

Cleft Palate Craniofacial Center Symposium

April 7, 2025

Welcome!



History

- Established 1985
- Coordinator
- Specialists: Genetics, Otolaryngology, Speech and Hearing, Plastics, Orthodontics, Pediatric (MD + DDS), OMFS, Parent advocate, Social Work etc.
- Team conferences (monthly)



Mission

- To provide **patient centered care** for children with CP/CF deformities
- To **guide and support families** to negotiate financial and social barriers



What We Do

- Provide diagnostics
- Prepare families for the timing and coordination of diagnostics and services
- Provide services
- Assist families and Communicate with managed care
- Assist primary care physicians
- Refer to specialists
- Educate community



Team Presentations

- Genetics
- Feeding/Lactation Specialist
- Plastic Surgeon
- Otolaryngology (ENT)
- Audiology
- Speech Orthodontics
- Maxillofacial Surgery
- Prosthodontist
- Social Work
- Caregivers



The Genetics of Cleft Lip And Palate

Barth, Shwin-Parson, MD
Professor, Clinical Genetics and Pediatrics
Director, Clinical Genetics

What is Cleft Lip and Palate?

A Cleft is a separation in the Lip or in the Roof of the mouth.
The lip forms at about the 7th Week of Pregnancy
Approximately 1 in 500 babies born in the United States has some type of Cleft.



What Causes this Condition?

In most cases, the exact cause of clefting is unknown.

There seems to be 2 interrelated factors:

Our Genes
The Environment

What Causes this Condition?

In most cases, the exact cause of clefting is unknown.

There seems to be 2 interrelated factors:

Our Genes
The Environment

Environment

This term refers to all those factors that are not in the genes and chromosomes. It can be exposure to drugs, chemicals and infectious agents as well as where the baby begins to grow in the uterus.

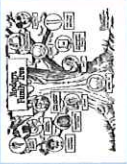
- These include heavy alcohol consumption, smoking and certain meds for acne and others necessary to control epilepsy in the mother. To cause a cleft, an environmental exposure must have occurred before the formation of these structures. Anything that happened after this time has no bearing on the cause of the cleft.

What is involved in a Genetic Evaluation

The most essential part of a genetic evaluation is verifying that the cleft is isolated, not part of a syndrome. Majority of children who have clefts do not have other genetic problems.

TOOLS

- History
 - Prenatal & medical
- Family history
- 3 generation pedigree
- Physical examination
- Further testing
 - Radiology
 - Genetic testing as indicated
- Determination of Recurrence Risk



Examples of Genetic Causes

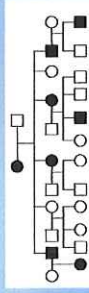
Genes

- Single Gene Defects
 - Dominant
 - Recessive
 - X-Linked
- Chromosomal Defects
 - Trisomy 13 (Patau)
 - 5p- (Cri-du-chat)
 - 4p- (Wolf-Hirschhorn)
- Tools
- Chromosome analysis, Chromosome Microarray, Gene Panels, Whole Exome Sequencing via blood, amniotic fluid or tissue/saliva.

Chromosomal Defects



Autosomal Dominant Inheritance



- Van der Woude Syndrome
- Facial Cleft (27%)
- 4x Inward Lip Flip (66%)
- 4x Family History
 - Single Most Common Inherited Cause of Isolated Cleft (p. 126)
- AD, Incomplete penetrance & variable expressivity



Velocardiofacial Syndrome

- Velo – Cleft palate (overt or submucous), hypomasal speech
- Cardio – Conotruncal defects (VSD, TOF)
- Facial – Microcephaly, long face with malar flattening, small palpebral fissures, long nose with bulbous tip and notched alae nasi, dysplastic ears, micrognathia
- CNS – Developmental delay/IMR
- Ext – Long, thin fingers



A Positive FISH Study on an Individual with Individual with Velocardio Facial Syndrome Using a 22q11 Probe



Stickler syndrome

- Cleft palate/Pierre Robin sequence
- Nearsightedness, vitreoretinopathy, myopia, Amblyopia, astigmatism
- Flat face, molar Hypoplasia
- Hearing loss, sensorineural hearing loss
- Hyperextensible joints/arthralgias
- Specific findings on bone x-rays: Knock-knees, epiphyseal dysplasia
- Mitral valve prolapse (46%)



Treacher Collins Syndrome

The child usually will show normal intelligence. Examination of the infant may reveal a variety of problems, including:

- Abnormal eye shape
- Protruding ears
- Clefts in the face
- Small jaw
- Low-set ears
- Abnormally formed ears
- Abnormal ear canal
- Defects in the eye (coloboma that extends into the lower lid)
- Decreased eyelashes on the lower eyelid
- Genetic tests can be done to look for mutations in the TCOF1 gene



Autosomal Recessive Inheritance – Smith-Lemli-Opitz Syndrome

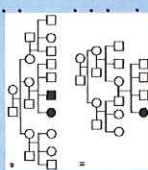
Facial clefting

- Distinctive appearance
- Upturned nose
- Wide mouth
- Low set ears

- Heart defects
- Male genital ambiguity
- Syndactyly of the 2nd & 3rd toes

Inheritance:

- Autosomal recessive
- Defect in cholesterol metabolism
- Deficiency of cholesterol reductase deficiency
- Low cholesterol, High 7-OH-cholesterol



Multifactorial or Polygenic Inheritance - Most cases of isolated non-syndromic cleft lip +/- cleft palate fall into this category.



Pierre-Robin Sequence
Vs
Pierre-Robin Syndrome



Isolated Posterior Cleft of the Soft Palate

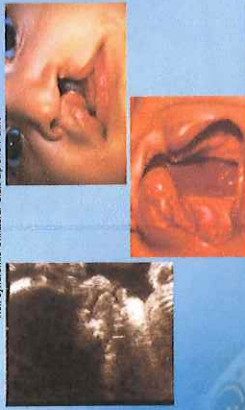
Environmental Teratogenic

- Main offending agents are
- Retinoic Acid
- Valproic Acid
- Warfarin or Coumadin
- Alcohol



Discussion of various kinds of Prenatal Diagnosis if couple is interested

Non-syndromic Unilateral Cleft Lip and Palate



FOLIC ACID

ALL WOMEN OF CHILD-BEARING AGE SHOULD BE ON A FOLIC ACID SUPPLEMENT - 0.4 MG OR FOR HIGH RISK, 4 MG

3 months preconception through the second trimester

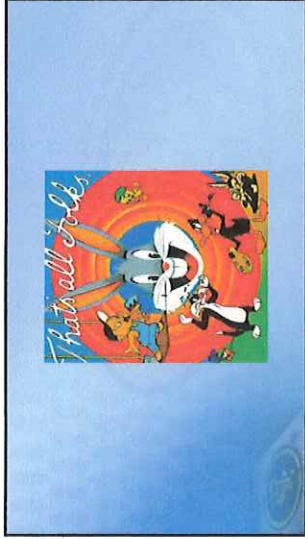
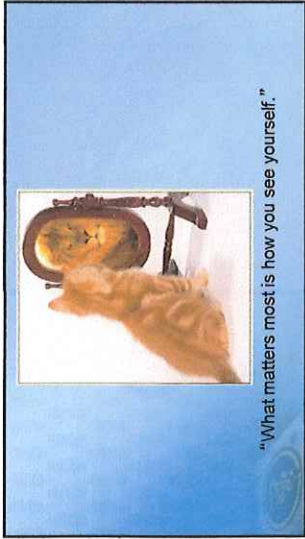
TO SUMMARIZE:

The most essential part of a genetic evaluation is verifying that the cleft is isolated, not part of a syndrome. Majority of children who have clefts do not have other genetic problems.

