

		
	Caring for Children with Cleft Lip and Palate: the first year of life Phayvanh Pecha, MD, MPH Associate Professor Pediatric Otolaryngology-Head & Neck Surgery, Cleft/Craniofacial Team Medical University of South Carolina	
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Disclosures

- ▶ No relevant financial disclosures

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Objectives

- ▶ Discuss interventions for cleft care in the first year of life:
 - ▶ Describe timeline of care specific to a newborn with cleft lip and palate
 - ▶ Differentiate the feeding needs of a baby with orofacial clefting
 - ▶ Managing the newborn with suspected Robin sequence



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Multidisciplinary team approach:

Standards set by the American Cleft Palate-Craniofacial Association, (ACPA) 2019

- | | |
|---------------------|-------------------------------|
| ▶ Audiologist | ▶ Otolaryngologist |
| ▶ Pediatric Dentist | ▶ Pediatrician |
| ▶ Geneticist | ▶ Cleft Surgeon |
| ▶ Neurosurgeon | ▶ Prosthodontist |
| ▶ Nurse | ▶ Psychologist |
| ▶ Ophthalmologist | ▶ Social Worker |
| ▶ Oral Surgeon | ▶ Speech-Language Pathologist |
| ▶ Orthodontist | ▶ Team Coordinator |

**1 of 3 nationally accredited
craniofacial teams in the state**

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Comprehensive Cleft and Craniofacial Team at MUSC:

Co-Directors



Krishna Patel, MD, PhD
Facial Plastic & Reconstructive Surgery



Melissa Montiel, CCC-SLP
Speech Language Pathology

Coordinator



Julia Black, LMSW
Social Work



Phayvanh Pecha, MD, MPH
Pediatric Otolaryngology



Byung Joo Lee, DDS
Maxillofacial Prosthodontics



Mark Miller, MD, DMD, FACS
Oral & Maxillofacial Surgery



Ramin Eskandari, MD
Pediatric Neurosurgery



Libby Infinger, MD, MPH
Pediatric Neurosurgery



Isis Raulino Scornaciao, MD
Plastic Surgery



Claire Hauschildt, AuD
Audiology



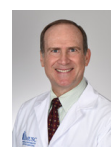
Kristen Lancaster, MD, FAAP
Pediatric Genetics



Stephanie Schank, CCC-SLP
Speech Language Pathology



Hannah Rustin DMD, MSD
Pediatric Dentistry



Timothy Tremont, DMD, MS
Orthodontics



Ildu Andrade Jr., DDS,
MS, PhD
Orthodontics

"It takes a village"

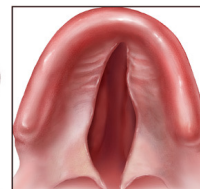
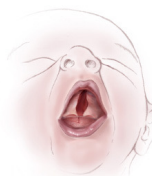


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Epidemiology of Cleft Lip and Palate

- ▶ Common congenital malformations with *variable degrees of deficient and displaced orofacial soft tissue, musculature, bone, and cartilage* involving the soft palate, hard palate and nasal sill
 - ▶ Cleft lip with or without palatal clefting
 - ▶ Cleft palate alone
- ▶ In the United States:
 - ▶ 1 in 1,600 babies is born with **cleft lip and palate**
 - ▶ 1 in 2,800 babies is born with **cleft lip** without cleft palate
 - ▶ 1 in 1,700 babies is born with **cleft palate** alone



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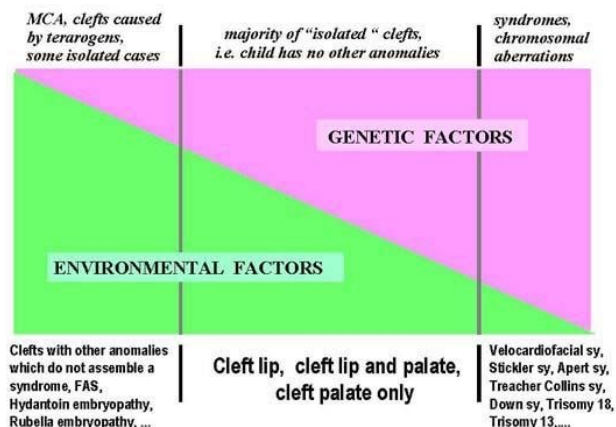
Cleft lip +/- palate		Cleft palate
1 per 1,000 newborns		1 per 1,700 newborns
Varies across ethnicities		Uniform across ethnicities
American Indian	3.6 : 1000	
Asian	1.7 : 1000	
White	1.0 : 1000	
African American	0.7 : 1000	
14% associated with syndromes		33% associated with syndromes
Male: Female = 1.5-2.0 : 1		Male: Female = 0.5-0.7 : 1

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Etiology of orofacial clefts

- ▶ Caused by interaction between genetic and environmental factors
- ▶ Significant congenital anomalies are present in up to 21% of patients overall
- ▶ Incidence of associated anomalies is dependent on type of clefting:
 - ▶ More common in cleft palate
 - ▶ Patients with bilateral clefting and midline clefting have higher risk of associated anomalies

