becomes 9%
- One parent – 4%
- 1 Parent 1 Child – 17%
- 1st, 2nd, 3rd degree relative – 4%, 0.7%

**Classification**

**Classification of Cleft Lip and Palate**
Craniofacial Clefts

Anatomy
The Cleft Nasal Deformity

Overview of Cleft Surgical Care and Timing

- Prenatal Counseling
- Pre-op Orthopedics
- Lip Repair* 3 months
- Palate Repair* 10-18 months
- Speech Surgery (VPI) 4-5 years
- Alveolar Bone Grafting* 7-10 years
- Orthognathic Surgery 14-18 years
- Rhinoplasty 14-18

Cleft Pre-Operative Orthopedics

- Active
  - Latham appliance

- Passive
  - Nasoalveolar molding
  - Lip taping
  - Lip adhesion
• General concept is to convert a difficult wide cleft to a simpler procedure
  – Narrower cleft
  – Convert complete to incomplete

Latham Appliance

Samuel Berkowitz, D.D.S., M.S., J.I.C.D., Marta Mejia, D.D.S., and Isaac Worth, Ph.D.
Nasoalveolar Molding (NAM)

Lip Adhesion
Lip Adhesion

• When is it useful
  – When alignment of a protruding prolabium would reduce dehiscence risk or decrease surgical difficulty
  AND
  – When finances do not allow for the prosthesis
  – No access to a team with NAM capabilities
  – Geographical access is difficult
  – Parental compliance is an issue.
So What is the Best Choice

- Avoid active orthopedics such as Latham device

- Passive measures
  - NAM: best in control, but will require multiple appointments and may be difficult from resources point
  - Lip Taping: Least invasive and easy to complete but requires parent cooperation
  - Lip Adhesion: Requires surgery, but requires least follow-up and parental cooperation
Timing

- **Rule of 10's**
  - 10 lbs
  - 10 g
  - >10 weeks

- **Modifications**
  - More pertinent medical needs
  - Need for presurgical orthopedics

Scarless Fetal Wound Healing: A Basic Science Review

Wounds scarlessly in age-dependent. In other words, fetal skin heals scarlessly before a certain gestational age, after which point typical scar formation occurs. In humans, scarring of wounds begins at approximately 24 weeks of gestation, whereas in mice scarring of wounds begins on embryonic day 18.5 (average gestation period for mice is 20 days). This transition point, however,

Definitive Tech

- **Straight Line Repair**
- **Rotation-Advancement**
- **Geometric (Triangular Flap)**
  - Randal Tenison
  - Fischer
Straight Line Technique

- DeLaire

Millard Type

- Adaptable to different cleft varieties
- May be adjusted during the procedure
- Scar resides on philtral column without inferior triangular scars

Challenges
- May be difficult to establish lip height in wide cases
Typical Millard Scar

Geometric Repair

• AKA - Tennison Repair
• Pros
  – Establishes excellent lip length
• Challenges
  – Requires exacting measurements and surgical plan cannot be altered once begun
  – Inferior triangular scar may be obvious
Unilateral Cleft Lip Repair: An Anatomical Subunit Approximation Technique
Bilateral Cleft Lip
Primary Bilateral Cleft Lip/Nose Repair Using a Modified Millard Technique

G.E. Ghazi, DDS, MD, FACS, Jason L. Ringman, DDS, MD
Unilateral nasal features

Bilateral Nasal Features

Fig. 1. Rhinoplasty (A) and facial photograph (B) of bilateral choanal atresia showing short medial crus and a deformed nasal bone.
Treatment of Cleft Nasal Deformity

- Primary – At time of cleft lip
- Intermediate – before completion of nasal growth
- Secondary (definitive septrhinoplasty) – after completion of nasal growth
  - 15 for females
  - 17 for males

Primary Rhinoplasty
Primary Rhinoplasty

Post-op Care

Cleft Palate Repair

- Timing
  - 9-18 months
  - 10 – 12 months on average
  - At least 12 months with Pierre Robin Sequence
The Battle of Speech vs Growth

- Historically, have had proponents of early, intermediate, delayed repair
  - No speech benefit shown with repairs completed before 9 months
  - Midface growth stunted with earlier surgeries
  - Growth maximized by delaying surgery, but speech permanently affected when repairs delayed beyond 18-24 months
  - Consensus has adopted 10-18 months for most children
**Bardach Two-Flap Palatoplasty**

- **Pros**
  - Can close wide clefts
  - Low fistula rate
  - Can complete intravelar veloplasty

- **Cons**
  - Large dissection for small clefts
Double Opposing Z-Plasty

Pros
• Retro displaces the palatal muscles
• Lengthens the palate by nature of Z-plasty
• Good for small clefts with limited dissection

Cons
• More difficult / more experience required for large clefts
• Muscles are reapproximated in a random pattern

Velopharyngeal Insufficiency
• Typically around age 4-5 years of age
• Must be able to cooperate with exam and speech therapist
• Evaluated by:
  – Nasopharyngoscopy
  – Videofluoroscopy
  – Nasometry