TOOLS

- History
- Prenatal & medical
- Family history
- 3 generation pedigree
- Physical examination
- Family testing
- Radiology
- Genetic testing as indicated
- Determination of Recurrence Risk
- Most cases are isolated and Multifactorial in origin
- Recommendations of Folic Acid for all future pregnancies





Feeding infants with cleft lip and palate



 Kent Ebermann, FSSLD
 M.S., CCFP, FACS
 Cleft Palate-Craniofacial Team
 Infant Feeding

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Feeding Goals

Create an individualize feeding plan to promote a positive feeding experience

- adequate nutrition
- weight gain

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Feeding Factors

- Time
- Calories
- Bottle Type
- Nipple flow

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Infants with Cleft Lip Only

- Typically feed with limited difficulty
- Maybe be able to breast feed
 - Latching baby to the breast can show what lip repair will resemble
- Standard bottles for expressed breasts milk or formula


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Infants with a Cleft Palate (with or without Cleft Lip)

Loss of/ reduced suction results in:

- Difficulty feeding at the breast or with standard bottles
- Longer feeding sessions
- Burning too many calories
- Poor weight gain patterns
- Reflux
- Feeding Avoidance

Exclusive breast feeding is often not possible

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Bottles and Nipples



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Popular Bottles

- Medela Special Needs Feeder (aka Haberman)
 - Feeder expressed
 - Valve that allows forward flow of nipple (slow, medium, fast)
- Dr. Brown's Specialty Feeding System
 - Infant paced feeding valve
 - Infant expressed with lingual/chomping movements
 - Various nipple options for improved flow rates

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Poor Weight Gain

- Increase Caloric Content of breastmilk with fortifier
 - Increase calories in formula
 - Strict feeding duration
 - Alternative feeding methods
 - Feeding Evaluation/therapy
- "Use it or lose it"*



Infant Healthcare Providers

- Support during acute hospital stay
- Refer to other healthcare providers and Early Intervention (if not already involved)
- Craniofacial team for continuity in care

Thank you.



Speech Pathology Intervention with Cleft Lip/Palate

Kerri Elorriaga M.S. CCC/SLP
Stony Brook University Medical Center

Speech Therapy Timeline

- Initial Meeting- feeding concerns/speech-language milestones, EI referral
- Speech-Language Evaluation ~2 YRS
 - Focus on articulation (sound production across word positions)
 - Assess for developmental, phonological, phonetic, phonological errors and maladaptive/compensatory errors (learned vs. CP)
- Speech Resonance Evaluation ~4 YRS.
 - Focus on resonance (oral vs nasal airflow)
- Videonasendoscopy- as needed

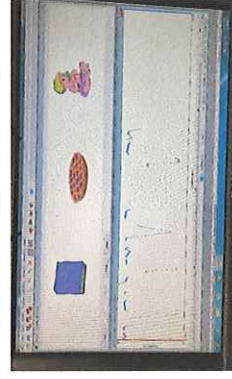
Speech Resonance Evaluation

- Articulation Testing
- Floxite Mirror for sounds and phrases
- Computerized Nasometry

Floxite Mirror Test



Computerized Nasometry

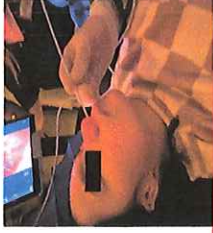


Videosoendoscopy

- Flexible endoscope passed through nose
- VF port viewed and videoed recorded during speech
- Sounds in isolation, phrases, connected speech
- Assess for pattern of closure (velar movement, lateral wall movement, posterior-velar wall)


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www.stonybrookchildrens.com

Videosoendoscopy



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Videosoendoscopy




Management of Velopharyngeal Insufficiency

- Surgical Intervention
- Speech Therapy
- Articulation drill with home carryover

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Thank you.



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Oral Health and Appliance Therapy for the Cleft Patient

Hygienic Concerns for both Patient and Appliance
 Kimberly A. Patterson, DDS, MS

Teamwork...

- Pediatric
- Pediatric Speech
- Pediatric Speech Pathologist
- OMRP
- Craniofacial
- Cleft Lip/Palate
- ENT
- Audiologist
- Speech Pathologist
- Prosthodontist
- Orthodontist
- New Center for Social Worker



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 Pediatric ENT/MSD

Neonatal Orthopedics - taping



Unilateral cleft lip and palate



Bilateral cleft lip and palate

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 Howek p. 82

Neonatal Orthopedics - taping



Time in proper counseling of parents regarding tape application....

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Neonatal Orthopedics - taping

Unilateral cleft lip and palate

Bilateral cleft lip and palate

Courtesy: Dr. Adewumi

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Neonatal Orthopedics

Courtesy: Dr. Adewumi

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Perspective on Size of Appliance

Dean Fig 24.14

Courtesy: Dr. Adewumi

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Neonatal Orthopedics

Taped lip guides alveolar growth along with adjustments to obturator

Weekly modifications to obturator guide alveolar growth

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Neonatal Orthopedics

Sequential maxillary arch dental models demonstrating maxillary orthopedic device use to gradually close unilateral complete cleft lip and palate.

Notice that as the cleft defect closes with time, lateral arch dimension is maintained, which produces optimal maxillary arch symmetry

1. REC. 2. BONDING 3. BONDING 4. BONDING

Dean Fig 24.18

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Neonatal Orthopedics

Relief areas to encourage directional growth

NAM appliance

Preauricular or molding plate / feeding plate

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Neonatal Orthopedics

NAM appliance

Nowak, p. 83

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Neonatal Orthopedics

Preauricular or molding plate / feeding plate

NAM appliance

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Pediatric Dentist's Role:

Role in the team:

- Coordinating with team members
- Provision of ongoing dental care
- Preventive treatments
- Restorative procedures
- Interceptive orthodontic therapy as requested

Pediatric Dentist's Role:

Preventive Role	Non-operative Role
<ul style="list-style-type: none"> Parent counseling for diet and oral hygiene maintenance Daily preventive advice for the child for caries prevention Consent of feeding plate Prevental appliances for correction or retard progression Early preventive advice for the child for caries prevention 	<ul style="list-style-type: none"> Post-operative oral hygiene maintenance through professional oral hygiene maintenance aids Prevention care (topical fluoride and sealants) application Restorative care for the various teeth and endodontic treatment for retained teeth Orthodontic correction of malaligned teeth Retained plate for correction of speech problems

Luzzi et al, Table 1

Oral Hygiene

- Caries Risk Factors:**
- Enamel defects (hypoplasia)
 - Parents overwhelmed, oral health low priority
 - Use of an acrylic obturator
 - Longer oral clearance times
 - Permissive parenting, highly cariogenic diet, less-than-adequate oral hygiene home care
 - Significant scarring / malalignment cleft area more difficult to clean
 - Orthodontic appliances
 - Oral aversion / fear of toothbrushing
 - Cognitive or motor impairment

Dean Table 24.3

Oral Hygiene

- Periodontitis Risk Factors**
- Poorly developed osseous support / connective tissue attachment
 - Anomalies of size, shape, and number, malalignment
 - Inadequate oral hygiene
 - Orthodontic appliances
 - Subgingival restorations
 - Sporadic and infrequent dental evaluations

Dean Table 24.3



Oral Hygiene

Swabs ("Toothbat") or Fingersip Brush accclimate infant to having oral tissues manipulated



Oral Hygiene

Smaller head toothbrushes – rubber grip of handle of a second toothbrush can be used to keep mouth open during oral hygiene



developed at University of Porto

Oral Hygiene

Denture brush – curve cleans cleft site, brush can clean appliances



Oral Hygiene

With orthodontic appliances...



Oral Hygiene

Without orthodontic appliances...



Infections

- Candida Albicans**
- opportunistic
 - Clean appliances with dish soap and warm water
 - Leave appliances out until infection resolves
 - Nystatin drops / gel tid x 10d



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Gwynson BH, Mallit D. Nasolabial Molding for Infants Born with Clefts of the Lip. *Alveolar Ridge Resection in Face Surg*. 2005 Nov;13(4):294-301. doi: 10.1055/s-2009-95994. PMID: 1742884748.

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All things are possible with a good team!

Thank you!

Kimberly.hathorn@stonybrook.edu

Stony Brook Children's

Cleft Lip and Palate

• Incidence: 1/700 births worldwide

• Can cause difficulty with:

- Eating
- Breathing
- Hearing
- Speaking
- Social integration
- Thriving

Stony Brook Children's

Cleft Care is Team Care

• **Multidisciplinary Team Approach**

- Genetics
- Feeding/Lactation
- Speech
- Audiology
- Otolaryngology
- Dentistry
- Orthodontics
- Prosthodontists
- Oral Surgery
- Social Work
- Plastic Surgery

• **Parents**

- 38 years of service
- Comprehensive, centralized patient care
- Continuity, long-term follow up
- Our overarching goal is outstanding outcomes for patients and parents in Suffolk County

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Cleft Lip and Palate

Alvin Wong, MD
Assistant Professor of Surgery
Craniofacial & Cleft Palate Symposium
April 7, 2025

Let us help you Personalize your experience in offering with our patients organizations and/or products mentioned in providing through report for this presentation.

Stony Brook Children's

Cleft Lip and Palate: Timing of Treatments Shows Need for Multidisciplinary Team

- Birth: Diagnosis, genetics, feeding, +/- presurgical orthopedics
- 3-6 months: Cleft lip repair
- 9-18 months: Cleft palate repair and myringotomy tubes, speech therapy
- 4-6 years: Pharyngoplasty if needed
- 5-6 years: Minor lip/nose revision if needed
- 8 years: Closure of alveolar cleft and orthodontic treatment
- 16-18 years: Orthognathic surgery if needed
- 16-18 years: Scar revisions +/- cleft rhinoplasty if needed

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Prenatal Diagnosis

- Allows for prenatal counseling, no surprise when child is born
 - Can increase parental anxiety
- About 15% of babies born with cleft lip/palate will have other associated anomalies
 - Fetal ECHO
 - Some parents may choose to have genetic testing

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Bents, Buey, Zuber. Pediatric Plastic Surgery, 2nd Edition.

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Cleft Lip Embryology

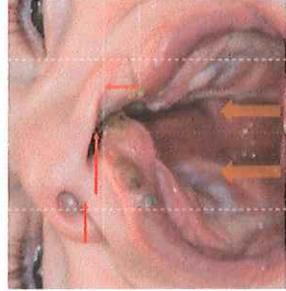
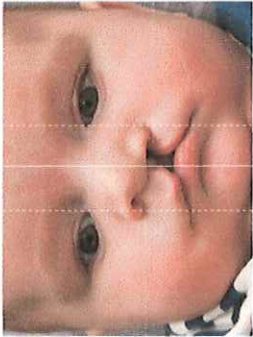
- frontonasal prominence
- maxillary prominence
- lateral nasal prominence
- mandibular prominence

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Nollman, Ed., Plastic Surgery, 2018.

Cleft Lip



- Congenital deformity of the lip and often nose secondary to abnormal development
- Missing and abnormal tissue
 - Shortened philtrum
 - Abnormal orbicularis oris muscle
- Abnormal vermillion
- Nasal deformity
 - Short columella
 - Cartilage collapse
 - Alar base malposition
- Bony deformity (complete clefts)



Cleft Lip Classification



Cleft Lip Classification

Presurgical Orthopedics

- Taping
- Naso-alveolar molding (NAM)
 - Custom appliance
 - Used to mold the protruding premaxillary segment and alveolar processes into more favorable position
 - Requires frequent adjustments by skilled orthodontist in order to properly mold alveolus and nares

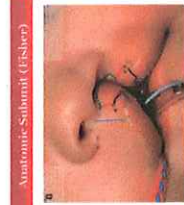


Surgical Goals

- **Lip:** normalize length, place scar along natural contours
- **Orbicularis:** re-establish continuity across cleft
- **Alar base:** raise, project
- **Nasal tip:** narrow and elevate stumped lower lateral cartilage
- **Septum:** Centralize



Types of Cleft Lip Repair



Unilateral Incomplete Cleft Lip



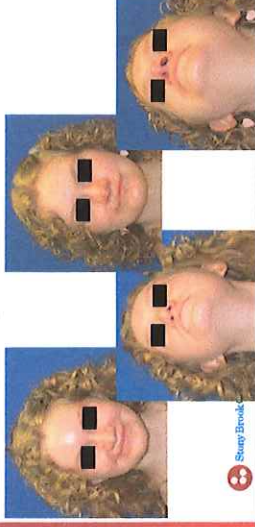
Unilateral Complete Cleft Lip



Bilateral Complete Cleft Lip



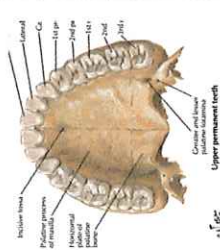
Cleft Lip Rhinoplasty



Cleft Palate

Cleft of the Primary Palate

- Anterior to incisive foramen
- Complete
 - Cleft of lip, alveolus, palate to incisive foramen
- Incomplete
 - Notch



Cleft Palate

Cleft of the Secondary Palate

- Incomplete bifid uvula to posterior to incisive foramen
- Complete from uvula to incisive foramen
- 15% associated with other syndromes/anomalies

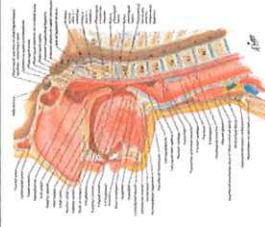


Cleft Palate Embryology



Palate Function

- Acts as a sphincter to separate the nasopharynx from the oropharynx during speech and eating
- Critical in production of speech
- Palate can be negatively impacted by either anatomic or functional causes
 - Cleft, scarring, velar dysplasia
 - Motor tone



Van der Woude syndrome

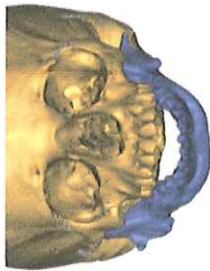


- Autosomal Dominant
- Associated with 2% of cleft lip cases
- Associated with congenital lip pits
- Mutation in the interferon regulatory factor 6 (IRF6) gene

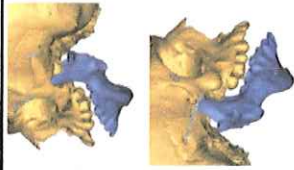
Treacher Collins



Treacher Collins



Sharp Break Children's



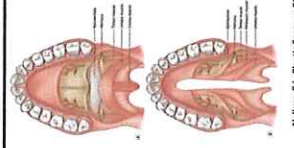
Associated Syndromes

- Velocardiofacial (DiGeorge) Syndrome
- Deletion of 22q11.2
- Congenital heart disease
- Hypoplastic thymus or athymic
 - Immune issue
 - Hypocalcaemia
- Speech issues
- Trisomy 13 (Patau Syndrome)
- Trisomy 18 (Edward's Syndrome)

Sharp Break Children's

Cleft Palate Anatomy

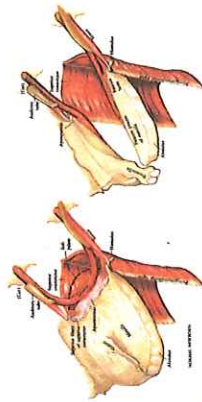
- Disruption of levator sling, central insertions of palatal aponeurosis
- Muscles instead insert along posterior edge of hard palate
- Sphincter function compromise
- Velopharyngeal insufficiency
- Problems with speech development
- Inadequate Eustachian tube drainage
- Chronic otitis media
- Reflux from oral to nasal cavity



Sharp Break Children's

Neilgan Ed., Plastic Surgery, 2018

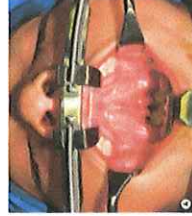
Functional Anatomy



Sharp Break Children's

Submucous Cleft

- Incidence 2-8/10,000
- Failure of fusion of palatal musculature, while overlying mucosa intact
 - Zona pellucida
- Bifid uvula, notched hard palate
- Rarely, can manifest as VPI



Sharp Break Children's

Neilgan Ed., Plastic Surgery, 2018

Velopharyngeal Insufficiency (VPI)

- Failure of complete closure of nasopharynx from oropharynx leads to air escape
- Manifests as hypernasal speech
- Children develop maladaptive speech patterns to compensate
- Early palate repair, before speech development is essential



Sharp Break Children's

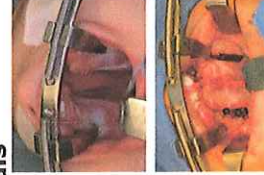
Neilgan Ed., Plastic Surgery, 2018

CLEFT PALATE REPAIR

Sharp Break Children's

Cleft Palate Repair Goals

- Achieve complete and intact closure of the palate
 - Nasal mucosa
 - Muscle
 - Oral mucosa
- Separate oral and nasal cavities
- Restore velopharyngeal function
- Normal swallow
- Normal hearing
- Optimize maxillary growth
- Prevent fistula formation



Sharp Break Children's

Timing for Palate Repair

- Controversial
- Very early repair (3-6 months)
- Early repair (6-18 months)
 - Improved speech
 - Improved hearing
 - Impaired midface growth
- Delayed repair (>24 months)
 - Impaired speech
 - Improved midface growth

Sharp Break Children's

Surgical Principles

- Early repair: increases likelihood of normal speech development
 - Theory: palate must be functional when palate related sounds are first learned to avoid poor habits
- Timing of surgery affects maxillofacial growth
 - Inherent growth retardation and maxillary deficiency because of clefting
- Impaired tensor function = reduced ventilation/drainage middle ear = Eustachian tube obstruction = recurrent OM = middle ear infections = hearing loss
 - Hearing loss is less with palate repaired earlier: 10% < 1 yr, 60% > 1 yr (Chamberlain 1976, Wilson 1986, Jansen 1979)
- Routine prophylactic insertion of tympanostomy tubes at palatal surgery

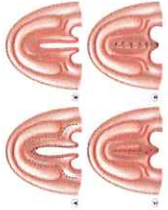


Sharp Brwolk Children's

Neilgan Ed., Plastic Surgery, 2018

Techniques: von Langenbeck

- Relaxing incisions along lateral edge of the hard palate, VZB mucosa also incised
- 2 bipedicated flaps of oral mucoperiosteum raised
 - Preserve greater palatine pedicle posteriorly and incisive pedicle anteriorly
- Close in 3 layers:
 - Nasal side: edges of cleft mucoperiosteum
 - Muscles: intra-velar veloplasty
 - Oral side: approximate bipedicated flaps in midline

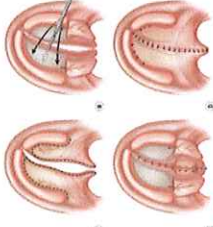


Sharp Brwolk Children's

Neilgan Ed., Plastic Surgery, 2018

Techniques: Two-Flap Palatoplasty

- Bardach variation
- Entire palatal mucosa elevated as 2 flaps, based on greater palatine pedicle posteriorly
- Careful dissection and release of pedicle
- Useful for larger clefts of the soft and hard palate



Sharp Brwolk Children's

Neilgan Ed., Plastic Surgery, 2018

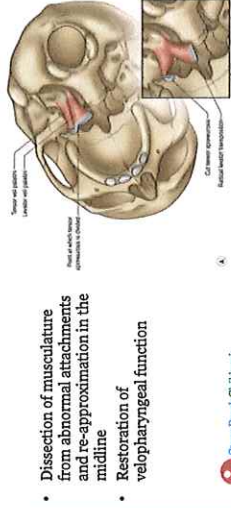
Techniques: Two-Flap Palatoplasty



Sharp Brwolk Children's

Neilgan Ed., Plastic Surgery, 2018

Techniques: Intra-velar veloplasty



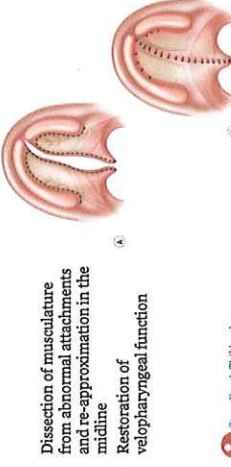
- Dissection of musculature from abnormal attachments and re-approximation in the midline
- Restoration of velopharyngeal function