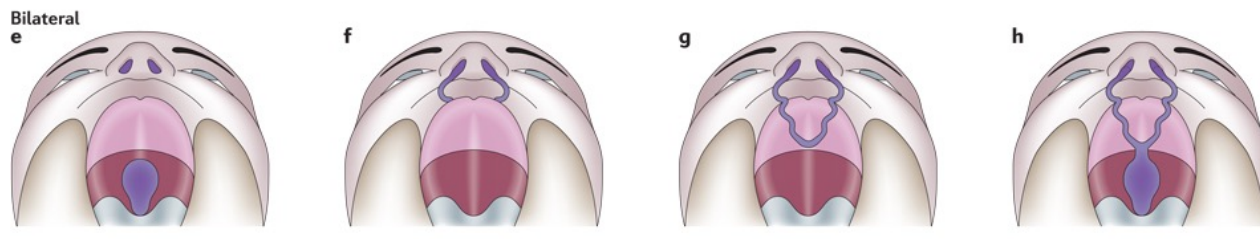
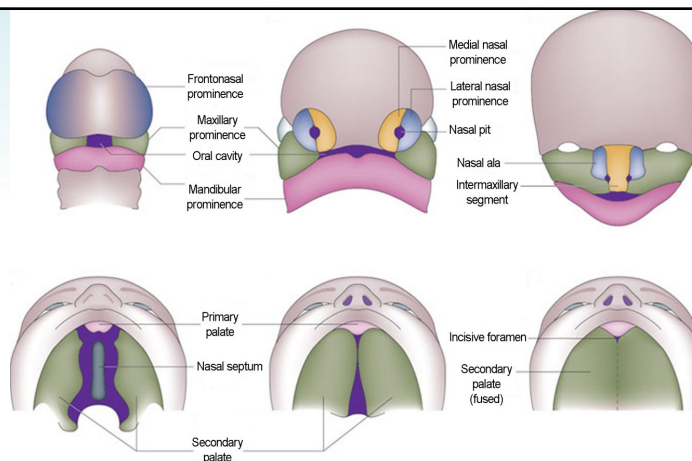
ETIOLOGY OF CLEFT LIP AND PALATE ANOMALIES



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Embryology

- ▶ 4-8 weeks gestation:
 - ▶ Lip/primary palate
- ▶ 8-12 weeks gestation:
 - ▶ Secondary palate
- ▶ 80% unilateral defects



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Multiple classification systems

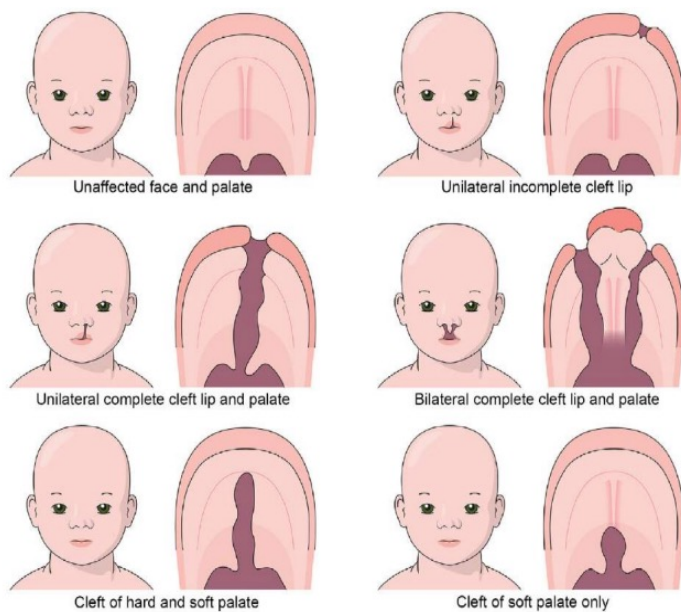
Veau Classification:

Type I: cleft of the soft palate only

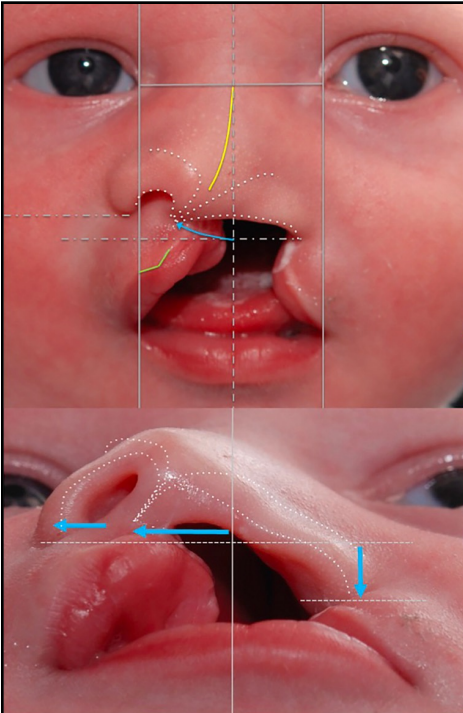
Type II: clefts of the hard and soft palate restricted to the secondary palate (palate posterior to the incisive foramen)

Type III: unilateral cleft involving secondary and primary palate (palate anterior to the incisive foramen)

Type IV: bilateral cleft with complete isolation of the median tubercle



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Typical nasal deformities associated with cleft lip include:

- ▶ Underprojection of the nose with poorly formed nasal tip
- ▶ Wide horizontal nostrils and a flat alar dome due to a lack of supporting bone structure and weak cartilage
- ▶ Lack of nasal floor on the cleft side
- ▶ Caudal septum and columella are deviated to the noncleft side in unilateral clefts

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Unilateral Cleft lips:



Microform lip

Incomplete
cleft lipComplete
cleft lip

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What are the goals of cleft care in the first year of life?



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Prenatal Counseling



- ▶ Consultation with families prenatally with craniofacial team (cleft surgeon, speech language pathologist, and coordinator)
- ▶ ~25% of cleft lips with or without cleft palate are diagnosed antenatally
- ▶ Cleft palate alone is difficult to detect on routine prenatal ultrasound

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Birth

- ▶ Goals: AIRWAY/GROWTH/FEEDING
- ▶ Intervention:
 - ▶ Airway Evaluation
 - ▶ Speech Language Pathology Evaluation → weekly weight checks
 - ▶ Genetics Evaluation

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What feeding accommodations are needed?

Cleft lip (tissue only, no involvement of alveolus):

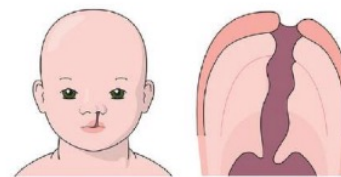
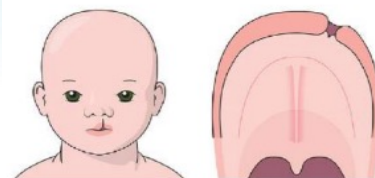
- ▶ No special accommodations

Cleft lip + alveolus:

- ▶ Possible accommodations required. Attempt mother's preferred feeding method and monitor baby closely with pre/post weights
- ▶ Consult cleft feeding specialist

Cleft palate (with or without lip):

- ▶ Special accommodations required. Will need cleft palate bottle
- Consult cleft feeding specialist
- Follow weekly weights, at risk for failure to thrive



Unilateral complete cleft lip and palate



Cleft of hard and soft palate

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Growth and feeding in babies with cleft palate

- ▶ Avoid feeding tubes in isolated cleft palate
- ▶ Cleft feeders
- ▶ Breast feeding for dessert
 - ▶ High risk of failure to thrive
 - ▶ Weekly weight checks



Mead Johnson



Habermann Nipple



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Birth – 6 months for babies with cleft lip

- ▶ Goal: Address the cleft lip

Potential Interventions based on type of cleft lip:

- ▶ Lip taping (+/- nasal hook)
- ▶ NAM (nasolabial molding) ?
 - ▶ Depends on family's ability to comply, time consuming process
- ▶ Lip adhesion ?
 - ▶ If unable to do NAM and wide cleft
- ▶ Cleft Lip Repair at 3-4 months of age
- ▶ Possible ear tubes



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Lip taping

- ▶ Recommended for babies with complete cleft lip
- ▶ Parents learn to do taping at home
- ▶ Can also use a nasal hook
- ▶ Tape until time of surgery
- ▶ Monitor for signs of skin irritation



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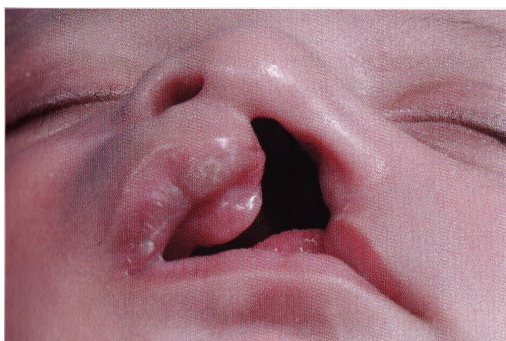
NasoAlveolar Molding (NAM) (birth - 5 months or until time of lip repair)



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NasoAlveolar Molding:

Before



After



Taping and NAM need to be ***started early (within 2 weeks of life)***, so need to be evaluated by the cleft team as soon as possible

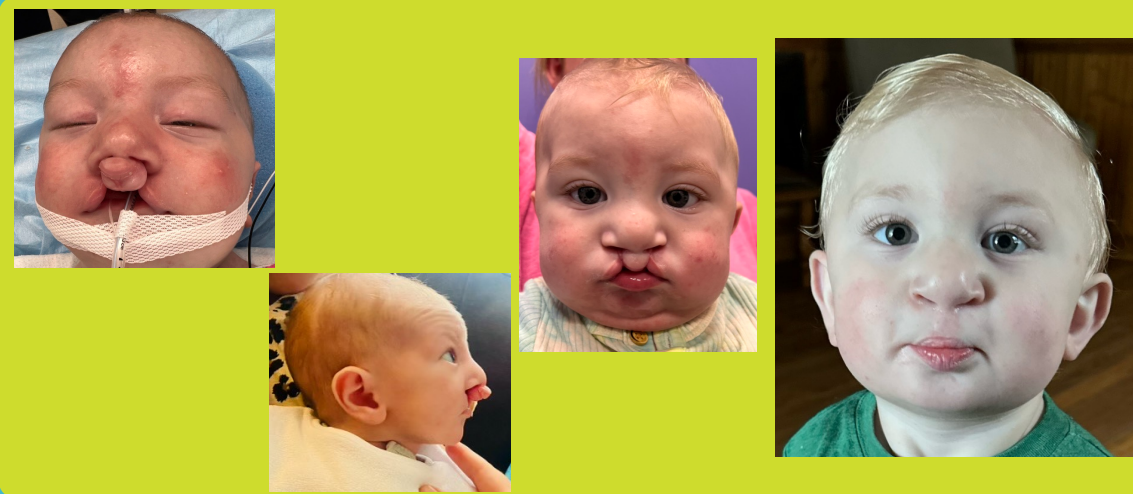
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Wide unilateral cleft lip and palate with NAM



