Cleft Lip and Palate

Cleft lip and palate occur as a defect of palatal growth in the first trimester. Patients may present with either one or the other or both. The etiology is multifactorial and includes genetic and environmental factors. Approximately 29% of patients will have a co-existing syndrome or other congenital disease/defect. Surgical repairs usually occur in neonatal period or first few years of life. Airway management may be difficult especially if certain syndromes are present (ex. Pierre Robin, Klippel-Feil). Patients are at risk for airway obstruction post-operatively.

ANESTHETIC CONSIDERATIONS FOR PATIENT:

- Considerations of the Pediatric or Neonatal patient
- Presence of syndrome or congenital defects associated with cleft lip and palate
  - more than 300 syndromes, more common with cleft palate
  - approximately 29% will have an associated condition
  - Pierre Robin, FAS, trisomy 21, Treacher Collins, Nager, Goldenhar's, Stickler, Klippel-Feil, velocardiofacial, CHD, kidney defects, subglottic stenosis, OSA
- Possible Difficult Airway (worse with bilateral cleft)
- Risk of airway obstruction with cleft palate (rarely with cleft lip) pre-operatively and post-operatively (pharyngeal edema, upper airway narrowing from surgery, or tongue obstruction of pharynx)
- Risk of FTT due to poor feeding mechanics
  - electrolyte abnormalities and iron deficiency anemia
- Risk of recurrent aspiration due to poor feeding mechanics

ANESTHETIC CONSIDERATIONS FOR PROCEDURE:

1. Shared Airway
2. Risk of ETT dislodgement with head movement and gag manipulation
3. Risk of nasal airway occlusion post cleft lip repair (some neonates are truly obligate nasal breathers).
4. Risk of airway occlusion with tongue post cleft palate repair (tongue suture may be placed x 24hrs)
5. Risk of perioperative bleeding (more common with cleft palate repair with bone graft)

ANESTHETIC GOALS:

- Anticipate possible difficult airway
- Inhalational induction is often preferred (especially in syndromic cleft lips or palates)
- Avoid trauma to cleft lip or palate during intubation - consider moist sterile gauze barrier
- Local anesthetics to minimize pain intra- and postoperatively (local infiltration or infraorbital nerve block)
- Smooth Emergence - undue crying may place excessive tension on the repair.
- Ensure patency of airway, return of protective airway reflexes and adequate bleeding control with clot evacuation prior to extubation
- Ensure patency of nasal airways post-procedure (stents in place or adequate passage of air with no stents)

PATHOPHYSIOLOGY AND EPIDEMIOLOGY

- Cleft lip and/or cleft palate are among the most common congenital anomalies
- Cleft lip and palate occur as a defect of palatal growth in the first trimester.
- Comprise a heterogeneous group of facial malformations
- Multifactorial genesis including:
  - Chromosomal abnormalities and familial trait
  - Drugs (e.g., steroids, antiepileptics [benzodiazepines]), chemotherapy, excessive maternal vitamin A intake), folic acid deficiency, maternal tobacco or alcohol abuse (fetal alcohol syndrome)
  - Maternal diabetes mellitus
  - Possible link with maternal age (younger than 20 years or older than 39 years of age) and increased paternal age
- Overall, a form of orofacial clefting is estimated to affect 1 in 500 to 600 newborns.
- Male: Female ratio 2:1—cleft lip and palate. Isolated cleft palate more common in females
- Incidence for cleft lip is 1/750 for Caucasians; more common in Asians; less in African Americans. Left cleft more common than right; both more common than bilateral, in the ratio of 6:3:1
- Incidence for cleft palate alone is 1:2500
- Associated anomalies are seen in ~29% of cleft lip cases

HISTORY

- Cleft lip can reliably be diagnosed by ultrasound at 18 to 20 weeks after conception
- Palatal cleft is diagnosed by examination after delivery
- These disorders are associated with more than 300 syndromes and diseases. The common ones include:
  - Pierre Robin Syndrome
  - FAS
  - Trisomy 21
  - Treacher Collins Syndrome
  - Nager Syndrome
  - Goldenhar's Syndrome
  - Stickler Syndrome
  - Klippel-Feil Syndrome
  - Velocardiofacial Syndrome
  - CHD
  - Kidney defects
  - Subglottic stenosis
  - OSA
- History should include evaluation for these syndromes/diseases