Sharp Brwolk Children's

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Techniques: Intra-velar veloplasty



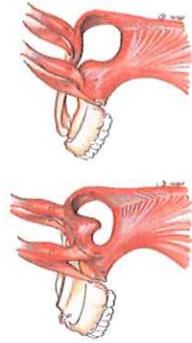
- Dissection of musculature from abnormal attachments and re-approximation in the midline
- Restoration of velopharyngeal function



Sharp Brwolk Children's

Neilgan Ed., Plastic Surgery, 2018

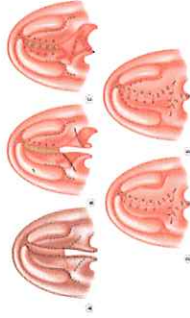
"Complete" IVVP: Total Release LVP



Sharp Brwolk Children's

Bardach, Sawyer & Bardach, Atlas, 2008

Techniques: Furlow Z-Plasty



Sharp Brwolk Children's

Neilgan Ed., Plastic Surgery, 2018

Furlow Z-Plasty

- Lengthens palate
- Prevents longitudinal scarring
- Reconstructs levator palatini sling
- Intra-velar veloplasty without dissecting muscle free
- Pharyngoplasty by lengthening palate and narrowing caliber of airway

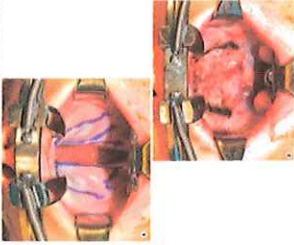


Sharp Brwolk Children's

Neilgan Ed., Plastic Surgery, 2018

Furlow Z-Plasty

- Several Studies show:
- Velopharyngeal Insufficiency (VPI)
 - 4-15% vs. 45-60% with straight line repairs
- Fistulae
 - 2-10% vs. 22-75% straight line repairs (selection bias?)



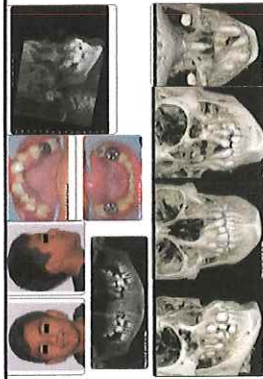
Maxillary Hypoplasia

- Maxilla is deficient
- Studies suggest growth restriction is due to surgery
- Secondary jaw surgery if necessary
- Sleep apnea possible in all patients, Pierre Robin at highest risk



Surgery During Later Childhood

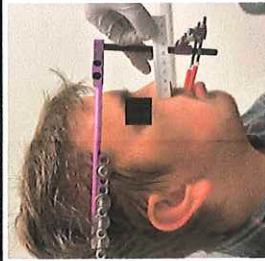
- More disruptive to daily life
- Child will have a memory of experience
- Speech surgery
- Alveolar bone grafting
- Orthognathic surgery
- Rhinoplasty



Alveolar Bone Grafting/rhBMP2



LeFort Distraction



Audiology and the Cleft Palate Child

Jamie Chung, AuD
Doctor of Audiology
Speech and Hearing Department
Stony Brook University Medical Center



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Role of Audiologist on Cleft Palate Team

- > Diagnose hearing loss (at any age)
- > Monitor hearing status
- > Refer for medical management
- > (Re)habilitation/assistive listening devices
- > Advocate for appropriate services



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Hearing loss in the cleft palate population

- Conductive hearing loss exists in 90% of children with cleft palate.
- Otitis media with effusion is the most common cause of hearing loss in the cleft palate child. This can cause a mild to moderate hearing loss in the affected ear.



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Causes of Hearing Loss in the Cleft Palate Child

- > Eustachian tube dysfunction
- > Ear malformation
- > Syndrome
- > Heredity factors



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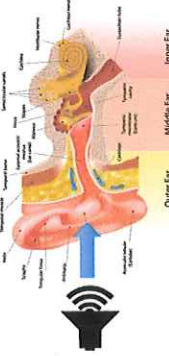
Conductive Hearing Loss (CHL)

- > Blockage/malfunction in outer or middle ear
- > Usually treatable/temporary
- > Common causes:
 - Cerumen
 - Ear malformation (otitis/otitic abnormalities)
 - Perforated eardrum
 - Fluid (otitis media)



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The acoustic pathway



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Sensorineural Hearing Loss (SNHL)

- > Inner ear/auditory nerve damage
- > Typically irreversible
 - Hearing aids
 - Cochlear implants
- > Common causes:
 - Syndromes associated with hearing loss (CHARGE, Pierre Robin, Cornelia de Langle)
 - Hereditary factors
 - OVI, Meningitis, Syphilis
 - Otitotoxic medications



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Three Types of Hearing Loss

- > Conductive
- > Sensorineural
- > Mixed



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Mixed Hearing Loss (MHL)

- > CHL & SNHL simultaneously present
 - Syndrome-related SNHL (ex. Stickler syndrome) in addition to middle ear fluid
- > Conductive portion medically treatable; Sensorineural component permanent



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"Minimal Hearing Loss"

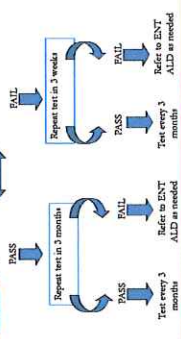
Even mild/temporary hearing loss can affect:

- Speech/language and cognitive development
- Peer relations (stress/low self-esteem)
- Behavior/attention (frustration/boredom)
- Academic achievement (37% higher failure rate in school)
- Central Auditory Processing abilities (difficulty understanding speech in background noise—life-long)

Untreated infections=tissue damage/permanent hearing loss



Newborn Hearing Screen



Thank you.



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Intermission



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ORTHODONTIC CARE OF PATIENTS WITH CLEFT LIP AND PALATE

RICHARD FABER, D.D.S., M.S.

CLINICAL PROFESSOR OF ORTHODONTICS AND DENTOFACIAL ORTHOPEDICS
 DIRECTOR OF DENTOFACIAL DEFORMITIES
 STONY BROOK UNIVERSITY MEDICAL CENTER
 PRIVATE PRACTICE MEDULLA, NY

ADA CERP
 Richard Faber, DDS, MS
 1000 Northern Blvd., Suite 1000, Great Neck, NY 11021
 Tel: 516-466-1000 Fax: 516-466-1001
 www.stonybrookchildrens.com



Dr. Richard Faber
 Dr. Zachary Faber
 Dr. Richard Faber



Role of the Orthodontist

- TIMING AND SEQUENCING OF ORTHODONTIC CARE—DETERMINED BY:
 - AGE OF PATIENT—
 - CHRONOLOGIC/SOMATIC
 - DENTAL DEVELOPMENT STAGE
 - PSYCHOSOCIAL DEVELOPMENT
 - SPEECH AND HEARING ISSUES
 - ORAL FUNCTION



Basic Four Time Periods of Treatment

- NEONATE/INFANT (BIRTH>2 yrs) Previously Discussed
- PRIMARY DENTITION (2-6 yrs) Limited to Orthopedics
- MIXED DENTITION (7-12 yrs)
- PERMANENT DENTITION (12yrs-Adult)



NEONATE/INFANT (Birth-2yrs)

- Align Cleft Segments
- Makes Lip repair easier
- Lip repair within 3-6 months
- Palatal repair at 12-24 months (speech)



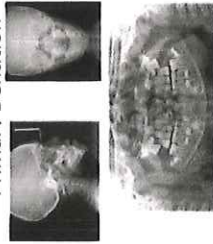
Primary Dentition (2-6yrs)

- ESTABLISH TYPE OF OCCLUSION
- ESTABLISH SKELETAL GROWTH TYPE
- CHECK FOR MISSING TEETH/SUPRANUMARY
- STAGE OF DEVELOPMENT OF TEETH
- DENTAL ARCH WIDTH AND SHAPE ISSUES
- OCCLUSAL FUNCTION AND SHIFTING BITE
- IDENTIFY FUTURE SURGICAL ISSUES

Primary Dentition



Primary Dentition



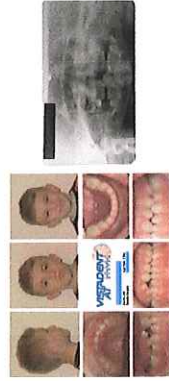
Mixed Dentition (7-12yrs)

- PALATAL EXPANSION-TRANSVERSE
- ALIGN ARCHES AND COORDINATE THEM
- BONE GRAFT FOR ERUPTING CUSPID
- DETERMINE GROWTH PATTERN
- ACCOUNT FOR MISSING/EXTRA TEETH
- ESTABLISH LONG TERM TXPL
- EVALUATE SOFT TISSUE DRAPE

Anthony's Journey 5.4



Anthony's Journey 7.3



Anthony's Journey 9.1



Anthony's Journey 9.3



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Anthony's Journey 9.8



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Anthony's Journey 10.5



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Anthony's Journey 11.2



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Permanent Dentition (12-Adult)

- ALIGN AND COORDINATE ARCHES
- LEVEL DENTITION
- SET UP FOR SURGICAL PROCEDURE
- EVALUATE GROWTH
- TIME FOR BONE GRAFT AND IMPLANT
- RETENTION—LONG TERM FOR CLEFT PT
- PROSTHETICS

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Anthony's Journey 12.3



Post Bone Graft—Cuspids Engaging

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Anthony's Journey 13.6

Cuspids Engaging and Growth Occurring CL III



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Anthony's Journey 14.2



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Along the Journey EAGLE SCOUT



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Anthony's Journey 17.8
Discussion of Orthognathic Surgery /Jawary Advancement

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Anthony's Journey –Surgical Simulation

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The Journey continues 18.4 yrs old
Waiting for the completion of growth

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Growth Finally Completed 19.11
Evaluated by Serial Cephalometric Radiographs

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Presurgical Orthodontic Appliances in Place

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Surgical Treatment Planning with VSP

VSP Orthognathic

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Precision Surgery Details

VSP Orthognathic

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Post Surgical Radiographs

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Post Surgical Occlusion

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The Journey is Over

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Life Goes On

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Nidhi 13.8

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Nidhi 13.8

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Nidhi 15.7

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Nidhi 16.6 Pontic

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Nidhi's Surgical Plan

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Nidhi 16.7 Immed PO

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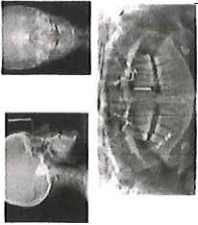
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Nidhi 18.11 Post Ortho

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Nidhi 18.7 Post Ortho



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Nidhi TX Completed



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THANKS FOR YOUR ATTENTION



IT TAKES A TEAM EFFORT

Thank you to Dr. Stephen Sachs and Dr. Sai Roggerio of the HRCO's group for the Surgery

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Hospital

Prosthetic Rehabilitation of the Cleft Patient

Tanya Somohano-Manquez DMD FACP

Let us know how your feedback improves our children with any questions, suggestions
and/or problems experienced in a previous Patient Center for the presentation.



School of Dental Medicine, Stony Brook University

Introduction

- Cleft lip and palate is one of the most common congenital anomalies
- Treatment will be dictated by its severity and usually not completed until the patient's second decade.
- Multidisciplinary management among the various specialties is essential to achieving optimal results.

Role of the Prosthodontist

- It has changed significantly over the years.
- Definitive prosthetics are usually one of the final therapies and they attempt to reduce any remaining anatomical or functional deficiencies.
- In the past, bulky removable prostheses were often necessary to replace missing teeth and correct vertical/horizontal discrepancies.
- Fortunately in recent years these have decreased with more effective surgical and orthodontic treatments in the multidisciplinary team settings.

Role of the Prosthodontist

- Nasoalveolar Molding, Feeding aids
- Interim Obturators or Speech Aids
- Management of Edentulous Spaces
- Treatment planning restorations for missing teeth
- Unrepaired Cleft Palates

Nasoalveolar Molding & Feeding Aids/Obturators

Nasoalveolar Molding

- Reduce the size of the intraoral alveolar cleft.
- Mold and position the surrounding soft tissues including the nose.
- Tissue expansion-columella.
- Aim:
 - Overall improve the esthetics of the naso-labial complex
 - Have less scarring
 - Minimize the extent/number of surgical procedures

Nasolabial Molding

Grayson Method:



Unilateral Cleft Lip/Palate



Stony Brook Technique



Stony Brook Technique: Nasal Stent



Interim Obturators & Speech Aids

Role of the Obturator Prosthesis

- Most velopharyngeal discrepancies for cleft patients are managed surgically and functional aspects improved with speech therapy.
- However 2 types of cleft patients might require an obturator prosthesis:
 - Clefts confined to the secondary palate with wide posterior manibulo-mandibular arch, residual tissues and anesthetic risks.
 - Patients that exhibit hypernasality and inadequate speech following surgical therapy.

Obturator and/or Speech Aid

- Prosthesis for these patients are usually fabricated of acrylic resin with adapted wire retainers.
- Some problems include:
 - Retention (no undercut, consider ortho brackets)
 - Access to area



Feeding Aids/Obturators



Leckli, P. J., & B. L. Schmalz & R. J. Schmalz, Schmalz, Schmalz, & Schmalz, J. O. J. & Schmalz, Schmalz & Schmalz, A. O. J. (2019). Fabrication of Feeding Plate in Cleft Palate Patient: A Case Report. Dental Journal of Advance Studies, 07, 10.1055/1-0039-1084196.


