Cleft Palate Craniofacial Center Symposium

April 7, 2025

Welcome!

Stony Brook Children's

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

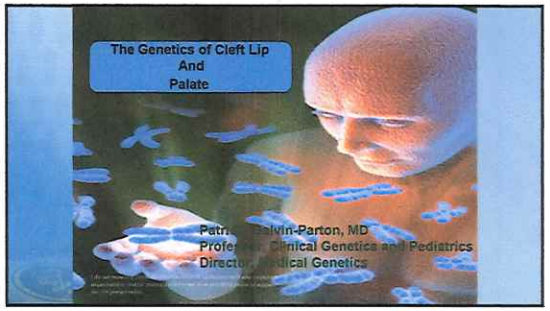
- To provide **patient centered care** for children with CP/CF deformities
- To **guide and support families** to negotiate financial and social barriers

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
- Pediatric Dentist
- Plastic Surgeon
- Otolaryngology (ENT)
- Audiology
- Speech
- Orthodontics
- Maxillofacial Surgery
- Prosthodontist
- Social Work
- Caregivers



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 600 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?

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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

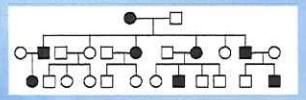
Genes

- *Single Gene Defects*
  - Dominant
  - Recessive
  - X-Linked
- *Chromosomal Defects*
  - Trisomy 13 (Patau)
  - 5p- (Cri-du-chat)
  - 4p- (Wolf-Hirschhorn

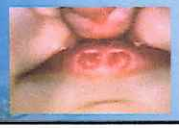
Tools

Chromosome analysis, Chromosome MicroArray, Gene Panels, Whole Exome Sequencing via blood, amniotic fluid or tissue/saliva.

Autosomal Dominant Inheritance



- Van der Woude Syndrome



- Facial Cleft (37%)
- +/- Lower Lip Pits (86%)
- +/- Hypodontia
- +/- Family History
  - Single Most Common Inherited Cause of Isolated Clefts (2-12%)
- *AD*, Incomplete penetrance & variable expressivity

Velocardiofacial Syndrome

- Velo – Cleft palate (overt or submucous), hypemasal speech
- Cardio – Contruncal 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/MR
- Ext – Long, thin fingers

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

Stickler syndrome

Cleft palate/Pierre Robin sequence

Nearsightedness, vitreoretinopathy, myopia, Amblyopia, astigmatism

Flat face, malar Hypoplasia

Hearing loss, sensorineural hearing loss

Hyperextensible joints/arthralgias

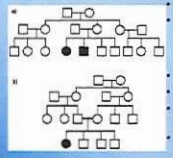
Specific findings on bone x-rays: Knock-knees, epiphyseal dysplasia

Mitral valve prolapse (46%)

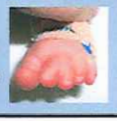
Treacher Collins Syndrome

- The child usually will show normal intelligence. Examination of the infant may reveal a variety of problems, including:
- Abnormal eye shape
- Flat cheekbones
- Clefts in the face
- Small jaw
- Low-set ears
- Abnormally formed ears
- Abnormal ear canal
- Hearing loss
- 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 TCS1 gene

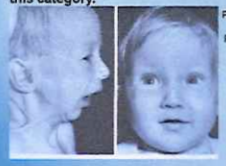
Autosomal Recessive Inheritance – Smith-Lemli-Opitz Syndrome



- Facial clefting
- Distinctive appearance
  - Upturned nose, droopy eyelids, low set ears
- Heart defects
- Male genital ambiguity
- Syndactyly of the 2nd & 3rd toes
- Inheritance: *Autosomal recessive*
- Defect in cholesterol metabolism
  - 7-dehydrocholesterol 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

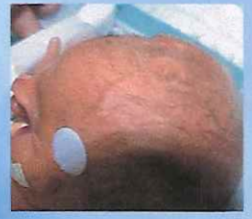
Main offending agents are

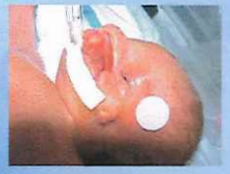
Retinoic Acid

Valproic Acid

Warfarin or Coumadin

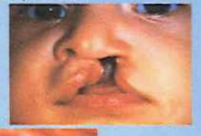
Alcohol

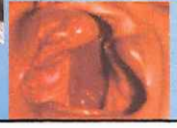




Discussion of various kinds of Prenatal Dagnosis if couple is interested

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
- Further 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

"What matters most is how you see yourself."

Feeding infants with cleft lip and palate

Kerri Elorriaga, TSSLD
M.S., CCC-SLP Stony Brook Medicine
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 breastfeeding 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 loose it"

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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

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Thank you.

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Speech Pathology Intervention with Cleft Lip/Palate

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

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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 errors and maladaptive/compensatory errors (related to CP)
- Speech Resonance Evaluation ~4 yrs.
  - Focus on resonance (oral vs nasal airflow)
- Videonasoendoscopy- as needed

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Speech Resonance Evaluation

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

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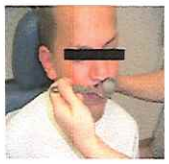
Floxite Mirror Test



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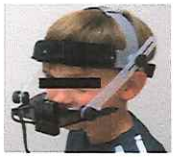
Computerized Nasometry



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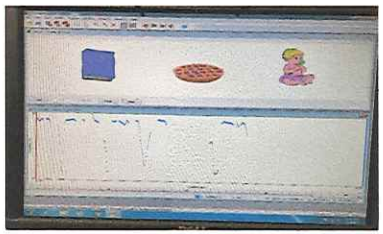

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Videonasoendoscopy

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

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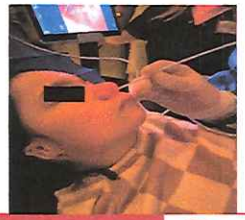
Videonasoendoscopy



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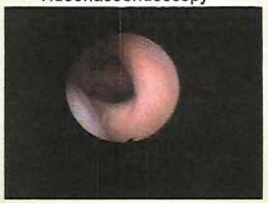
Videonasoendoscopy



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 K. Patterson DDS MS

Stony Brook School of Dental Medicine

Teamwork...

- Pediatrician
- Plastic Surgeon
- Feeding Specialist
- OMFS
- Orthodontist
- Pediatric Dentist
- ENT
- Audiologist
- Speech Pathologist
- Prosthodontist
- Psychiatrist
- Geneticist
- Nurse Coordinator
- Social Worker

Patient and FAMILY

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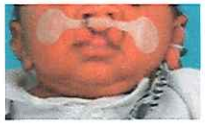
Neonatal Orthopedics - taping

Unilateral cleft lip and palate



Bilateral cleft lip and palate

Nowak p. 82

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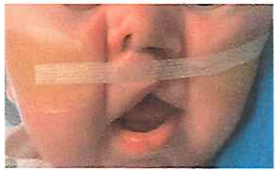
Neonatal Orthopedics - taping



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

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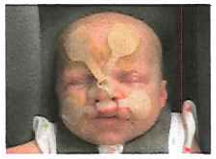
Neonatal Orthopedics - taping

Unilateral cleft lip and palate



Bilateral cleft lip and palate

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Neonatal Orthopedics

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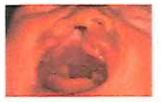
Perspective on Size of Appliance



Dean Fig 24.14

Courtesy Dr. Adewumi

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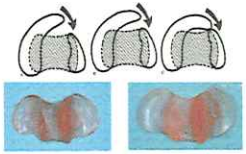
Neonatal Orthopedics

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 molding in an infant with a 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

Dean Fig 24.18

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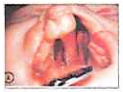
Neonatal Orthopedics



Relief areas to encourage directional growth

Presurgical or moulding plate feeding plate

NAM appliance

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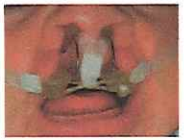
Neonatal Orthopedics



NAM appliance

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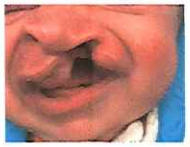
Neonatal Orthopedics



Presurgical or molding plate / feeding plate

NAM appliance

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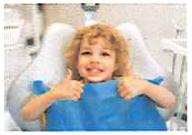
Pediatric Dentist's Role:

Role in the Team:
Coordinating with team members
Provision of ongoing dental care
Preventive treatments
Restorative procedures
Interceptive orthodontic therapy as requested



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Pediatric Dentist's Role:

Pre-Operative Role	Post-Operative Role
Parental counselling for diet and oral hygiene maintenance	Post-operative oral hygiene maintenance through professional oral hygiene maintenance aids
Early preventive advice for the child for caries prevention	Preventive care: topical fluoride and sealants application
Construction of feeding plate	Restorative care for the carious teeth and endodontic treatment for involved teeth
Presurgical orthopedics for correction of rotated premaxilla	Orthodontic correction of misaligned teeth
Instill positive attitude towards the dental treatment in a child by behaviour shaping or modification as required	Palatal plate for correction of speech problems

Luzzi et al. Table 1

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Oral Hygiene

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

Dean Table 24.3

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Oral Hygiene

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

Dean Table 24.3

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Oral Hygiene

Swabs ("Toothette") or Fingertip Brush acclimate infant to having oral tissues manipulated

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Oral Hygiene

developed at University of Porto -

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

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Oral Hygiene

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

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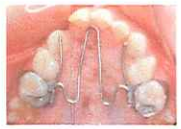
Oral Hygiene

With orthodontic appliances...

end-tufted toothbrush



Proxy brushes

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Oral Hygiene

Without orthodontic appliances...

Proxy brushes

Three-sided toothbrush

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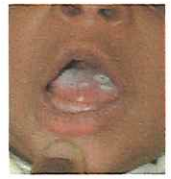
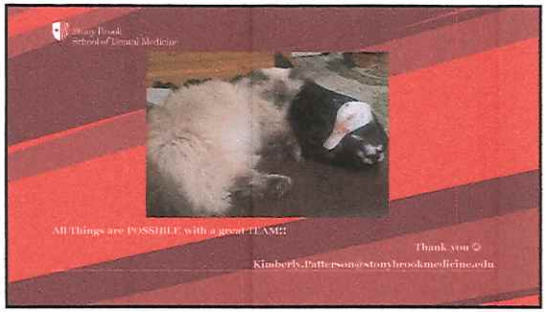
Infections

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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Cleft Lip and Palate

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

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I do not have any financial arrangements or affiliations with any corporate organizations and/or products mentioned in or providing financial support for this presentation.

