Cleft Lip/Palate Guideline

Objectives:

1) Be able to classify types of cleft lip/palate, understand the timeline of surgical correction
2) Be able to care for the newborn cleft baby, participate in parent education
3) Understand coexisting conditions associated with clefts

Assessment:

1) Prenatal Diagnosis
   a. Usually cleft lip can be seen by about 18-20 weeks on ultrasound
   b. 3D/4D ultrasound helpful
   c. Palatal clefts difficult to diagnose
2) Prenatal Consultation
   a. Great for families to help understand and feel better about the diagnosis
   b. Focus on Feeding issues
   c. Discuss timeline for surgeries and what to expect
   d. Review pictures of surgical results
   e. Answer all questions
3) Diagnosis after born
   a. Classification of Cleft Lip/Palate
      i. Lip
         1. Unilateral or Bilateral
         2. Complete or Incomplete
      ii. Palate
         1. Primary
            a. Alveolus to the incisive foramen
         2. Secondary
            a. Incisive Foramen posterior
            b. Complete or incomplete
         3. Submucous Cleft Palate
            a. Muscles dehiscent only
   b. Associated Syndromes
      i. Consider Genetics Consult
         1. Important for diagnosis
         2. Parental counseling
      ii. Common Syndromes
         1. Stickler’s
            a. AD-variable penetrance
            b. Pierre Robin Sequence
            c. Ocular
2. **Velocardiofacial**
   a. AD – variable penetrance
   b. Cleft palate or other palatal abnormalities
   c. Facial characteristics- small ala, squared root of nose, long philtrum
   d. Cardiac abnormalities
   e. FISH test – 22q11 deletion (DiGeorge Syndrome)

3. **Treacher-Collins**
   a. AD
   b. Microtia, Hypoplastic midface, micrognathia, downward slanting palpebral fissures

4. **Goldenhaar syndrome**
   a. Mostly sporadic
   b. Hemifacial microsomia, microtia, clefts

4) **Airway Issues**
   a. Usually less of problem in cleft lip patients – more air space
   b. Pierre Robin Sequence
      i. Micrognathia
      ii. Glossoptosis
      iii. Cleft Palate
   c. Management
      i. Positioning
         1. Side
         2. Prone
      ii. Urgent
         1. Oral airway
         2. Tongue stitch
      iii. Intubation
         1. Can be difficult
         2. ENT – intubation over Hopkins telescope
         3. Intubate through laryngeal mask
      iv. Tongue-Lip adhesion
         1. Not usually recommended due to feeding, speech and scarring problems
      v. Mandibular Distraction
         1. Prevent tracheostomy
         2. Aids in Feeding
      vi. Tracheostomy

5) **First Visit**
   a. Optimal to be seen within the first week of life – NICU, inpatient, or outpatient
   b. If not seen prenatally, discuss diagnosis, treatment plan, and timeline for repairs
   c. Emphasis on Feeding options
      i. Breastfeeding not usually effective if cleft palate present
      ii. Teach/Experiment with different bottles
         1. Pigeon – personal favorite
2. Haberman
3. Mead Johnson

iii. Feeding Issues
1. May be unable to form suction- bottles are designed to create flow without suction
2. Breastfeeding- can attempt, but need to understand anatomy and why it may not be possible. Cleft lip only- usually can.
3. Expect more nasal congestion with clefts- parents needs much reassurance
4. Feeding time needs to be 20 min or less

iv. Nasogastric tubes
1. Only if adequate trial of bottles done
2. Can temporarily allow infant to strengthen with bottle
3. If used longer than 2 month, consider g-tube

v. G-tube
1. Last resort after all else fails
2. Can be combined with Nissen if reflux an issue

vi. Reflux
1. Can be worse in cleft infants because swallow more air
   a. Need more frequent burping
   b. Head of bed up/wedge/positioning
   c. May need anti-reflux meds
   d. Nasal regurgitation is normal if palate is open- need much reassurance

vii. Every patient sees our nutritionist to discuss growth and feeding goals including time and volumes
1. higher calorie formulations, if needed, so that infant is not expending too much energy on eating
2. families are asked to call/email weekly weight checks

   d. One month follow up for weight monitoring
      i. Earlier if not gaining weight
      ii. May need admit for failure to thrive

Management:

1) Timeline for surgical repair
   a. Lip adhesion surgery- 4-8 weeks (if necessary)
   b. Cleft lip Repair – 2-3 mos
   c. Cleft Palate Repair –10-12mos
   d. Columellar Lengthening – 2-3yrs (bilateral only)
   e. Tip Rhinoplasty – 6-7 yrs
   f. Speech Surgery – 3-8 yrs (if necessary)
   g. Bone Grafting – 7-10 yrs
   h. Orthodontics – 7-teens
   i. Rhinoplasty – 14-18 yrs
   j. Orthognathic Surgery – late teens
2) Factors affecting timing of surgery
   a. Ability to withstand surgery/anesthesia
      i. Lip Surgery
         1. 3months to allow weight gain, adequate hemoglobin, lessen post anesthetic apnea
   b. Airway adequacy
      i. Palatoplasty
         1. Narrow a large breathing passage to small one
         2. Will have postoperative swelling
   c. Facial growth
      i. Alveolar Bone Graft
         1. Dissection affects growth center – stunt maxillary growth
         2. Once bone in place, need to move teeth in place to prevent resorption
      ii. Rhinoplasty
         1. Use septal cartilage grafts to reconstruct alar cartilage/tip
            a. Harvest of septum too early can effect facial growth
            b. Want full facial growth to fit nose to face

3) Associated conditions with Clefts
   a. Otitis Media
      i. Over 95% patients with cleft palate will have chronic otitis media with effusion
      ii. Placement of ventilating tubes at the time of palatoplasty
      iii. If unable to pass newborn hearing screen, place tubes at 3 month of age to get accurate assessment of CNVIII function
      iv. Placement of ventilating tubes before palatoplasty may result in excessive tube otorrhea, including formula
      v. Antibiotics not needed unless acutely infected
      vi. May need tubes until older, may be into teens
      vii. Otorrhea treated initially with drops, if not effective after 5-7 days, then refer for ear suctioning prior to oral antibiotics
      viii. Patient will follow up every 9 months for audiology and tube checks until tubes no longer needed
   b. Speech
      i. Begin speech therapy at around 12-18 months of age
      ii. Monitor pressure consonants such as P, K, and S to assess palatal function as early as 18 months
      iii. Assess for Velopharyngeal insufficiency (VPI) every 6-9 months through puberty to ascertain adequate palatal function or need for intervention
      iv. Typical timing for speech surgery including pharyngeal flap or sphincterpharyngoplasty is between 4-8 years old depending on severity
      v. BE CAREFUL, with tonsillectomy/adenoidectomy in cleft palate patients
         1. If indicated for obstructive sleep apnea or tonsillitis, should be performed by person experienced in cleft care to prevent postoperative VPI.
4) Cleft Team Care
  a. All cleft patients should be followed by an accredited cleft team (ACH has the only accredited cleft team in the state of Arkansas)
  b. Provides coordinated, streamlined patient care in this complex patient population
  c. Team members
     i. ENT - Facial Plastic surgery
     ii. Audiology
     iii. Speech
     iv. Genetics
     v. Nursing
     vi. Nutrition
     vii. Social Services
     viii. Orthodontics
     ix. Dental
     x. Periodontics
     xi. Orthognathic surgery
  d. Patients begin seeing our cleft team at the age of 6-7
     i. Begin Orthodontic treatment in prep for bone graft
     ii. Consider tip rhinoplasty in combination with lip revision if needed
  e. Patients are followed in team clinic every 1-2 years until completion of treatment in late teens/20's.

References