Grayson Method

- Early intervention: <1 week.
- Labor intensive: weekly appointments for 12-24 weeks
- Unilateral: 3 months then lip repair.
- Bilateral: 5-6 months, then lip repair.

Management of Edentulous Spaces


Management of Edentulous Spaces

- Removable Partial Dentures




Management of Edentulous Spaces

- Resin Bonded Bridges



Management of Edentulous Spaces


- Essex Appliances
- Retainers with Pontics



Treatment Planning Restorations

Restoration of Missing Dentition

- Dental discrepancies include:
 - Missing lateral incisors - 40%
 - Unerupted maxillary central incisors next to cleft
 - Shortened roots of teeth in premaxilla
 - Missing premolars - 20%
 - Hypocalcified enamel present
 - Hypodontia - 50%




Implants in Growing Children

- Advisable to wait until two consecutive cephalometric films one year apart show no evidence of growth.
- Site development due to horizontal and vertical discrepancies present.

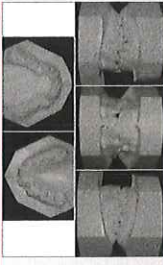


Complex Treatment Planning



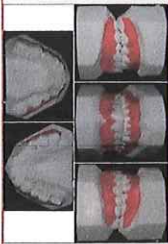
J. Craniofac. Surg. 2014; 76: 1124-1124

Complex Treatment Planning



www.dentistryjournal.com 1511-1516

Complex Treatment Planning



© 2017, Mosby, 978-0-323-35124-8

Unrepaired Cleft Palates

Unrepaired Cleft



- Adults mostly partially or completely edentulous.
- Challenges and difficulties include:
 - Active denture foundation area
 - Excessive intramaxillary space
 - Lack of bony palate
 - Scarring from lip closure
 - Presence of oro-nasal communication
 - Opposing natural dentition.

Prosthetic Considerations

- Vertical Dimension:
 - Increased in patients with small maxilla and alveolar ridge
 - Tendency to a Class III relationship; posterior teeth set in crosbite and anterior edge to edge
- Evaluator:
 - Anterior tooth display in harmony with lip thickness and scar
 - Preexisting lateral incisors could mask scar
- Records:
 - Responsibility might be an issue, consider process record bases for retentive

Prosthetic Options

- Overlay or Overdentures
 - Partial or Complete
- Implant retained and/or supported
- Tooth borne retained and/or support



Adult Cleft



Adult Cleft



Cleft Lip and Palate Symposium University Hospital at Stony Brook

Salvatore L. Ruggiero DMD, MD, FACS
Clinical Professor
Department of Oral and Maxillofacial Surgery
Stony Brook School of Dental Medicine



Thank you!

Email: Tanya.ScoroblancoMarques@stonybrookmedicine.edu

Treatment Sequence

1. Maxillary orthopedics w/ lip adhesion (4-6 weeks)
2. Definitive lip repair (rule of 10's)
3. Repair of the soft and hard palate (12-18 months)
4. Ear tubes
5. Speech therapy
6. Orthodontic therapy
7. Correction of velopharyngeal incompetence
8. Dentoalveolar cleft repair (canine eruption)
9. Orthognathic surgery
10. Nasal surgery
11. Lip revisions



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Treatment Sequence



The timing of dentoalveolar cleft repair is dependent on the canine eruption



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Goals of Alveolar Cleft Bone Grafting



- Closure of the oronasal and palatal fistula
- Stabilization of the expanded arch and the maxilla
- Provide bone support for the erupting canine
- Support the bone support and periodontal status of the incisor(s)
- Provide bone support for the air base of the nose



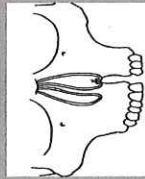
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Alveolar Cleft Grafting



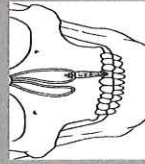
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Alveolar Cleft Grafting



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Alveolar Cleft Grafting



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Alveolar cleft grafting



- Autologous bone marrow
- Bone morphogenic protein (BMP)



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Corrective Jaw Surgery

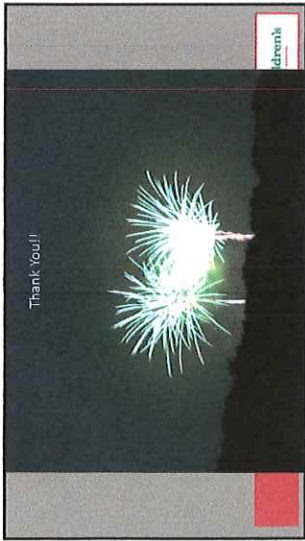


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Corrective Jaw Surgery



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**Cleft Lip and Palate Symposium:
Treatment of the Adult Cleft Patient**

University Hospital at Stony Brook

Michael Proothi, DMD, MD, FACS
Department of Oral and Maxillofacial Surgery
Stony Brook University School of Dental Medicine
1991-2020

Treatment Sequence

1. Maxillary orthopedics vs lip adhesion (4-6 weeks)
2. Definitive lip repair (rule of 10's)
3. Repair of the soft and hard palate (12-18 months)
4. Ear tubes
5. Speech therapy
6. Orthodontic therapy
7. Correction of velopharyngeal incompetence
8. Dental/occlusal cleft repair (canine eruption)
9. Orthognathic surgery

Treatment Sequence

9. Orthognathic surgery

14 Year Old Female

14 Year Old Female: Past Procedures

- Lip adhesion
- Lip repair @ 3 months
- Hard and soft palate repair @ 12 months
- Lip and nose repair @ 4 years
- Alveolar cleft graft @ 10 years

14 Year Old Female

14 Year Old Female

14 Year Old Female

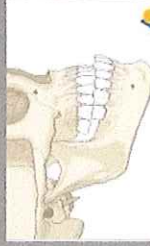
Lefort 1 Maxillary Advancement

- Horizontal advancement of the maxilla
- Correction of facial disharmony (maxillary hypoplasia/delayancy)



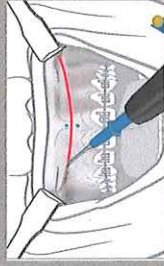
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Pediatrics • Cancer • Transplant

Lefort 1 Maxillary Advancement



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Lefort 1 Maxillary Advancement



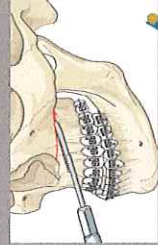
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Lefort 1 Maxillary Advancement



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Lefort 1 Maxillary Advancement



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Lefort 1 Maxillary Advancement



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1.4 Year Old Female



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14 Year Old Female



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Thank You!!