Cleft Care is Team Care

- Stony Brook Cleft Palate-Craniofacial Center
  - 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
- Multidisciplinary Team Approach
  - Genetics
  - Feeding/Lactation
  - Speech
  - Audiology
  - Otolaryngology
  - Dentistry
  - Orthodontics
  - Prosthodontists
  - Oral Surgery
  - Social Work
  - Plastic Surgery
  - **Parents**

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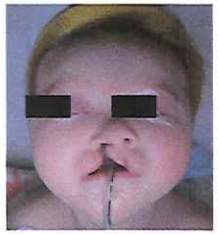
Cleft Lip and Palate

- Incidence: 1/700 births worldwide
- Can cause difficulty with:
  - Eating
  - Breathing
  - Hearing
  - Speaking
  - Social integration
  - Thriving



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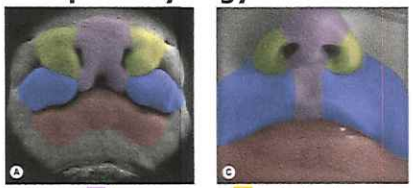
Cleft Lip Embryology



frontonasal prominence
lateral nasal prominence
maxillary prominence
mandibular prominence

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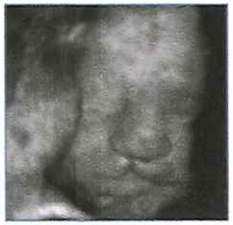
Neligan *Ed.*, Plastic Surgery, 2018

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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Bentz, Bauer, Zuker. Pediatric Plastic Surgery, 2nd Edition.

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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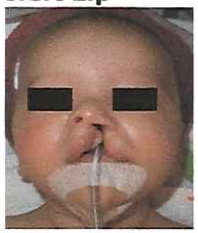
Cleft Lip



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

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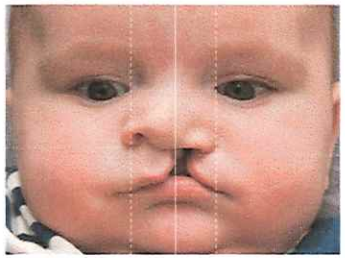

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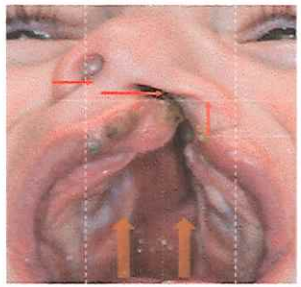
Tse et al, 2020



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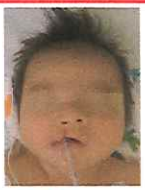
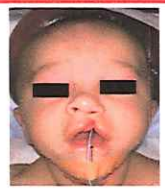
Tse et al, 2020

Cleft Lip Classification

Microform



Incomplete



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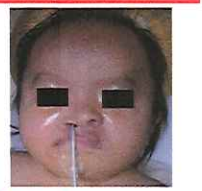
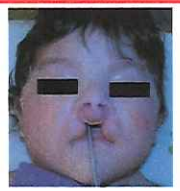
Cleft Lip Classification

Complete Unilateral



Complete Bilateral



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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

Worley et al, 2018

Losee, Joseph. Comprehensive Cleft Care, 2nd Edition.

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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 slumped lower lateral cartilage
- **Septum:** Centralize

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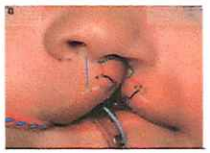
Types of Cleft Lip Repair

Millard Rotation Advancement

Anatomic Subunit (Fisher)



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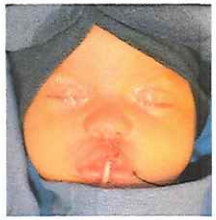
Losee, Joseph. Comprehensive Cleft Care, 2nd Edition

Unilateral Incomplete Cleft Lip



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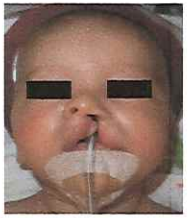
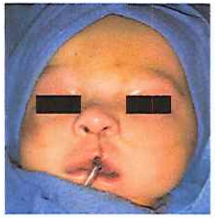
Unilateral Complete Cleft Lip





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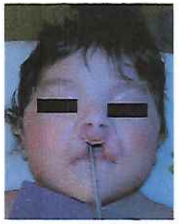
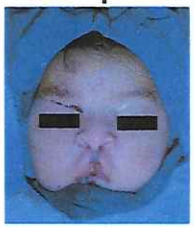
Bilateral Complete Cleft Lip





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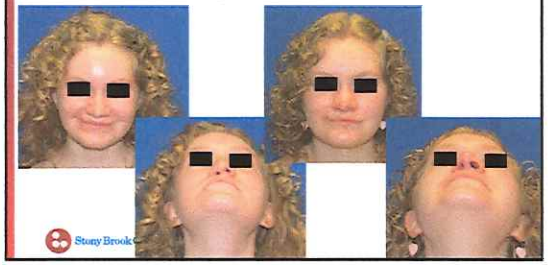
Cleft Lip Rhinoplasty



