Pierre Robin Sequence and Mandibular Distraction

- Kaiser SCPMG

- Stacey Francis, MD
- Craniofacial Surgeon

Obstructive Sleep Apnea in the Neonate and Infant

- The correlation between micrognathia and neonatal airway obstruction was originally described by Pierre Robin in 1920's
  - Glossoptosis
  - Micrognathia or retrognathia
  - Respiratory distress
  - Often cleft palate (wide U shaped)
  - 1/20,000 to 1/30,000 live births
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  -
-

Artist's rendition of fetal growth in weeks.

Marques IL, de Sousa TV, Carneiro AF, Barbieri MA, Bettiol H, Gutierrez MR. Clinical experience with infants with Robin sequence: a prospective study. Cleft Palate Craniofac J. 2001;38(2):171–178

Clinical Red Flags

- Desaturations in supine position
  - ANY desat in prone or lateral position
- Work of breathing, retractions
- Apnea, hypopnea, tachypnea
- Poor feeding
- Consideration of feeding tube
- Failed car seat test

Daniel M, Bailey S, Walker K, Hensley R, Kol-Castro C, Badawi N, Cheng A, Waters K. Airway, feeding and growth in infants with Robin sequence and sleep apnoea. Int J Pediatr Otorhinolaryngol. 2013;77(4):499-503.

Pre-op Work-up

- Labs: HCO3 (CBG or Chemistry)
- Documentation of desaturations- degree, position, length
- Sleep Study
- 3D CT scan
- Multidisciplinary team- consults
  1. Neonatologist
  2. Pulmonologist/Sleep Medicine Specialist
  3. ENT
  4. Plastic Surgeon/Craniofacial Surgeon

Bull MJ, Givan DC, Sadove AM, Bixler D, Hearn D. Improved outcome in Pierre Robin sequence: effect of multidisciplinary evaluation and management. Pediatrics. 1990;86(2): 294-301.

Polysomnography

- Can be done as inpatient at FMC
- Specific parameters used: AHI, hypoventilation, hypoxia
- NAPTIME study not as effective
- Importance to distinguish central vs obstructive sleep apnea

Anders T, Parmalee AH, Emde RA. Manual of Standardized Terminology: Technology and Criteria for Scoring of States of Sleep and Wakefulness in Newborn Infants. UCLA BIS/BRI Publications, Los Angeles (1971)

Crowell DH, Brooks LJ, Colton T, Corwin MJ, Hoppenbrouwers TT, Hunt CE, Kapuniai LE, Lister G, Neuman MR, Peucker M, Ward SL, Weese-Mayer DE, Willinger M. Infant polysomnography: reliability. Collaborative Home Infant Monitoring Evaluation (CHIME) Steering Committee. *Sleep.* 1997;20(7):553-560.

Freed G, Pearlman MA, Brown AS, Barot LR. Polysomnographic indications for surgical intervention in Pierre Robin sequence: acute airway management and follow-up studies after repair and take-down of tongue-lip adhesion. *Cleft Palate J.* 1988;25(2):151-155.