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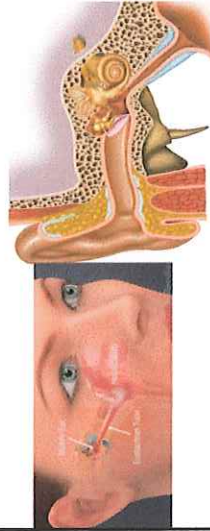
Ear Problems in Children with Cleft Palate

Huseyin Isildak, MD FACS

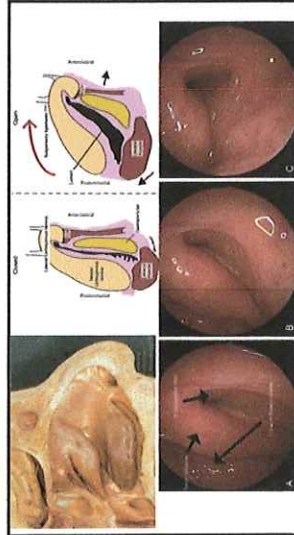
Department of Otolaryngology, Head and Neck Surgery, Stony Brook Children's Hospital

- Children with cleft palate are at a higher risk for ear problems primarily due to dysfunction of the Eustachian tube, which results from structural and muscular abnormalities associated with the cleft
- ETD is nearly universal in infants and young children with an unrepaired cleft palate.
- Majority continue to have ETD even after cleft palate repair, though some improve with age.
- ETD often leads to chronic otitis media with effusion (OME) and conductive hearing loss.

Eustachian tube



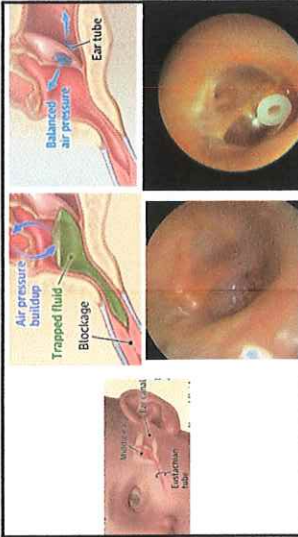
- The tensor veli palatini (TVP) muscle originates from the skull base (sphenoid bone) and inserts into the palatine aponeurosis of the soft palate.
- the levator veli palatini (LVP) muscle runs from the skull base (temporal bone) and also inserts into the soft palate.
- These muscles contract to lift the soft palate and open the Eustachian tube during swallowing and yawning.
- The TVP is the primary muscle responsible for opening the Eustachian tube, pulling it laterally.
- In individuals with cleft palate, the palatal muscles, including the levator veli palatini (LVP) and tensor veli palatini (TVP), are abnormally oriented. Instead of inserting into the soft palate, these muscles are often directed anteriorly, attaching to the posterior edge of the hard palate



- the levator veli palatini muscle → the elevation of the palate and medially rotating the torus tubarius.
- the tensor veli palatini muscle → the excursion of the anterolateral wall.

- Tubal closure depends on the adhesion of the intraluminal mucous blanket, elastic forces of the supporting tissues, and hydrostatic pressure of venous blood. The position of the Ostmann fat pad in the posterolateral half of the tube enables it to assist in restoring the tube to its closed position.
- Although under normal conditions, the middle ear maintains a slight negative pressure a few millimeters of H₂O lower than the nasopharynx, unless the tube periodically opens, the absorption of gas would result in a large negative pressure differential.
- Middle ear baroreceptors may have a greater role in eustachian tube function than mechanoreceptors
- More recent evidence, however, does suggest that mechanoreceptors in the tympanic membrane may also aid in pressure equalization

- Mucin (major component of the mucous secretions) lubricates the epithelial lining of the middle ear. Mucin secretions have been identified, particularly MUC5AC, have been found to be upregulated in otitis media.
- The middle ear mucosa and respiratory epithelium of the tube are covered with a thin layer of mucous blanket from the tympanic orifice of the tube toward the nasopharynx.
- The density of ciliated and goblet cells gradually increases in a distal and inferior direction. It has been suggested that the lower half of the tube is principally involved with clearance of the mucociliary blanket, whereas the upper half is important to pressure equilibration and gas exchange.
- Tube transport is an active process that is not gravity dependent.
- The mucociliary clearance mechanism is affected by the viscosity of the mucus.
- Conditions of hyperviscosity—such as those found in sinusitis, middle ear effusions, and cystic fibrosis—lead to reduction in beat frequency and efficiency of mucociliary clearance.
- After viral damage, ciliated cells may take 4 weeks to regenerate.



- Other the causes of ETD;
 - Laryngopharyngeal reflux
 - Immunodeficiency
 - Allergies/Samters (aspirin sensitivity, nasal polyps, asthma)
 - adenoid-like tissue in the tubal tonsil and tubal lumen
 - Nasopharyngeal mass

- Questions?
- Thanks!

Services for Cleft Palate Children & Health Insurance

- > Services are driven by medical necessity VS. reimbursement. What to do if your specialist is outside your plan? Single case agreement!
- > Know your appeal options and COB (coordination of benefits) Keep EOB's (explanation of benefits)
- > Insurance emphasis is on stock price, collecting premiums and denial or claims therefore making patients and healthcare providers jump through hoops for pre-identification, payments and denials of such. Appeal Options must be used timely.

Geoffrey O'Connell, LCSW/R



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Managed Medicaid Programs

- > Medicaid and Child Health Plus of NY slamming: forcing patients into an HMO) VS. having straight Medicaid. What is a fair hearing?
- > The need for disabled children to have straight Medicaid with no referrals for the many, specialists needed and should be requested at the Medicaid office. Parental Reimbursement !
- > IEP's, Early interventions!

Geoffrey O'Connell, LCSW/R



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Disabilities for Children

- > Children with disabilities may be entitled to SSI (supplemental security income) 1-800-772-1213
- > Patient's families must call to file a claim; they do not offer this benefit unsolicited.
- > Assess for Mental Health, school bullying and Stigma association and reaction

Geoffrey O'Connell, LCSW/R



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Insurance Issues for Cleft Lip/Palate Families



Geoffrey O'Connell, LCSW, CAC
 Licensed Clinical Social Worker
 Department of Care Management
 Stony Brook University Medical Center
 631.444.4343



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- In craniofacial anomalies:
 - The levator and tensor veli palatini muscles may be inherently weak or underdeveloped.
 - The most commonly observed abnormality is decreased action of the tensor veli palatini muscle, impairing the elevation of the anterior nasal wall.
 - There may easily be associated clefts. The levator palatini has been observed to relax more easily, affecting the stable scaffold by which the tensor muscle must contract in order to dilate the lumen.

Gatekeeper's of Our Own Health

- > We must be the gatekeepers of our own family's health and insurance.
- > Ask Ques. And take names.
- > <http://www.suffolkcountyny.gov/departments/healthservices/children.aspx>
- > NYSDFS Dept. of Financial Services Hotline
800-342-3736

Geoffrey O Connell, LCSW/R



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Thank you.



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Geoffrey O Connell, LCSW/R



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Parent Speaker
Staci Landi