Micrognathia

Children's Mercy KANSAS CITY | Evidence Based Practice

Inclusion Criteria:
Infants and children with suspected micrognathia and **any** of the following:
- Stertor/stridor/increased work of breathing when supine
- Feeding difficulty

Features of Robin Sequence
- Micrognathia
- Glossoptosis
- Upper airway obstruction with or without cleft palate

Criteria for *Otherwise Unfavorable Sleep Study*
- Any AHIo with severe hypoventilation:
  - End-tidal or transcutaneous CO_2 > 50 mmHg for > 25% of total sleep time (TST) ***- or -***
  - CO_2 increase > 10 mmHg from awake baseline to sleep
- Any AHIo with saturation nadir (*with obstructive events*) < 80%
- Any AHIo with prolonged severe events

Supplemental O_2 Response
- Decrease in AHIo > 50% from baseline
- No hypoventilation

QR code for mobile view

Patient admitted with suspected micrognathia* and airway concerns
**preferred terminology to retrognathia*

Consults
- Plastic Surgery
- Otolaryngology (ENT): Bedside flexible fiberoptic laryngoscopy performed to evaluate for synchronous airway lesions
- Genetics
- Pulmonology

Has Robin sequence been diagnosed?
- **No** → *Off Pathway*. Primary and consulting teams to determine further management
- **Yes** →

Does airway obstruction require intubation or noninvasive support (CPAP or HFNC)?
- **No** →
  - ***Discuss*** need for sleep study with Pulmonology ***- and -***
  - ***Consult*** Occupational Therapy for feeding evaluation

  → What are the sleep study results?
    - **AHIo ≤ 10** → **Outpatient Follow-Up**:
      - Sleep clinic in 1-2 months after discharge
      - JAWS clinic in 6 months
    - **AHIo > 10 - 39.9** → ***Consider*** a repeat sleep study with O_2, if additional data are needed to determine whether supplemental O_2 is an appropriate treatment option → Is there shared decision-making that supplemental O_2 is an appropriate management strategy?
      - **Yes** → ***Discharge*** with supplemental O_2 (*flow determined by Pulmonology based on sleep study)*
      - **No** → **AHIo ≥ 40 -or- *Otherwise Unfavorable Sleep Study***
- **Yes** → ***Discuss*** need for CT scan (maxillofacial) with Plastic Surgery (*Plastic Surgeon will place CT order*)

AHIo ≥ 40 -or- *Otherwise Unfavorable Sleep Study*

Are factors favorable or unfavorable for mandibular distraction?
- **Unfavorable** → **Multidisciplinary Approach**
  Options may include:
  - Noninvasive support
  - Tracheostomy
  - Other surgical interventions

  → **Outpatient Follow-Up**:
  - JAWS clinic in 6 months
- **Favorable** → Surgery for MLB +/- endoscopic intervention, +/- concurrent or staged mandibular distraction

Did mandibular distraction take place?
- **No** → **Multidisciplinary Approach**
- **Yes** →
  - ***Consult*** Pulmonology to determine plan for post-operative sleep study in 4-6 weeks (*location of sleep study to be determined pending patient status*)
  - ***If persistent OSA,*** consulting teams to determine further management

  → **Outpatient Follow-Up**:
  - Sleep clinic in 1-2 months after discharge
  - JAWS clinic as determined by Plastics team
  - Cleft clinic for patients with cleft palate
  - Ascertain need for separate ENT follow-up

Timing of Diagnostic Evaluation
- ***If patient is a preterm infant***, consider obtaining sleep study, and other relevant evaluations once the infant is term-corrected and on lower levels of respiratory support, if clinically feasible

Mandibular Distraction Considerations
- Determined by plastic surgeon (*imaging may be ordered*)
- Factors that may be unfavorable:
  - Chronic lung disease
  - Multiple anomalies/complex syndromes
  - Aspiration/dysphagia present
  - Abnormal neuromuscular status
  - Other surgical considerations
  - Multi-level airway obstruction
    - In some instances may be treated separately, followed by mandibular distraction (e.g., laryngomalacia)

Abbreviations (laboratory & radiology excluded):
AHIo = Apnea-hypopnea index obstruction
JAWS = Jaw, Airway and Sleep clinic