SYSTEMS

- Airway
  - Careful assessment is necessary as associated anomalies may affect airway or lungs.
  - Possible Difficult Airway
  - Newborns with a cleft lip usually do not have problems maintaining airway patency
  - The tongue may fall into the cleft and obstruct the airway (nose breathing) for infants with a cleft palate or cleft lip/palate
  - Isolated cleft palate may be more discrete, sometimes requiring meticulous inspection, and palpation of the hard and soft palate
- Breathing/Respiratory
  - Upper respiratory tract infections are common in this age group

- Risk of recurrent aspiration from poor feeding mechanics
- CVS
  - Risk of CHD – assess for CHD on physical exam
  - Pediatric cardiology consult for those with known CHD
  - However, in
- Other
  - Recurrent or chronic otitis media is common – may require myringotomy and tube insertions.
  - Poor nutrition due and FTT to feeding difficulties
    - Assess nutritional status from physical exam and comparison to expected growth for age.
  - Delayed development of speech is common in the older child with cleft palate. Some of these children may be hearing impaired.
  - Many patients with orofacial congenital malformations require multiple procedures; emotional support and psychological assessment of these patients are essential.

INVESTIGATIONS

- High incidence of iron deficiency anemia
- Other investigations based on nutritional status or co-existing diseases

TREATMENT

- Surgical correction of cleft lip defect is usually performed at 3 months of age to allow sufficient time for maturation and associated abnormalities to become apparent.
- Variation in age range at which the cleft palate is repaired varies significantly (most often neonatal period to approximately 18 months of age), before the development of speech.
- In children with isolated orofacial clefting, the prognosis is excellent, although in some cases (mainly in cleft palate patients), further corrective surgeries (e.g., pharyngo- and palatoplasty) may be required.
- Cleft lip
  - May be either unilateral or bilateral, associated frequently with clefts of the alveolus and palate.
  - Surgical repair involves the design and execution of geometric flaps on the medial and lateral sides of the cleft and primary repair of the cleft nasal deformity.
  - Nasal repairs involve extensive mobilization of the alar cartilages and transfer of tissue up into the cleft nasal vestibule and floor, with nasal stents often placed.
    - All of these factors can decrease or occlude nasal airway breathing.
  - In large clefts, a lip adhesion may be performed as an initial stage several months before the actual definitive correction of the cleft lip
    - This procedure basically involves creating a wound on either side and suturing the muscles, mucosa, and skin together.
  - Presurgical orthopedic devices may be placed and manipulated, instead of a lip adhesion, to bring a wide, bony cleft into better opposition for a tension-free complete repair.
    - These are custom-fitted and may be fixed with pins to the palate.
    - They are removed in the OR at time of repair.
- Cleft Palate
  - Cleft-palate repair usually is done when the child is 9–18 months old,
  - Repair involves mobilizing the lateral soft tissue and moving it toward the midline to close the cleft and elongate the palate, if necessary.
  - An important goal of palate repair is normal anatomic approximation of the levator palati muscles, which are responsible for oronasal valving in speech and swallowing.
  - The most important goal of cleft-palate repair is the attainment of normal speech.
    - Children with unrepaired or inadequately repaired clefts develop nasal-sounding speech patterns termed rhinolalia.
  - Traction with a tongue suture may be placed as this often proves helpful in restoring patient's airway (maintained 24 h).
  - 15% of children undergoing cleft palate repair will need some type of secondary palatal lengthening procedure after 3 yr.
- Alveolar Bone Cleft
  - The size of the cleft is variable; it may be unilateral or bilateral and is associated with cleft lip and palate.
  - The alveolar segments are often collapsed such that orthodontic expansion is required before bone graft and repair.
  - A bone graft is placed in between these two layers to consolidate the upper arch.
    - Cancellous bone usually is taken from the iliac crest or corticocancellous bone from the outer table of the skull.
  - Increased risk of blood loss.
  - Most nasal and lip revision surgery should be put off until the alveolus is reconstructed, since this is the base on which the lip and nose sit.

PRE-OPERATIVE MANAGEMENT AND OPTIMIZATION

- However, many surgeons and anesthesiologists find the 'rule of ten' helpful: the child should have an Hb >10 g, be 10 wk old, and weigh 10 lbs
- Optimize any co-existing syndromes/diseases prior to surgery
- Assess nutritional status from physical exam and by comparison to expected growth for age. Treat FTT and blood (iron deficiency anemia) and electrolyte abnormalities in these patients prior to surgery
- Patients without signs of airway obstruction > 9 mo old, may benefit from sedation pre-operatively
  - Midazolam (0.5–0.75 mg/kg po) or ketamine (5 mg/kg po) ~30 min preop may be used.
- Type and Crossmatch for 1 U PRBC (especially for cleft palate + alveolar bone cleft repair)

ANESTHETIC OPTIONS

- General Anesthesia Only

ANESTHETIC SETUP

- Standard CAS Monitors
- Difficult Airway Cart as indicated by Co-Existing Diseases and Airway Exam
- Other Monitors as per co-existing diseases

INTRA-OPERATIVE AND MANAGEMENT OF ANESTHESIA

- **Induction**
  - Inhalational induction is usually preferred especially with syndromic clefting
    - Consider fiber optic intubation in patients with suspected difficult airway. Also consider elective tracheostomy under local anesthesia in patients with severe airway abnormalities.
  - An oral Ring-Adair-Elwyn (RAE) endotracheal tube is preferred.

- Throat pack is often placed
- **Maintenance**
  - Standard GA +/- muscle relaxants
  - For cleft palate surgery a Dingman mouth gag may be used.
    - If used considerations should be made to have it released intermittently to allow reperfusion of the tongue; each manipulation of the gag may affect the ETT placement.
  - Adequacy of ventilation should be checked after every position change.
  - For children with difficult airways and a small posterior pharynx, consider preop dexamethasone 0.5 mg/kg IV to decrease palatal edema
  - Multimodal analgesia is important: Opioids, Local anesthetics, Tylenol, Possibly NSAIDs
- **Emergence**
  - Ensure removal of Throat pack
  - Pediatric patients should wake up in an unagitated state
    - Undue crying may place excessive tension on the repair
  - Immediate elbow restraints for children × 2 wk.

POST-OPERATIVE MANAGEMENT AND CONSIDERATIONS

- Cool mist in the postoperative period has been used successfully to keep these children comfortable and prevent airway complications.
- The nose may be stented initially.
- The arms are often splinted to prevent removing of nasal stents or disrupting the repair.
- Patients with difficult airways or severe obstructive sleep apnea undergoing palatal surgery should be evaluated for ICU monitoring postoperatively.
- Cleft Palate surgery patients should be admitted to hospital overnight (Risk of bleeding and OSA)
- Cleft lip patients can be day surgery patients if appropriate age
- Analgesia
  - Local anesthetics should be used to minimize pain intra- and postoperatively.
    - Local infiltration +/- an infraorbital or sphenopalatine nerve block
  - Opioids, Tylenol, Possible use of NSAIDS

COMPLICATIONS

- Retained throat pack if there are Sx of airway obstruction in immediate postop period
- Patients with velocardiofacial syndrome may have medially displaced internal carotid arteries placing these major arteries in harm's way during dissection along the posterior pharyngeal wall
- Airway edema/croup
  - Treatment includes cool, humidified, 100% O_2 mask, or nebulization 2.25% racemic epinephrine (0.5 ml in 3 mL NS. If posterior pharyngeal edema is present
  - Consider dexamethasone 0.5 mg/kg iv
  - May require reintubation – possible difficult reintubation
- Hemorrhage
- Post-Operative Respiratory Failure (esp. with obstructive sleep apnea)

REFERENCES

- Stoelting's Anesthesia and Co-Existing Disease, 5th edition
- Anesthesiologist's Manual of Surgical Procedures, 4th edition
- Manual of Pediatric Anesthesia, 5th Edition
- Anesthesiology Review 3rd Edition
- Cote