Background
• 3 Main Terms
  – Velopharyngeal insufficiency
    • Due to anatomical deficiencies
      – Shortened soft palate
      – Inappropriate repair of levator muscle
  – Velopharyngeal incompetence
    • Poor muscle function
      – Myasthenia gravis, or syndromes with hypotonia
  – Velopharyngeal mislearning
    • Failure to coordinate speech despite normal structures and physiology
Velopharyngeal Valve

Variations in Function
- Coronal Pattern
- Sagittal Pattern
- Circular Pattern

Treatment Options
- Posterior Pharyngeal Fat Injection
  - Reserved for small gaps
- Palatal Push Back
  - May be combined with larger procedures
- Sphincter Pharyngoplasty
  - For deficient lateral wall motion with preserved AP motion
- Superior Base Pharyngeal Flap
  - For large gaps
  - Especially helpful with preserved lateral wall motion
Evaluation of VPI Speech Disorders

- **Multiview videofluoroscopy**
  - Provides several views for better 3-dimensional view
    - Speaking in lateral w/ and w/o barium
    - Swallowing barium in lateral
    - Speaking in base view after barium is instilled in nostrils
    - Speaking in frontal view

- **Pressure-flow method**
  - 1 pressure tube in the lip and nostril
  - 1 flow tube in 2nd nostril
  - VP gap calculated based on Bernoulli’s principle
  - May calculate nasal ram pressure (NRP) in infants

- **Nasometry**
  - Output is called % nasalance
  - Use nasal and oral microphone to measure acoustic energy
  - Normal when reading “Zoo” is <32%
Additional Considerations

• Ideally sleep study should be completed prior to pharyngeal surgery
  – If OSA present, must be careful with the degree of correction
• C in syndrome

Speech Appliances

• 3 primary indications
  – VP gap is large with limited movement of structures
  – Perceived symptoms are severe
  – Medical contraindications to surgery
• Disadvantages
  – Compliance may be problematic
  – Not recommended w/ poor hygiene

Superior Based Pharyngeal Flap

• Allows dissection of the posterior pharynx with constrictors to be inset into the soft palate
Superior Based Pharyngeal Flap

- N = 65
- 83.1% showed normal velar function
- 66.1% showed normal or near normal speech
- 89.2% noted snoring
- ECG eval’d in 33 pts showed only one with right ventricular hypertrophy signifying OSA

Pharyngeal Flap and OSA

- 90% had immediate post-op OSA
- 20% had OSA at 3 months

Sphincter Pharyngoplasty
Sphincter Pharyngoplasty

Comparison of Flap Outcomes
- Group 1 – Pharyngeal Flap
- Group 2 – Pharyngoplasty
- Procedures where tailored based on videofluoroscopy

Alveolar Cleft Management
- Gingivoperioplasty (GPP)
- Alveolar Bone Grafting
  - Autogenous bone
  - Allogenic bone
  - BMP
- Closure of cleft in conjunction with orthognathic surgery
Alveolar Cleft Management

- Purpose of the bone graft
  - Closure of the oro-nasal fistula
  - Establishment of continuity with the premaxilla in bilateral cases
  - Provide bony support of the nasal base
  - Provide bone for eruption of permanent dentition
  - Bonus: Provide bone for implant placement

Alveolar Bone Grafting

- Classically completed when the canine root is 1/2 - 2/3 formed
- Prefer to have arch expansion completed prior during phase 1 or
Anterior Iliac Crest Bone Harvest

Alveolar Bone Grafting

• Defect is pyramidal
• Requires closure at the nasal floor, palatal, buccal mucosas
Alveolar Bone Grafting

Challenging Cases and Alternative Treatments
• What will be next?
Missing Lateral Incisor Management

- Permanent Options
  - Canine Substitution
  - Implant Reconstruction
  - Fixed Prosthesis
- Interim options
  - Removable Appliance
  - Temporary Mini-implant

Their studies validated previous studies in which the lay public preferred canine substitution.
Canine Substitution

- **Pro's**
  - Decreased cost of therapy
  - No intermediate removable appliance

- **Con's**
  - Esthetic work required to convert canine to a lateral
  - Difference between substituted canine and c (i.e. gingival margins)
  - 1st premolars are shorter and narrower than

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Dental Implants

- **Pros**
  - Can re-establish Class I relationship
  - No need for adjustment of the canine

- **Cons**
  - Require interim option for lateral incisor
  - Often require additional bone grafting for implant placement
  - Cost
  - Typically challenging cases
Dental Implant

• Reviewed 7 retrospective and 4 prospective
  – Overall success rate 91.5%
  – Additional bone grafting required 43.1%

• Recommended CBCT for all patients to receive implants
  secondary to high rate of inadequate bone

• Dehiscence increases to 25% when implant is placed
  simultaneous with secondary grafting

Temporary Prosthesis

• Mini-Implant Reconstruction