Micrognathia

Children's Mercy KANSAS CITY | Evidence Based Practice

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

QR code for mobile view

Patient admitted with suspected micrognathia* and airway concerns

*preferred terminology to retrognathia

Pediatric Specialty Consults:
- Plastic surgery
- ENT:
  - Bedside Flexible Laryngoscopy
  - Determine need for difficult airway designation
- Genetics
- Pulmonology

Beside Flexible Laryngoscopy:
- Used to evaluate for obstruction or other cause of symptoms
- May be false negative for glossoptosis due to crying

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

Has Robin Sequence/Pierre Robin Sequence been diagnosed?

No → Primary and consulting teams to determine further management

Yes

Criteria for *Otherwise Unfavorable Sleep Study*:
- Any AHIo with severe hypoventilation (*% TST with end-tidal CO_2 or trans-cutaneous*) CO_2 > 50 mmHg for >25% of TST or > 10 mmHg for baseline awake
- Any AHIo with saturation nadir (*with obstructive events*) < 80%
- Any AHIo with prolonged severe events

Sleep Study Results
AHIo Classification

Score	Classification
< 1	No OSA
> 1 - 10	Mild OSA
> 10 - 39.9	Moderate OSA
≥ 40	Severe OSA

For central sleep apnea or mixed sleep apnea, Primary and consulting teams to determine further management

Does airway obstruction require intubation or noninvasive support (CPAP)?

No → Discuss need for sleep study with Pulmonology and consult Occupational Therapy for feeding evaluation

Yes

Tracheostomy:
- For alternative options, shared decision-making with family involvement is very important

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

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 ≥ 40 Or ***Otherwise Unfavorable Sleep Study***

AHIo >10 - 39.9 → Second sleep study with O_2

Did patient respond to supplemental O_2?

Yes → Discharge with supplemental O_2 (*flow determined by Pulmonology based on sleep study)*

No

Are factors favorable or unfavorable for mandibular distraction?

Unfavorable → **Consider Tracheostomy:**
- Arrange pre-tracheostomy meeting with aid of Social Work

Outpatient Follow-Up:
- Infant Home Vent clinic in most cases
- JAWS clinic in 6 months

Favorable → Surgery for MLB +/- endoscopic intervention, with anticipated mandibular distraction

Are results of MLB clear to proceed with mandibular distraction?

No

Yes

- Consult Pulmonology prior to mandibular distraction to 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

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):
ENT = Ear, Nose & Throat
CPAP = Continuous positive airway pressure
AHIo = Apnea-hypopnea index obstruction
TST = Total sleep time
JAWS = Jaw, Airway and Sleep clinic
MLB = Microlaryngoscopy and bronchoscopy
OSA = Obstructive sleep apnea

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Link to synopsis and references

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