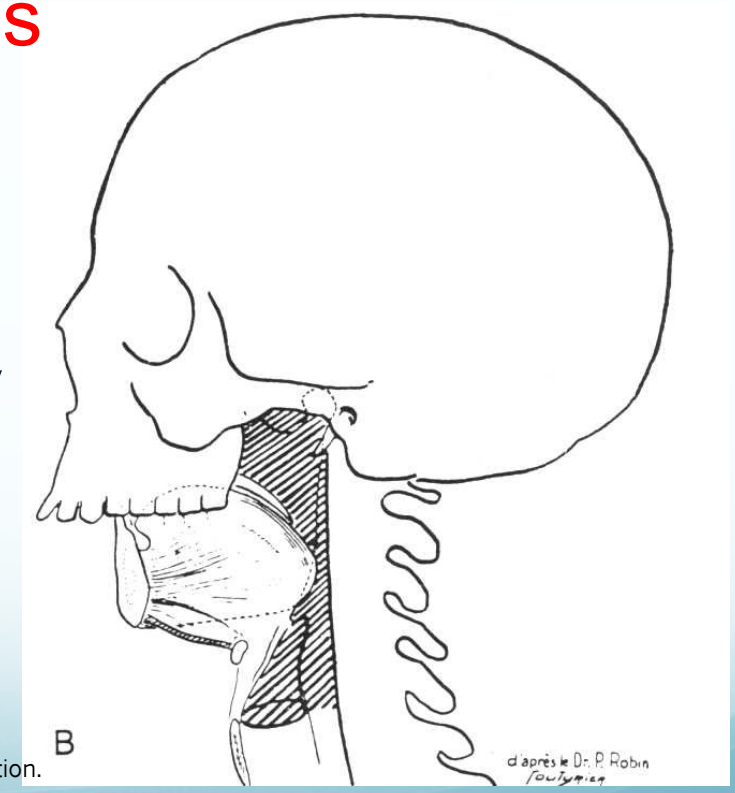
Management Options

Acute Conservative:

- Lateral or Prone positioning
- Supplemental oxygen
- Nasopharyngeal airway

Surgical:

- Tongue lip adhesion
- Mandibular Distraction
- Tracheostomy



Jarrahy, R. Controversies in the Management of Neonatal Micrognathia: To Distract or Not to Distract, That Is the Question. Journal of Craniofacial Surg. 2012;23(1):243–249.

Management based on respiratory status

1. Respiratory distress
   - Initial conservative measures failed:
     - prone position and supplemental oxygen
   - Sleep study performed to evaluate the Apnea Hypopnia Index(A.H.I.), hypoxemia, and hypoventilation
2. Respiratory failure
   - Required intubation for oxygenation
   - No sleep study performed

Hammoudeh J, Bindingnavele VK, Davis B, Davidson Ward SL, Sanchez-Lara PA, Kleiber G, Nazarian Mobin SS, Francis CS, Urata MM. Neonatal and infant mandibular distraction as an alternative to tracheostomy in severe obstructive sleep apnea. Cleft Palate Craniofac J. 2012;49(1):32-38

Contraindications to Distraction

- Central sleep apnea
- Severe esophageal reflux
- Primary lung disease
- Airway lesions
- Co-morbidities
- Poor prognosis

Shen W, Jie C, Chen J, Zou J, Ji Y. Mandibular distraction osteogenesis to relieve Pierre Robin severe airway obstruction in neonates: indication and operation. J Craniofac Surg. 2009;20 Suppl 2:1812-1816.

The Risk of No Intervention

- Persistent OSA > pulmonary hypertension > cor pulmonale
- 2011 event- KP infant presented with heart failure

Dykes EH, Raine PA, Arthur DS, Drainer IK, Young DG. Pierre Robin syndrome and pulmonary hypertension. J Pediatr Surg. 1985;20(1):49-52.

Freeman MK, Manners JM. Cor pulmonale and the Pierre Robin anomaly. Airway management with a nasopharyngeal tube. *Anaesthesia.* 1980;35(3):282-286.

Johnson GM, Todd DW. Cor pulmonale in severe Pierre Robin syndrome. *Pediatrics.* 1980;65(1):152-154.

Cogswell JJ, Easton DM. Cor pulmonale in the Pierre Robin syndrome. *Arch Dis Child.* 1974;49(11):905-908.

What are the Goals?

- For Respiratory Distress:
  - Wean off supplemental O2
  - Prevent long term OSA complications
    - CHF
- For Respiratory Failure:
  - Extubation
    - Avoid tracheostomy

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Tracheostomy Morbidity

- Published morbidity ranges from 5%-70%
- Hemorrhage – early or late :
- Erosion of innominate artery
- Tracheoinnominate fistula
- Pneumothorax
- Obstruction with mucous, blood
- Displacement
- Infection
- Tracheal stenosis
- Tracheocutaneous fistula
- Delayed language development
- Significant psycho-social impact on family
- Financial impact as a surgical burden of disease

Tonsager SC, Mader NS, Sidman JD, Scott AR. Determining risk factors for early airway intervention in newborns with micrognathia. Laryngoscope. 2012;122 Suppl 4:S103-104.
Handley SC, Mader NS, Sidman JD, Scott AR. Predicting Surgical Intervention for Airway Obstruction in Micrognathic Infants. *Otolaryngol Head Neck Surg.* 2013;148(5):847-851
Meyer AC, Lidsky ME, Sampson DE, Lander TA, Liu M, Sidman JD. Airway interventions in children with Pierre Robin sequence. *Otolaryngol Head Neck Surg.* 2008;138(6):782–787