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Lip Adhesion



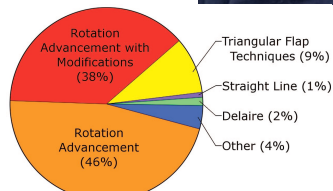
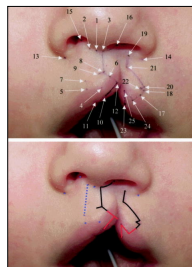
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Cleft Lip Repair/Primary Rhinoplasty (3 - 6 months)



Rule of 10's:

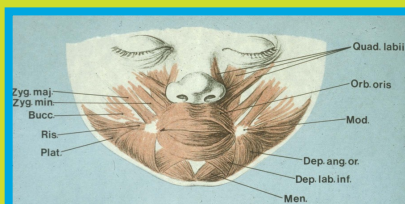
- ▶ > 10 weeks
- ▶ > 10 lbs
- ▶ > 10 Hgb



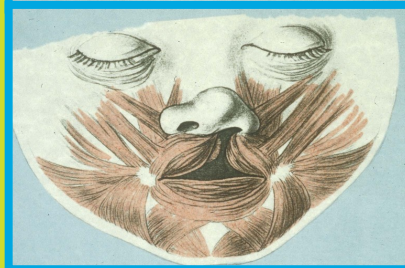
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Orbicularis Oris muscle

Normal Anatomy



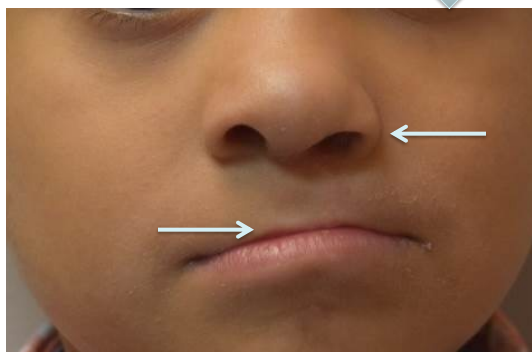
Unilateral cleft lip



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Cleft lip repair goals:

- ▶ **Function**
 - ▶ Restore orbicularis sphincter
- ▶ **Aesthetics**
 - ▶ Symmetric lip height
 - ▶ Symmetric cupid's peak
 - ▶ Continuous vermilion roll
 - ▶ Scar camouflage
 - ▶ Nasal symmetry

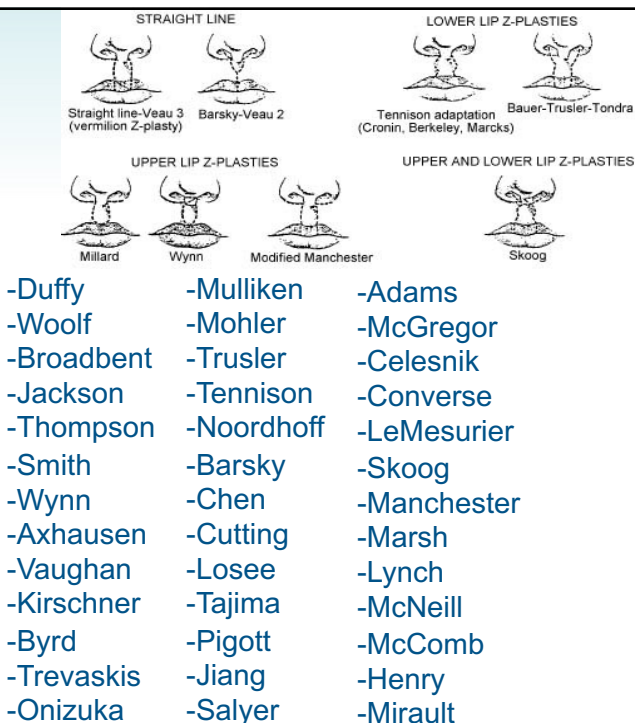


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Repairs:

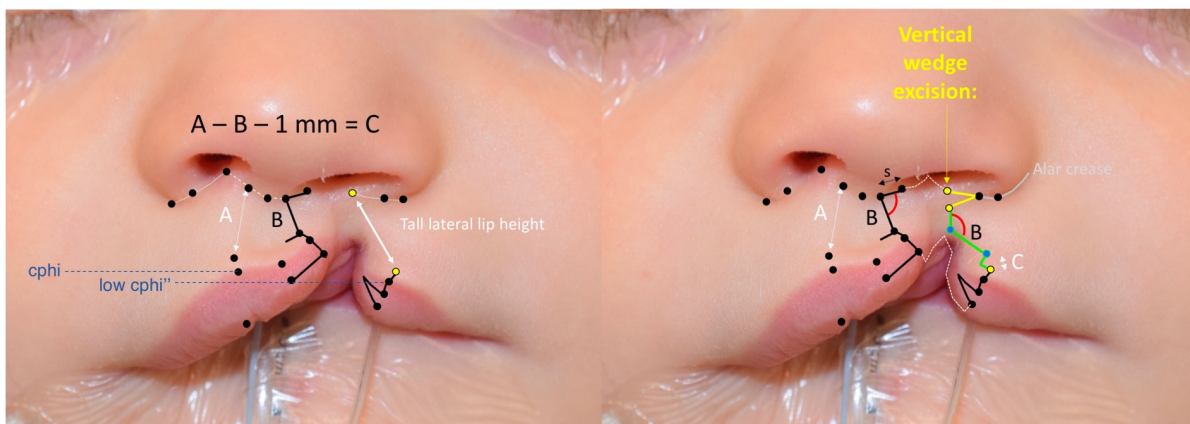
-Dupuytren
-Sims
-Rose
-Crorin
-Van Roonhunze
-Malgaigne
-Desault
-Geogiade
-Latham
-Kilner
-Potter
-Van Bardeleben
-Shultz
-Veau
-Matthews
-Monroe
-Pancoast
-Ternovsky