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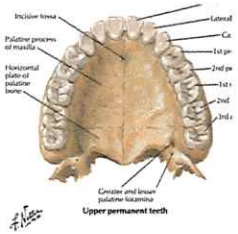
Cleft Palate

Cleft of the Primary Palate

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



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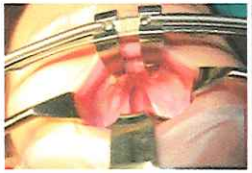
Cleft Palate

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



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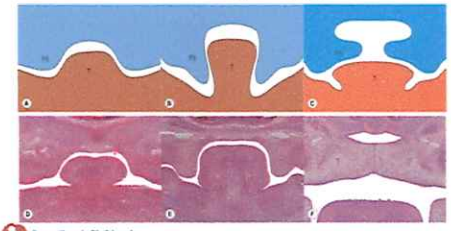
Cleft Palate Embryology



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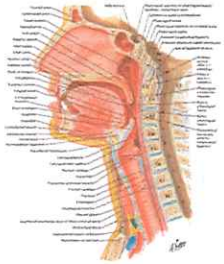
Neligan *Ed.*, Plastic Surgery, 2018

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



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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

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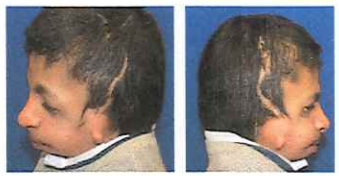
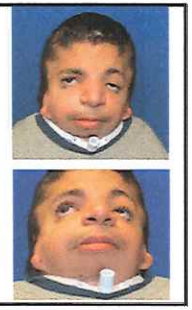
Wieprzowski et al. 2024

Treacher Collins





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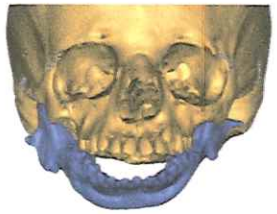

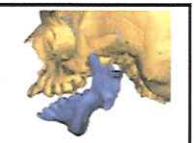

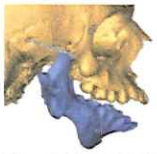

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Associated Syndromes

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

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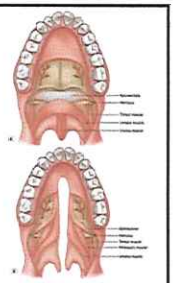
Cleft Palate Anatomy

- 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



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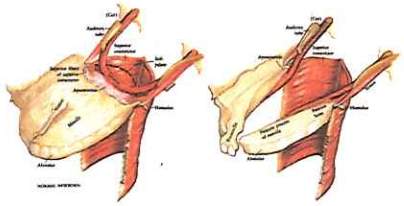
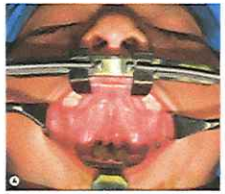
Functional Anatomy



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



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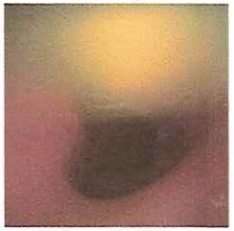
Velopharyngeal Insufficiency (VPI)

- 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



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CLEFT PALATE REPAIR

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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

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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

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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 *(Chaudhuri 1978, Watson 1986, Bennett 1972)*
  - Routine prophylactic insertion of tympanostomy tubes at palatal surgery

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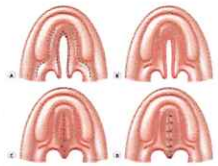
Techniques: von Langenbeck



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

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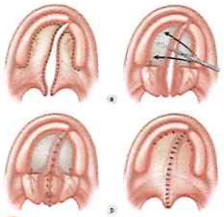
Techniques: Two-Flap Palatoplasty



- 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

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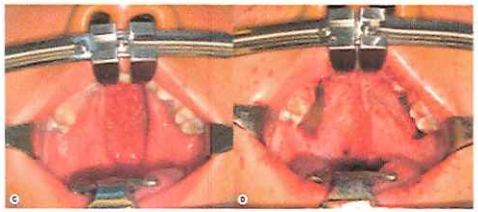
Techniques: Two-Flap Palatoplasty



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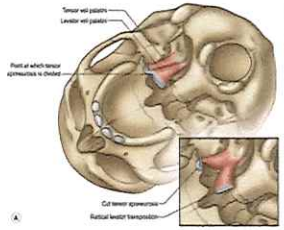
Techniques: Intra-velar veloplasty

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



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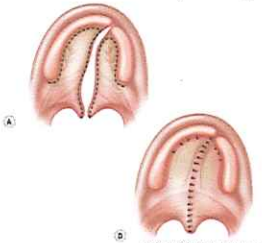

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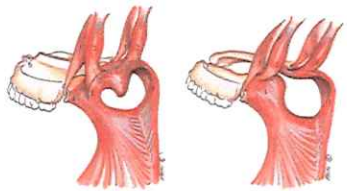
"Complete" IVVP: Total Release LVP