Distraction Osteogenesis (slow growing bone)

- Principles of Distraction Osteogenesis
  - Latency phase (24 hours)
  - Distraction phase (2-3 weeks)
    1.8 mm/day (2 turn/every 8 hours)
    extubate during this phase
  - Consolidation phase (3 months)

- 3 stages (2 surgeries) (requiring anesthesia)
  a. Distractor placement (distract for 2-3 weeks)
  b. Removal distractor activation arms
  c. Removal of distractors (3-4 months after)

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Peri-op HNS Work-up

- Flexible laryngoscopy
  - Posterior tongue
  - Epiglottal collapse
  - Laryngomalacia
  - Hemangiomas
- Rigid bronchscopy
  - Tracheal webs
  - Vascular tumors

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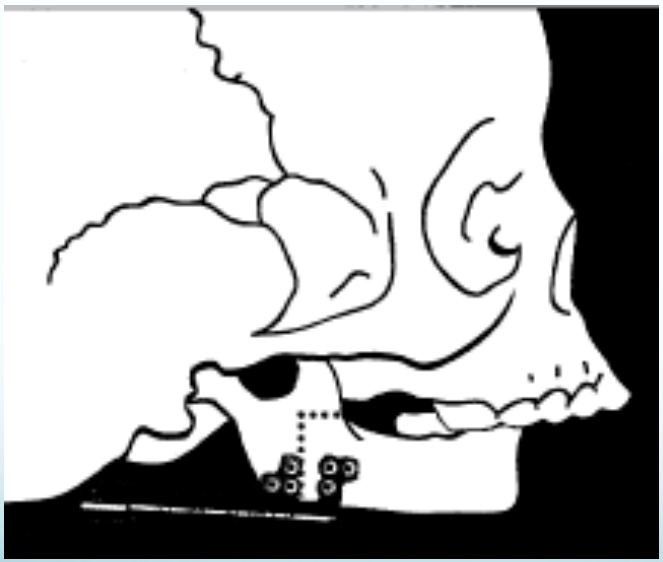
Schematic Pre and Post



Post Op NICU/PICU Protocol

- KLS Screwdriver and Distraction Log Bedside
- Begin distraction 24 hours post op
  - Plastic surgery to do first distraction, then RN vs family
- Extubation TBD (max swelling) / Collaboration CF, NICU, HNS
- Feeding attempts begin early post extubation
- Plastic surgery to monitor post op via PA or pictures virtually
- Distraction usually lasts 16 days/2 weeks (1.8 mm/day for 30 mm)
- Biweekly HCO3 (chemistry vs CBG) until normal
- Repeat sleep study after distraction complete
- Discharge after when no positional desaturations and oral feeding ok
  - Apnea monitor generally not necessary (depending on PSG results)

During Distraction

- Pain management
  - First few days IV, then Tylenol, then nothing before distraction
- Antibiotic ointment around pin sites with each distraction
- Keep Log of distance distracted
- Things to watch for:
  - Pin site infections (common)
  - Asymmetry of the jaw (common)
  - Lack of advancement of the jaw
    - Maxillo-mandibular discrepancy
  - Continued poor feeding

General Pediatric Care

- After mandibular distraction, treat baby as a normal baby with cleft palate
- Will need special bottle, but no changes in solid food recommendations (other than will be messy)
- Should follow in craniofacial clinic yearly for surgical and speech and orthodontic care
- For PRS babies that are mild and not distracted, monitor for work of breathing, failure to thrive
- Ophthalmology consult (usually done by craniofacial team)
  - Usually no other consults needed
  - Stickler syndrome

SCPMG Protocol

- Development of a regional standardized care plan
- Regional craniofacial management
- Prospective IRB study with Dennis Hwang
- Would like input from NICU and peds MD's and RN's
- Will support KPCCN efforts and regional care

Conclusion

- Pierre Robin leads to respiratory distress of varying degrees
- Requires a comprehensive multidisciplinary team approach
- Work up for management includes:
  - Sleep study
  - ABG/CBG
  - Consults to Craniofacial Surgery
- Feeding issues may indicate need for intervention
- Mandibular distraction is a viable alternative to tracheostomy and conservative treatment