-Brauer
-Slaughter
-Brown
-McDowell
-Kolesov
-Lorenz
-Gabarro
-Pitanguy
-Penn
-Wardill
-Wolff
-Obukhova
-Payne
-Tondra
-Cresswell
-Randall
-Millard
-Hong

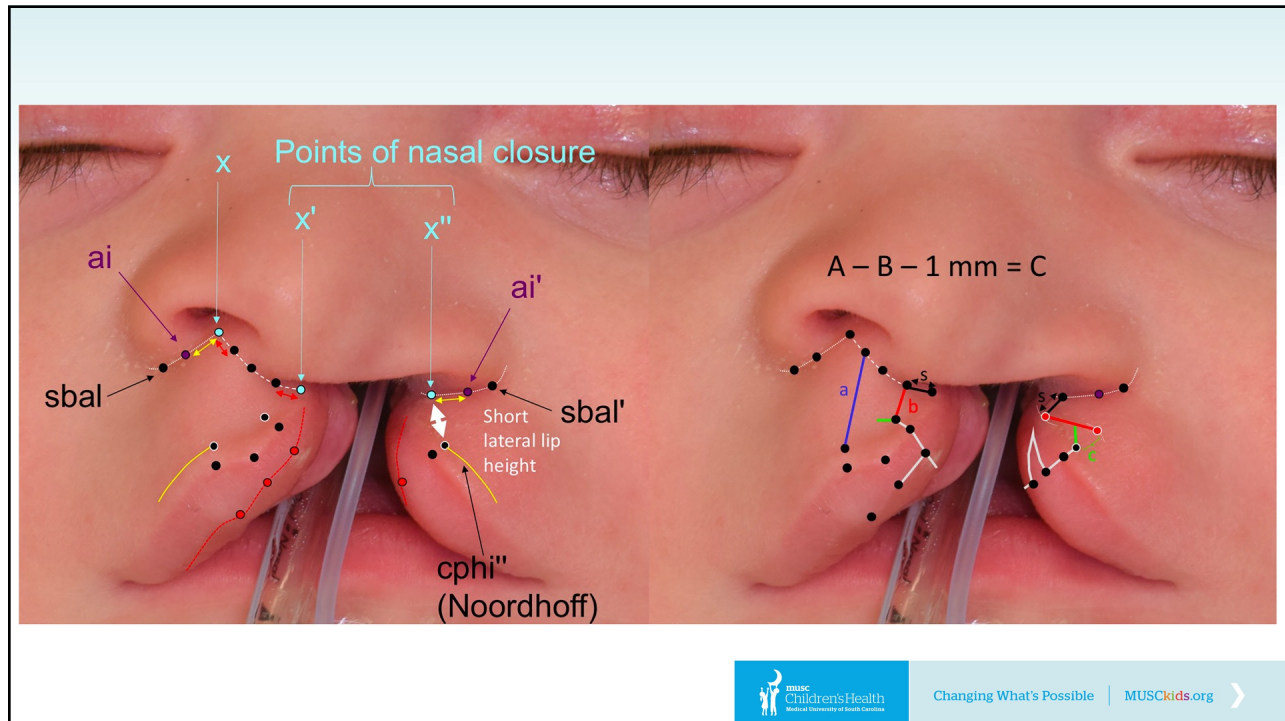


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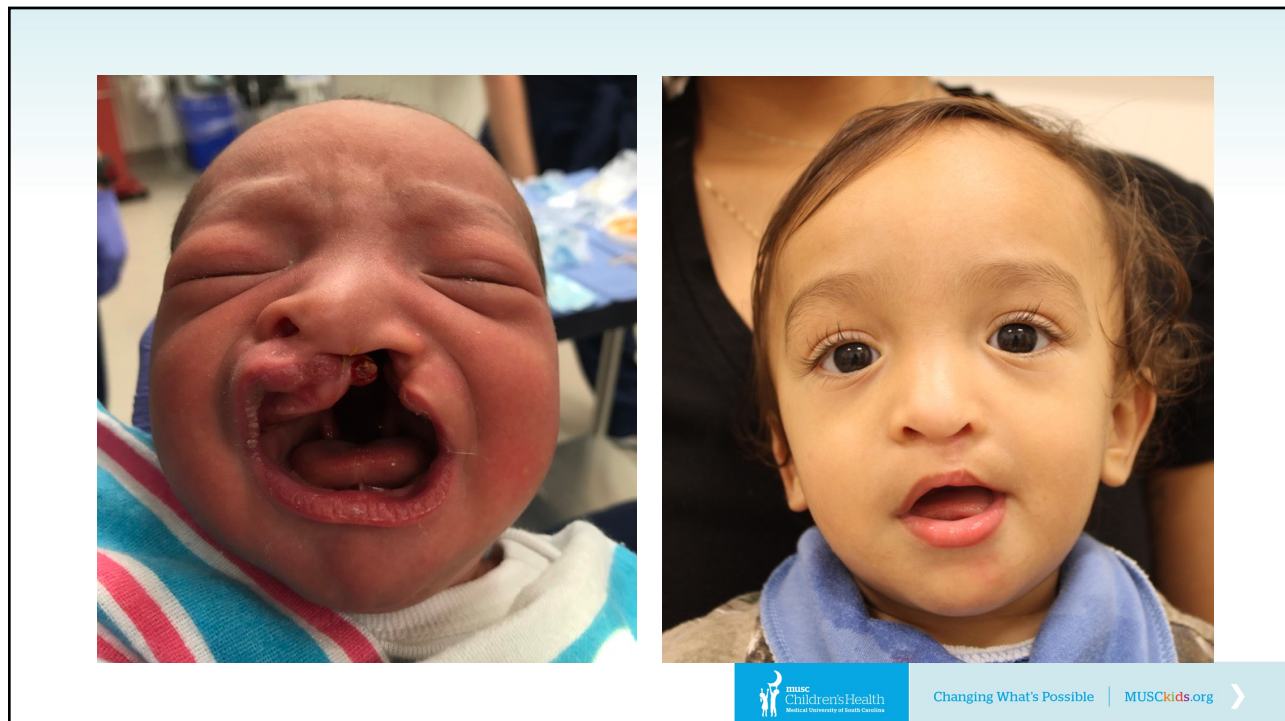
Fisher Anatomic Subunit Approximation