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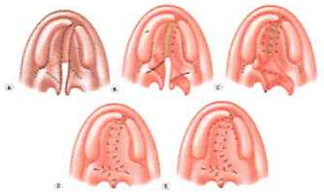
Bardach, Salyer & Bardach's Atlas, 2008

Techniques: Furlow Z-Plasty



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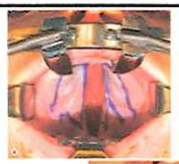
Furlow Z-Plasty

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



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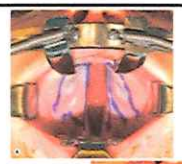
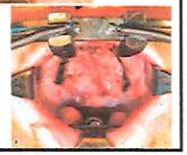
Furlow Z-Plasty

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





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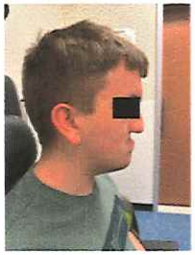
Maxillary Hypoplasia

- 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



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Surgery During Later Childhood

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

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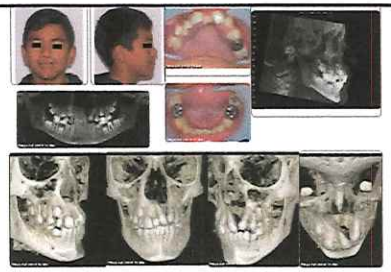

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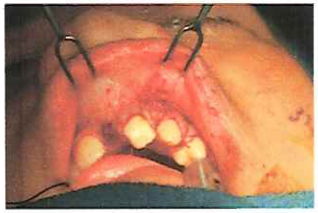
Alveolar Bone Grafting/rhBMP2



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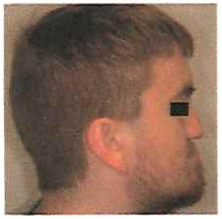
LeFort Distraction



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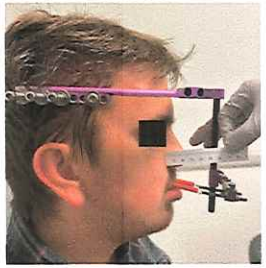

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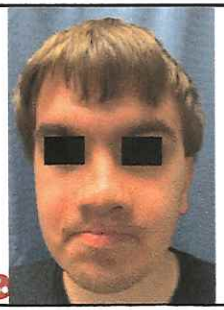

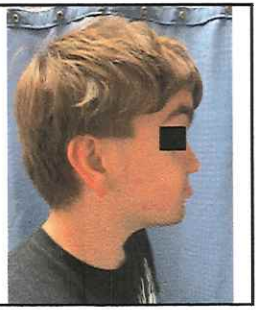

Audiology and the Cleft Palate Child

Jamie Cluna, 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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The acoustic pathway

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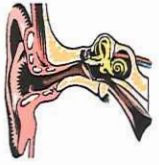
Three Types of Hearing Loss

- Conductive
- Sensorineural
- Mixed



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Conductive Hearing Loss (CHL)

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


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Sensorineural Hearing Loss (SNHL)

- Inner ear/auditory nerve damage
- Typically irreversible
  - Hearing aids
  - ALDs (FM system)
  - Cochlear implant
- Common causes:
  - Syndromes associated with hearing loss (CHARGE, Pierre Robin, Cornelia de Lange)
  - Hereditary Factors
  - CMV, Meningitis, Syphillis
  - Ototoxic medications

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Mixed Hearing Loss (MHL)

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

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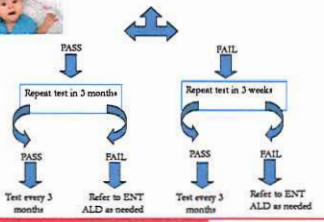
"Minimal Hearing Loss"

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



Early identification and (re)habilitation is essential in facilitating a more favorable outcome AND promoting normal development!!!

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.
CLINCAL PROFESSOR OF ORTHODONTICS AND DENTOFACIAL ORTHOPEDICS
DIRECTOR OF DENTOFACIAL DEFORMITIES
SCHOOL OF DENTAL MEDICINE
STONY BROOK UNIVERSITY MEDICAL CENTER
PRIVATE PRACTICE MELVILLE, NY

ADA CERP

Dr. Richard Faber Dr. Zackary Faber
Dr Hechang Huang

Role of the Orthodontist

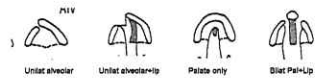
- TIMING AND SEQUENCING OF ORTHODONTIC CARE –DETERMINED BY:
  - AGE OF PATIENT–
  - CHRONOLOGIC/SOMATIC
  - DENTAL DEVELOPMENT STAGE
  - PSYCHOSOCIAL DEVELOPMENT
  - SPEECH AND HEARING ISSUES
  - ORAL FUNCTION

Basic Four Time Periods of Treatment

- NEONATE/INFANT (BIRTH>2 yrs) Previously Discussed
- PRIMARY DENTITION (2-6 yrs) Limited tx /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)



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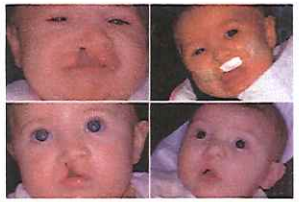

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Primary Dentition (2-6yrs)

- 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

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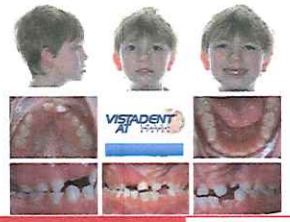
Primary Dentition



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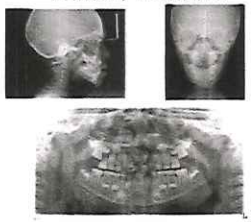
Primary Dentition



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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

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Anthony's Journey 5.4

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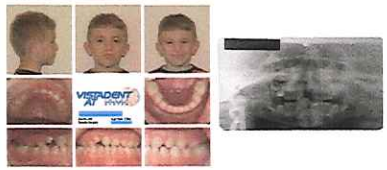
Anthony's Journey 7.3



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Anthony's Journey 9.1

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Anthony's Journey 9.3

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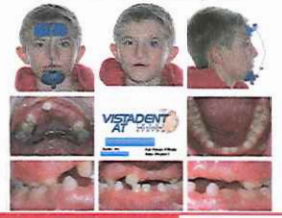
Anthony's Journey 9.8



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

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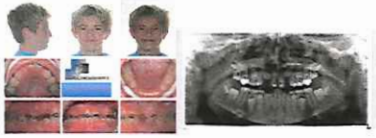
Anthony's Journey 12. 3

Post Bone Graft –Cuspids Erupting



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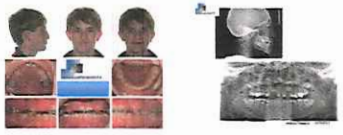
Anthony's Journey 13.6

Cuspids Erupting and Growth Occurring CL III



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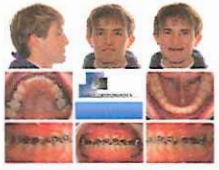
Anthony's Journey 14.2



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Along the Journey EAGLE SCOUT

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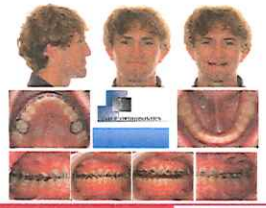
Anthony's Journey 17.8
Discussion of Orthognathic Surgery Maxillary Advancement



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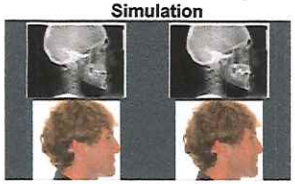
Anthony's Journey –Surgical Simulation



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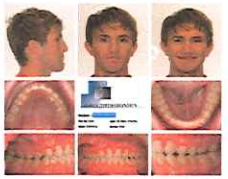
The Journey continues 18.4yrs old
Waiting for the completion of growth



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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 Orthognathics

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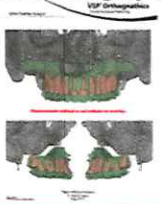
Precision Surgery Details

VSP Orthognathics



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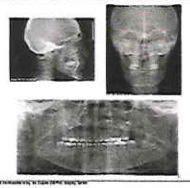
Post Surgical Radiographs



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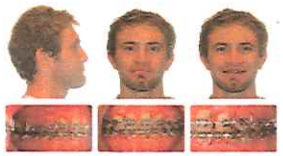
Post Surgical Occlusion



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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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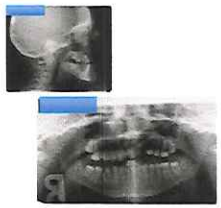
Nidhi 13.8



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Nidhi 15.7

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Nidhi 16.6 Pontic

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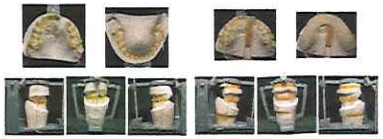
Nidhi's Surgical Plan



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Nidhi 16.7 Immed PO

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Nidhi 18.11 Post Ortho

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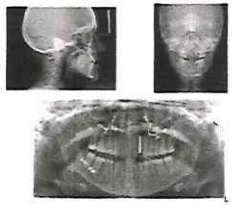
Nidhi 18.7 Post Ortho



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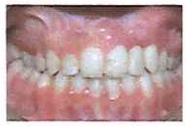
Nidhi TX Completed



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THANKS FOR YOUR ATTENTION

IT TAKES A TEAM EFFORT

Thank you to Dr Stephen Sachs and Dr Sal Ruggiero of the NYCOMS group for the Surgery

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Prosthetic Rehabilitation of the Cleft Patient

Tanya Somohano-Marquez DMD FACP

I do not have any financial arrangements or affiliations with any corporate organizations and/or products mentioned in or providing financial support for this 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

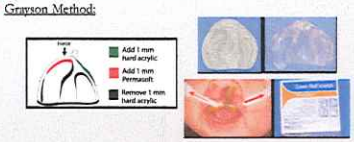
Nasoalveolar Molding & Feeding Aids/Obturators

Nasolaveolar 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 scaring
  - Minimize the extent/number of surgical procedures

Nasolaveolar Molding

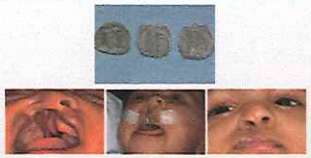
- Grayson Method:



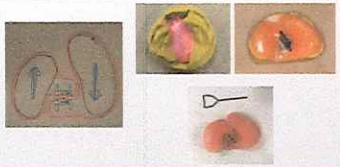
Grayson Method

- 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.

