Caring for Kids with Clefts or Craniofacial Differences

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Overview

1. Introduction
2. Cleft lip and palate
3. Pierre Robin sequence
4. Craniosynostosis
5. Craniofacial syndromes
300 infants are born with a craniofacial anomaly in Virginia each year

CDC Vital Statistics 2006

How to provide the best care for children with special needs: (Koop, 1987)

- Share unbiased information with families
- Be sensitive to cultural differences
- Professionals must collaborate with families
- Provide emotional & financial support
- Encourage parent-to-parent support
- Care should be interdisciplinary
- Incorporate developmental needs into health care plans
- Comprehensive social, emotional, & cognitive services should be available
Key Points for Families

a continuum of care

team approach is standard of care

CHoR Team

- Pediatric Plastic Surgery
- Pediatric Neurosurgery
- Oral Surgery
- Speech Therapy
- Audiology
- Child Psychology
- Pediatric Dentistry
- Pediatric Dermatology
- Orthodontics
- Interventional Radiology
- Pediatric Orthopedics

- Pediatric Otolaryngology
- Pediatric Surgery
- Nursing
- Genetics
- Early Intervention
- Neuropsychology
- Care Coordination
Cleft Lip and Palate
Isolated Cleft Palate
Functional anatomy

- Tensor veli palatini
- Levator veli palatini

These muscles allow the nose to be closed off from the mouth and are critical for eustachian tube function.

Functional anatomy

This means children with a cleft palate

- Can not generate any suction force to draw milk out of a breast or bottle,
- Experience nasal regurgitation of food and drink,
- Have fluid in the middle ear chronically.
Caring for a child with a cleft lip and palate

Prenatal Diagnosis

- Cleft lip and palate 33.3%
- Isolated cleft lip 20.3%
- Isolated cleft palate 0.3%

Key Issues

- Support
- Education
- Feeding Techniques

Prenatal Feeding Consultation

- Demonstration of feeding techniques/bottles
- Family practice with various bottles
- Provide bottles to out of town families
- Breastfeeding review
- Provision of literature, feeding video
- Try to call when you go into labor!
Feeding Management of Newborns

Feeding the Newborn with Cleft Lip

- Usually no problems with feeding
- Breastfeeding recommended
- If bottle feeding, use wide-based nipple
Feeding the Newborn with Cleft Palate (+/- Cleft Lip)

- Difficulty generating adequate negative intraoral pressure.
- Babies frequently latch on and suck (breast or bottle) but expend an enormous amount of energy and cannot meet their nutritional needs.
- Breastfeeding may look very deceiving even to experienced nurses.
- Our goal in feeding is to minimize effort and maximize appropriate weight gain.

Feeding Evaluation

- Newborns should be evaluated on Day 1, or as soon as possible, by a feeding specialist experienced with cleft and craniofacial diagnoses to establish optimal feeding management
- Phone consultations for out of town hospital nurseries and families
- Out of town families are prioritized and seen as outpatients ASAP
What About Breastfeeding with Cleft Palate?

• Difficult to stimulate and maintain milk let-down
• Always bottle feed first
• Encourage skin to skin contact after bottle feeding
• Encourage pumping of breast milk
Feeding Consultation

- Families are provided with adequate supply of appropriate bottles/nipples
- Moms are encouraged to provide skin to skin contact after bottle feeding
- Encouraged to pump breast milk
- Feeding log if required
- CPF Feeding Video, available in English, Spanish, Mandarin
- Referrals to Feeding Clinic, EI, Care Connection as needed

Working with the Pediatrician

- Weekly monitoring of weights by pediatrician on same set of scales for at least 6-8 weeks
- Family calls in weights
- Close communication with pediatrician
  - Appropriate volumes/formula types
  - Increased caloric content/fortifying breast milk
  - Need for supplemental feedings
Pierre Robin Sequence

The CHoR protocol

Defined by:
- small jaw
- retro-displaced tongue
- breathing issues
Pierre Robin Sequence

Most children have “U” shaped cleft palate

Pierre Robin Sequence

- May or may not be related to a craniofacial syndrome
Pierre Robin Sequence

- Mandible of non-syndromic patients are likely to grow at normal rate
- Children tend to resolve breathing issues by age 2
- Syndromic children more likely to have abnormal mandibular growth

Prone Positioning
Prone Positioning

Nasopharyngeal Airway +/− Gavage Tube

Endoscopy with Tongue Retraction
Feeding the Newborn with Pierre Robin Sequence

- Size of jaw, tongue position and cleft palate may cause significant feeding problems
- Must have adequate airway for PO feeding
Feeding the Baby with Pierre Robin

- Positioning in prone or side-lying
- NG tube feeds may be necessary
- Nasal trumpet may help airway
- Apnea monitor for home use
- Close follow-up with families
Abnormal Skull Shapes

Positional Plagiocephaly

Etiology?

• Intrauterine positioning?

• “Back to sleep?”
Positional Plagiocephaly

- Sutures patent
- Most commonly see flat Parallelogram deformity, with ear forward on side of flat occiput.
- Frequent history for torticollis.

Some data suggests that conservative treatment results in same outcome.

Craniosynostosis

Single Suture Craniosynostosis:

- Alters head shape by restricting growth in a direction perpendicular to fused suture
- 1 in 2000 live births
- <20% have known genetic mutation
Single Suture Craniosynostosis: **Sagittal Suture**

Causes scaphocephaly, “keel shaped head”

2 weeks post op

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Single Suture Craniosynostosis: **Metopic Suture**

Causes trigonocephaly, “triangle shaped head”
“Minimally invasive”

1990’s - strip craniectomy, +/- endoscope.

Postoperative remodeling helmet, generally required for 10 - 12 months, used to encourage improvement in head shape.

Craniosynostosis

Advantages:
• Shorter operation (2 vs 4 hrs), shorter hospital stay (1 vs 2 nights)

Disadvantages:
• Concern for negative impact on school achievement compared to open approach
• Lack of long term results
• Extended treatment course
• Increased frequency of bony defects and irregularities reported
Craniofacial Syndromes

- Pfeiffer
- Crouzon
- Saethre-Chotzen
- Carpenter
- Muenke
- Apert
Treatment of Midfacial Hypoplasia

LeFort III Halo-distraction

- Better Maxillary Advancement.
- Better Treatment of sleep apnea.
- No increase complication rate.


Microtia Reconstruction
Hemifacial Microsomia

- 1 in 1500 incidence
- Small upper and lower jaw
- +/- Microtia
- +/- Facial weakness

One Final Thought………

Girl’s & Boy’s Night Out !!!
Thank You!!!