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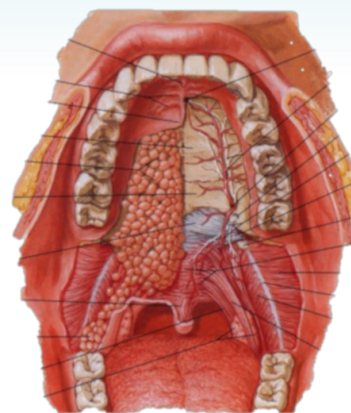
Cleft Palate Repair

- ▶ Goal: Speech and Swallowing
- ▶ Performed at 9-12 months (ACPA guidelines state by 18 months)
- ▶ Interventions:
 - ▶ Palatoplasty
 - ▶ Earlier palatoplasty - helps prevent poor speech habits
 - ▶ Later intervention - interferes less with growth of palate
 - ▶ Pressure Equalization tubes (70-90%)
 - ▶ If fluid in middle ears or conductive hearing loss
 - ▶ Often coordinate with sedated ABR

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Principles of Palatoplasty

- ▶ Minimize wound tension
- ▶ Atraumatic technique ("no touch" technique)
- ▶ Two-layer closure of oral and nasal layers
- ▶ Recreation of soft palate muscular sling



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The elusive submucous cleft palate...

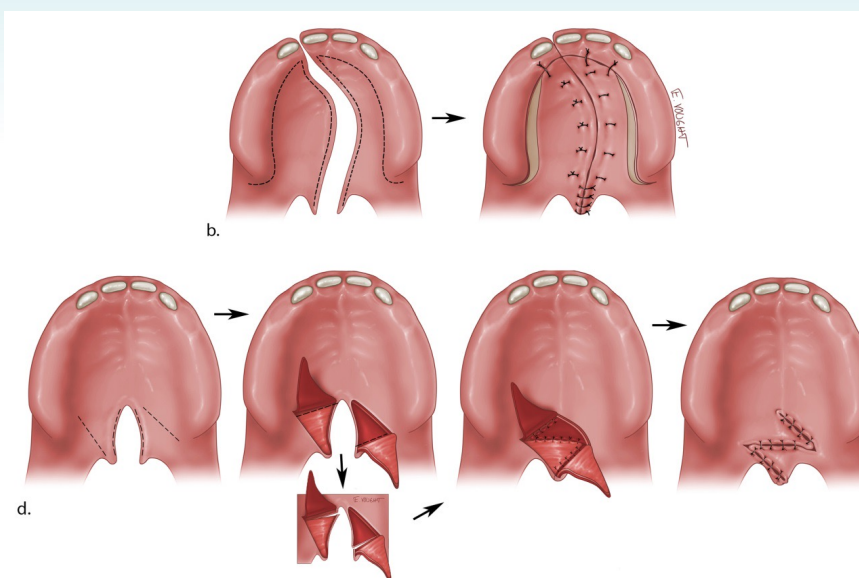
- ▶ Findings may include:
 - ▶ Bifid uvula
 - ▶ Bony notch palpable at the posterior margin of the hard palate
 - ▶ Zona pellucida: lucency or blue discoloration of the midline of the palate due to muscle diastasis
- ▶ Decision to repair made in conjunction with speech pathology (e.g., concerns for velopharyngeal insufficiency)



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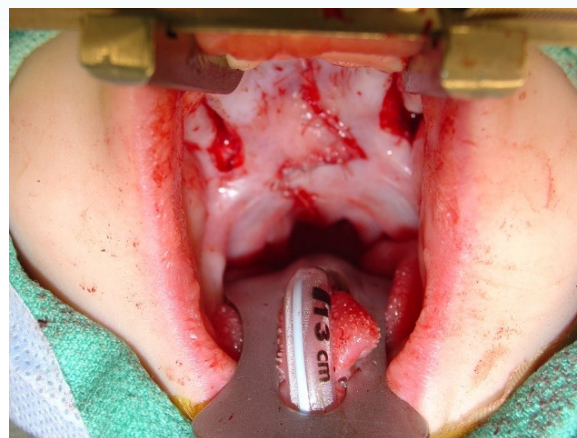
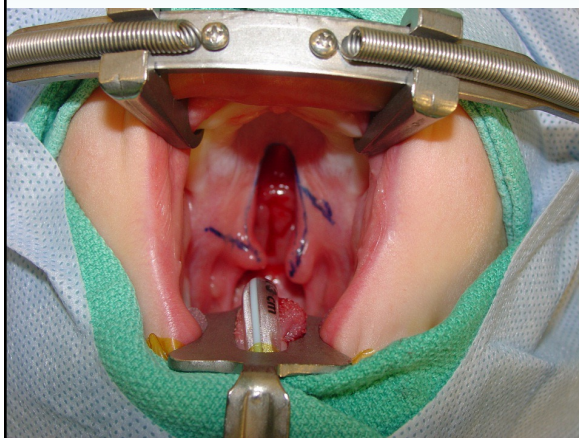
2-flap Advancement

Double opposing Z-plasty Furlow



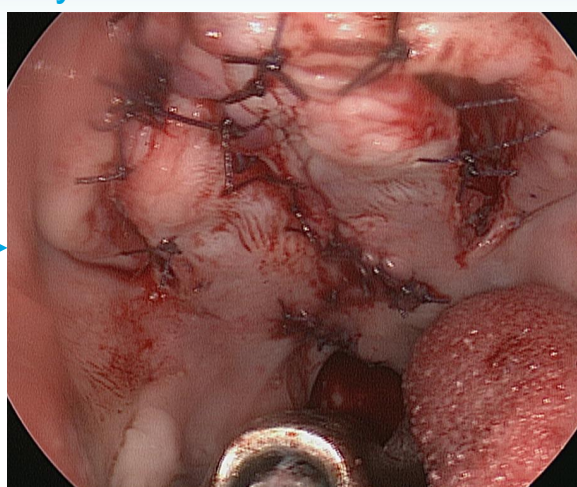
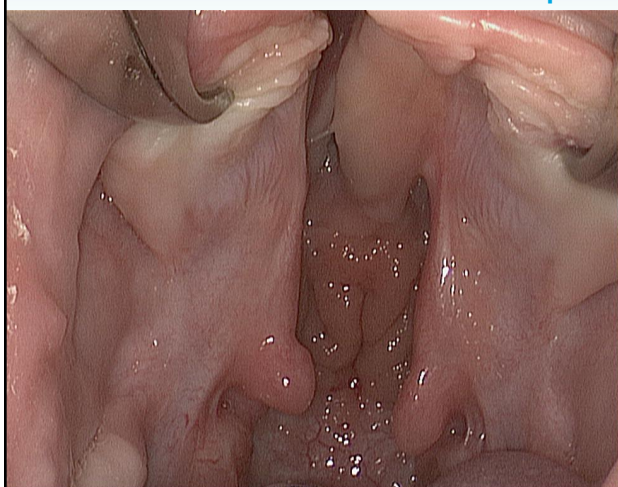
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Palatoplasty: Furlow/Double reversing Z-plasty



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Wide complete unilateral cleft palate: 6-flap modified Furlow palatoplasty



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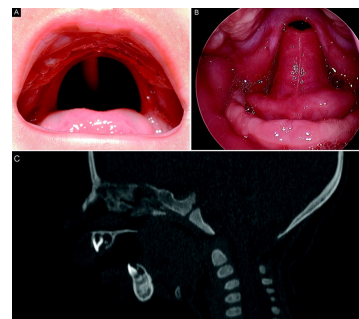
Management of Infants with Robin Sequence



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Pierre Robin Sequence

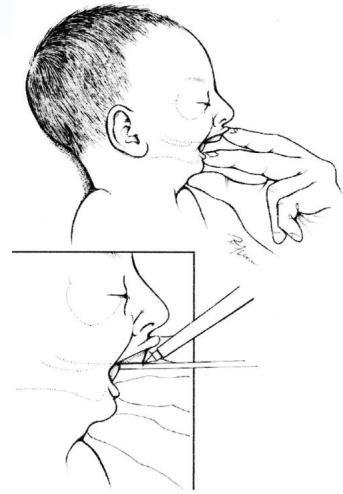
- ▶ **Incidence:** 1 in 5,000 to 1 in 14,000 births
- ▶ **Sequence:** a collection of abnormalities resulting from an initial developmental anomaly
- ▶ **Etiology:** mandibular hypoplasia before 9th week of gestation displaces tongue posteriorly and superiorly between the palatal shelves
- ▶ **Classic triad:**
 - ▶ Micrognathia
 - ▶ Glossoptosis
 - ▶ Airway obstruction
 - ▶ (Cleft palate 73-90%, typically U-shaped)



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Workup in neonate with Robin sequence

- ▶ **Airway Assessment**
 - ▶ Maxillary-mandibular discrepancy
 - ▶ Stertor, retractions
 - ▶ Oxygen saturation, **PCO₂**, **bicarbonate**
 - ▶ Fiberoptic nasopharyngoscopy to evaluate base of tongue position
 - ▶ Polysomnography? Airway evaluation (multilevel obstruction)?
- ▶ **PO intake**
- ▶ **Genetics consultation for associated syndrome**
 - ▶ 52-83% association with other abnormalities
 - ▶ Stickler and 22q11.2 deletion syndromes most common



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Parental counseling and potential complications

- ▶ **Conservative management**
 - ▶ Positioning (side-lying or prone)
 - ▶ Nasopharyngeal airway placement (monitor for alar necrosis, plugging)
 - ▶ CPAP, supplemental oxygen
- ▶ **Surgical management**
 - ▶ Tracheostomy
 - ▶ Tongue-lip adhesion
 - ▶ Mandibular distraction osteogenesis (MDO)
- ▶ **MDO directly addresses the primary problem of micrognathia and can provide long-term benefits by lengthening the mandible.**



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Mandibular Distraction Osteogenesis

Advantages:

- ▶ Definitive treatment
- ▶ Avoid tracheostomy
- ▶ Avoid g-tube
- ▶ Can provide long-term cosmetic benefits

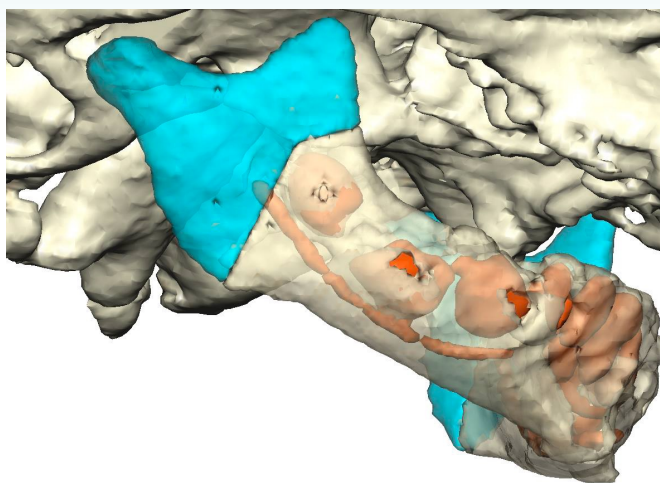
Complications:

- ▶ Damage to the inferior alveolar and facial nerves
- ▶ Damage to tooth buds
- ▶ Infection
- ▶ Failure to cure
- ▶ Device failure

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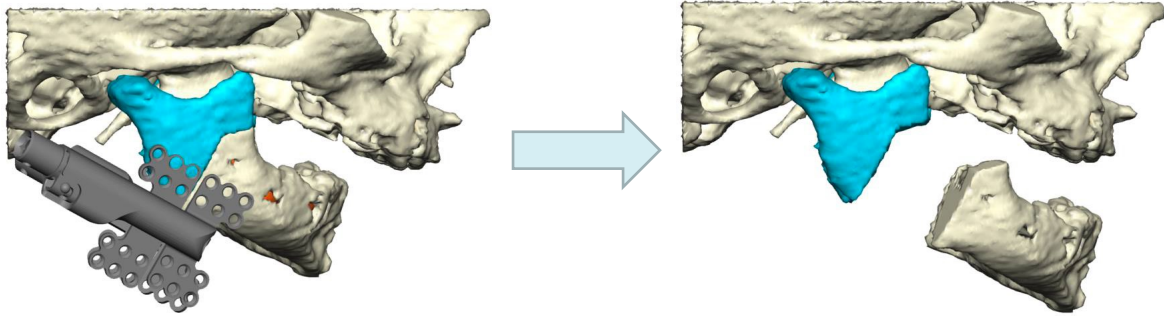
Presurgical planning and anatomy

- ▶ Type of device
 - ▶ Internal
 - ▶ External
- ▶ Companies
 - ▶ KLS, Synthes, Stryker
- ▶ Pre-surgical planning
 - ▶ Vector of distraction
 - ▶ Guides for osteotomy
 - ▶ Avoid surrounding structures



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Planned osteotomy and mandibular advancement



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Postoperative Management*

- ▶ Transported to ICU intubated
- ▶ Extubation 2-3 days post-operatively
- ▶ Distractor device is activated after observing a latency period of 2-5 days
- ▶ Start working with SLP for PO intake 1-2 days after extubation
- ▶ Rate of activation 1-2mm per day until mild class III occlusion is obtained
- ▶ Activator arms removed before discharge



After a period of 6 weeks of consolidation

- ▶ Distraction device removed in a second operation

*Protocol varies by institution and surgeon



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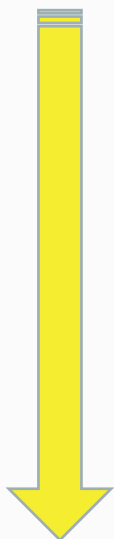
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BASIC TIMELINE OF TREATMENTS FOR CLEFT LIP AND PALATE



AGE	MEDICAL TREATMENTS:	SURGERY:
Prenatal to Birth	- Genetic counseling - Speech pathology consultation for feeding baby with a cleft	
0-5 months	- Speech pathology follows for feeding and growth - Monitor hearing - Nasoalveolar molding if needed (NAM)	- Cleft lip repair - Ear tubes if fluid in ears
9-12 months		- Cleft palate repair (Palatoplasty) - Ear tubes if fluid in ears persist
1-4 years	- Introduction to pediatric dentist - Assess language development	
4-6 years	Speech pathology assesses if surgery needed to correct speech for velopharyngeal insufficiency/(VPI)	- Speech corrective surgery - Lip revision if needed - Minor nasal surgery if needed
6-12 years	- Orthodontics (Dental appliances and braces) - Assess school/psychosocial adjustment	Alveolar bone grafting
12-21 years	Orthodontics (braces)	- Orthognathic surgery to correct facial structure and jaw - Major nasal surgery (rhinoplasty)

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Take Home Points

- ▶ Challenges in the first year of life for infants with orofacial clefting may include distortion of anatomy, ET dysfunction, breathing and feeding problems.
- ▶ Infants with cleft palate are at risk of failure to thrive and need assessment and initiation of specialized feeding strategies in the immediate neonatal period.
- ▶ Newborns with Robin sequence need to be followed closely for airway management, feeding, and potential interventions.
- ▶ Long-term interdisciplinary care is necessary with a cleft team.



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Special thanks to Dr. Krishna Patel for images



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