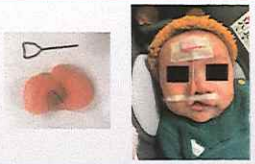
Unilateral Cleft Lip/Palate



Stony Brook Technique



Stony Brook Technique: Nasal Stent



Feeding Aids/Obturators

Lodhi, Tulsi & Patil, Surendra & Bahetwar, Surendrakumar & Sharma, Aparna & Nenavre, Nupur & Dolas, Arti. (2019). Fabrication of Feeding Plate in Cleft Palate Patient: A Case Report. Dental Journal of Advance Studies. 07. 10.1055/s-0039-1684106.

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 maxillo-mandibular width, residual tissues and anesthetic risks.
  - Patients that exhibit hypernasality and inadequate speech following surgical therapies.

Obturator and/or Speech Aid

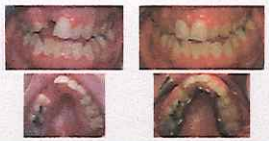
- Prosthesis for these patients are usually fabricated of acrylic resin with adapted wire retainers.

- Some problems include:
  - Retention (no undercuts, consider ortho brakets)
  - Access to area

Management of Edentulous Spaces

Management of Edentulous Spaces

- Removable Partial Dentures



Management of Edentulous Spaces

- Resin Bonded Bridges

Management of Edentulous Spaces

- Essix Appliances

- Retainers with Pontics

Treatment Planning Restorations

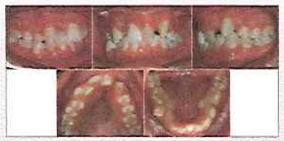
Restoration of Missing Dentition

- Dental discrepancies include:
  - Missing lateral incisors ~40%
  - Undersized 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

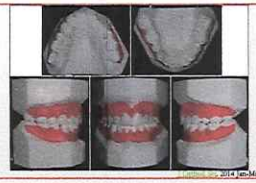


J Orthod Sci. 2014 Jan-Mar; 3(1): 17–24.

Complex Treatment Planning

J Orthod Sci. 2014 Jan-Mar; 3(1): 17–24.

Complex Treatment Planning

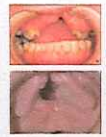


Implant Dent. 2014 Jan-Mar; 23(1): 17–24.

Unrepaired Cleft Palates

Unrepaired Cleft

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



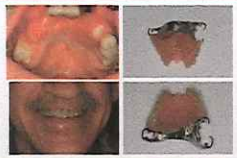
Prosthetic Considerations

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

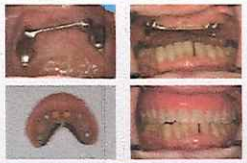
Prosthetic Options

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

Adult Cleft



Adult Cleft



Thank you!

Email: Tanya.SomehanoMarquez@stonybrookmedicine.edu

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

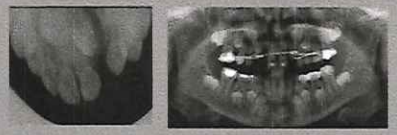
Stony Brook Children's

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. Dentoalveolar cleft repair (canine eruption)
9. Orthognathic surgery
10. Nasal surgery
11. Lip revisions

Stony Brook Children's

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 premaxilla
- Provide bone support for the erupting canine
- Improve the bone support and periodontal status of the incisor(s)
- Provide bone support for the alar base of the nose

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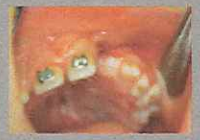
Alveolar Cleft Grafting



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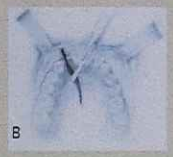
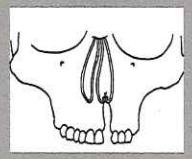
Alveolar Cleft Grafting





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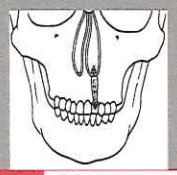
Alveolar Cleft Grafting



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Alveolar cleft grating

- Autologous bone marrow
- Bone morphogenic protein (BMP)

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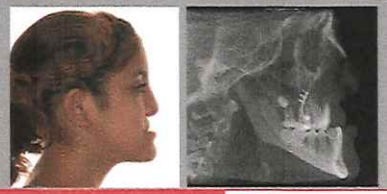
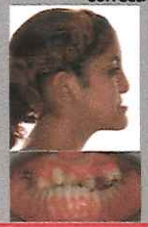
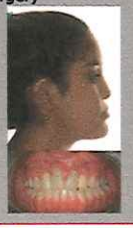
Corrective Jaw Surgery



Stony Brook Children's

Corrective Jaw Surgery





Stony Brook Children's

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

University Hospital at Stony Brook

Michael Proothi, DMD, MD, FACS
Clinical Assistant Professor
Department of Oral and Maxillofacial Surgery
Stony Brook University School of Dental Medicine
April, 2015

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. Dentoalveolar cleft repair (canine eruption)
9. Orthognathic surgery

Treatment Sequence

9. Orthognathic surgery

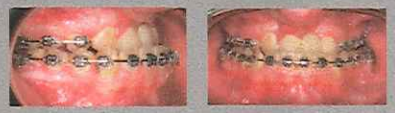
14 Year Old Female

14 Year Old Female: Past Procedures

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

14 Year Old Female

14 Year Old Female

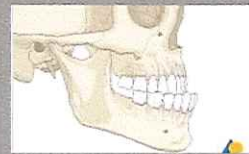


14 Year Old Female

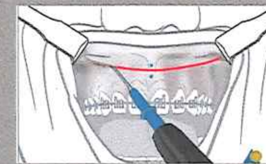
Lefort 1 Maxillary Advancement

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

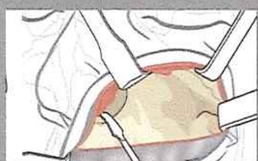
Lefort 1 Maxillary Advancement



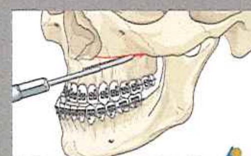
Lefort 1 Maxillary Advancement



Lefort 1 Maxillary Advancement



Lefort 1 Maxillary Advancement



Lefort 1 Maxillary Advancement

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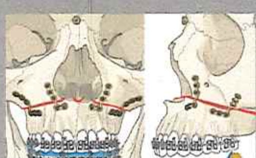

Lefort 1 Maxillary Advancement

14 Year Old Female

14 Year Old Female

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Thank You!!

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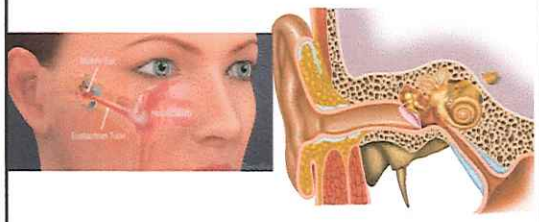
Ear Problems in Children with Cleft Palate

Huseyin Isildak, MD FACS

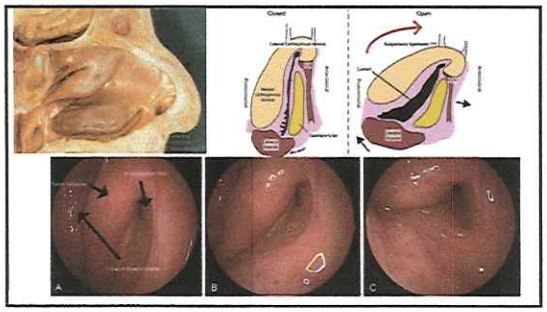
I do not have any financial arrangements or affiliations with any corporate organizations and/or products mentioned in or providing financial support for this presentation.

- 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 coordinate 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



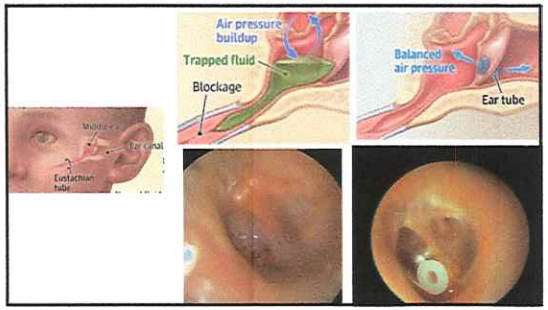
- the levator veli palatini muscle→the elevation of the palate and medially rotating the torus tubarius.
- the tensor veli palatini muscle→the excursion of the anterolateral wall.

- 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 very slight negative pressure a few millimeters of H2 O 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 surface and traps bacteria and viruses. Mucin genes (9 human mucin genes 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 cilia that transport the 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 equalization and gas exchange.
- Tube transport is an active process that is not gravity dependent
- The ***ciliary*** beat frequency in the middle ear and ET is involved in the clearance of mucus and pathogens from the middle ear and 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/Samter's (aspirin sensitivity, nasal polyps, asthma)
  - adenoid-like tissue in the tubal tonsil and tubal lumen
  - Nasopharyngeal mass

- 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 excursion of the anterolateral wall.
  - There may even be discoordination. The levator muscle has been observed to relax prematurely, eliminating the stable scaffold by which the tensor muscle must contract in order to dilate the lumen.



Questions?

- Thanks!

Insurance Issues for Cleft Lip/Palate Families

Stony Brook Children's

Geoffrey O'Connell, LCSW, CAC
Licensed Clinical Social Worker
Department of Care Management
Stony Brook University Medical Center
631 444 4343

Stony Brook Children's

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 of claims therefore making patients and healthcare providers jump through hoops for precertification, payments and denials of such. Appeal Options must be used timely.

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 Refusalment !
- IEP's, Early interventions!

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

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

Thank you.

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Parent Speaker

Staci Landi