A 2-month-old male with Pierre Robin Sequence presents for bilateral sagittal split mandibular osteotomies and placement of external distractors.

Moderators: Smokey Clay, M.D.; Paul Samuels, M.D.; Nishan Goudsouzian, M.D.

Institutions: Cincinnati Children's Hospital Medical Center, Massachusetts General Hospital

Objectives:

1. Review Pierre Robin Sequence.
2. Discuss syndromes associated with micrognathia.
4. Identify approaches to the micrognathic airway.

Case history:

A 3.2 kg 2-month-old former 29 week premie twin presents for bilateral sagittal split mandibular osteotomies and placement of external distractors. At birth he was intubated for a period of 3 days. He was transitioned from nasal CPAP to nasal cannula and then room air. He was maintained in the prone or lateral positions due to frequent airway obstruction in the supine position resulting in oxygen desaturation and bradycardia. His family resisted early recommendations for tracheostomy and sought other surgical options.

Describe Pierre Robin Sequence:

Pierre Robin Sequence describes the triad of micrognathia, glossoptosis, and a U-shaped cleft palate.

- Micrognathia: abnormally small lower jaw
- Glossoptosis: downward displacement or retraction of the tongue
- U-shaped cleft palate

Pierre Robin first focused on the description and study of this disorder. It had been written about much earlier by others (1822 Saint-Hilaire and 1846 Fairbain and 1911 Shukowsky).

He noted that these anatomical abnormalities lead to upper airway obstruction.
The initial insult in the sequence is failure of anterior growth of the mandible that results in the tongue physically blocking the fusion of the palatine shelves, resulting in a U-shaped cleft palate. Problems encountered include difficulty with feeding and respiratory distress.

The tongue base is retrodisplaced, resulting in oropharyngeal narrowing.

**Conditions associated with micrognathia:**

- Pierre Robin Sequence
- Hallerman-Streiff Syndrome
- Trisomy 13
- Trisomy 18
- XO Syndrome
- Progeria
- Treacher-Collins Syndrome
- Smith-Lemli-Opitz Syndrome
- Russell-Silver Syndrome
- Cri Du Chat Syndrome
- Marfan Syndrome

**Options for treatment:**

- Supportive measures: prone positioning and nasal CPAP
- Invasive measures: 23% of patients with micrognathia have a tongue-based obstruction that requires intervention beyond these noninvasive measures.
  - Tracheotomy.
  - Tongue-lip adhesion procedures.
    - Glossopexy
  - Mandibular distraction osteogenesis.
- Discuss relevant pros and cons to these procedures

**Describe the anesthetic plan for induction and maintenance of anesthesia:**

Anesthesia considerations for micrognathia should include a formal approach to airway management. Pediatric difficult airway management is almost always due to difficult laryngoscopy, not difficult mask ventilation.

- Spontaneous ventilation with inhalational induction or IV induction.
- Fiberoptic (other indirect methods) versus direct laryngoscopy.
- Rescue with intubating stylette or LMA.
- Notification and presence of otolaryngologist with rigid bronchoscopy available.
Case continued:

The child underwent a mask inhalation induction in the lateral position. We progressively controlled ventilation. Ventilation was then possible with bimanual jaw thrust and oral airway when turned supine. The view was moderate to difficult to expose with direct laryngoscopy using a size one glide scope. Otherwise, the glottis was normal. A 3.0 uncuffed endotracheal tube was placed. Placement was confirmed and the endotracheal tube was sutured by the plastic surgeon to the anterior mandible at a depth of 10 cm.

Sagittal osteotomies performed and external distractor placed.

The patient was moved to the ICU intubated without event.

The mandibular distractor was advanced at a rate of 2-3 mm per day.

The patient was extubated on post-operative day 7 and weaned to room air.

He returned to the operating room for removal of the mandibular distractors on post-operative day 23. He has been maintained on room air without a single episode of oxygen desaturation. He continues to grow with minimal difficulty eating.

References:


