Diagnosing Tongue Base Obstruction in Infants with Pierre Robin Sequence: Is Sleep Endoscopy Superior to Awake Endoscopy?

Jake J. Lee, BS[1]; Matthew D. Ford, MS, CCC-SLP[2]; Noel Jabbour, MD[3]

[1]University of Pittsburgh School of Medicine; [2]Cleft-Craniofacial Center, Children's Hospital of Pittsburgh of UPMC; [3]Division of Pediatric Otolaryngology, Children's Hospital of Pittsburgh of UPMC

Abstract

Objectives
To investigate whether awake endoscopy can diagnose base-of-tongue obstruction as reliably as sleep endoscopy in infants with Pierre Robin sequence (PRS).

Study Design
Individual case-control study.

Methods
A retrospective review of 141 infants with PRS managed between 2005 and 2015 was conducted. Thirty-five underwent both bedside awake flexible fiberoptic nasopharyngolaryngoscopy and drug-induced sleep endoscopy in the operating room. Base-of-tongue collapse was considered pathologically obstructed if described as moderate or severe or classified as Yellon grade 2 or 3. Sensitivity, specificity, and positive likelihood ratio of awake endoscopy findings were calculated with 95% confidence intervals (CI). McNemar's test was also utilized to examine inter-test differences.

Results
Awake endoscopy was performed at a median age of 5.5 days (range 0 to 61), and drug-induced sleep endoscopy was performed at a median age of 21 days (range 0 to 216). Awake endoscopy had 50.0% sensitivity (CI 27.2-72.8%) and 86.7% specificity (CI 59.5-98.3%) for detecting base-of-tongue obstruction compared to drug-induced sleep endoscopy; false negative rate was 28.6% (*N*=10). Positive likelihood ratio was 3.75 (CI 0.96-14.65). Compared to awake endoscopy, sleep endoscopy demonstrated significantly more cases of base-of-tongue obstruction (p = 0.039).

Conclusion
Bedside awake endoscopy has low sensitivity for detecting base-of-tongue collapse in infants with Pierre Robin sequence. Due to the substantial number of false negatives, awake endoscopy may not be a reliable diagnostic modality for ruling out tongue base obstruction in this susceptible population. Drug-induced sleep endoscopy may be indicated in high-risk patients in order to avoid missing the diagnosis of upper airway obstruction.

Introduction

- Pierre Robin sequence (PRS): microretrognathia, glossoptosis, & possible cleft palate (Fig 1) → upper airway obstruction, feeding difficulty, & growth retardation.[1]
- Upper airway obstruction may be subtle, manifesting only during sleep. 85-100% of PRS infants have concurrent sleep-disordered breathing.[2,3]
- Awake flexible fiberoptic nasopharyngolaryngoscopy has been used to assess for sites of obstruction and to grade base-of-tongue (BOT) collapse in patients with PRS.[4]
- Drug-induced sleep endoscopy (DISE) in adults and children has been shown to identify more cases of obstruction than awake endoscopy (AE).[5,6]
  - Use of DISE in infants with PRS to has not been described.
- **Can awake endoscopy diagnose base-of-tongue obstruction as reliably as sleep endoscopy in infants with PRS?**

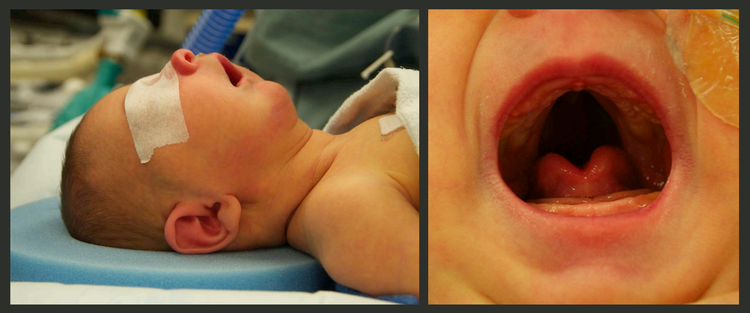

Fig 1. An infant with PRS that demonstrates microretrognathia, glossoptosis, and cleft palate.

Methods

- Retrospective case series with chart review of the 141 consecutive infants with PRS managed between January 1, 2005 and July 31, 2015.
  - 35 patients underwent both AE and DISE.
- Awake flexible fiberoptic nasopharyngolaryngoscopy was performed and interpreted by pediatric otolaryngologists at the bedside.
- DISE was performed and interpreted by pediatric otolaryngologists in the operating room.
  - Sedation was achieved using continuous intravenous propofol infusion.
- BOT collapse was graded as normal, mild, moderate, or severe or 0, 1, 2, or 3 according to the Yellon classification[7].
  - Moderate (2) or severe (3) BOT collapse was considered pathologically obstructed (Fig 2).
- Sensitivity, specificity, positive predictive value, negative predictive value, positive likelihood ratio, and negative likelihood ratio of AE findings compared to the gold standard of DISE were calculated with 95% confidence intervals (CI).
- McNemar's test was utilized to examine inter-test differences between AE and DISE.

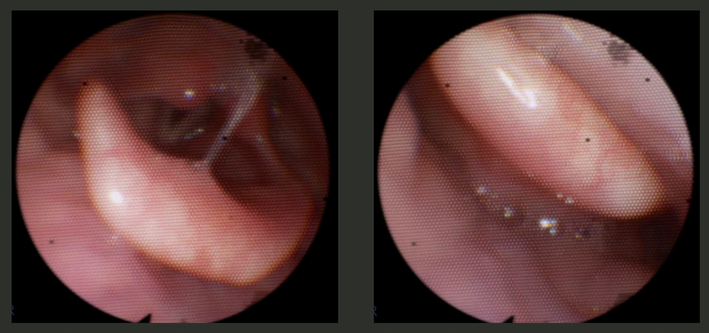

Fig 2. Left: no BOT collapse on endoscopy. Right: moderate-to-severe BOT collapse on endoscopy.

Results

- Awake endoscopy performed at a median age of 5.5 days (range 0 to 61 days).
- DISE performed at a median age of 21 days (range 0 to 216 days).

	(+) BOT collapse on DISE	(-) BOT collapse on DISE
(+) BOT collapse on AE	10	2
(-) BOT collapse on AE	10	13

Table 1. Prevalence of BOT collapse as determined by AE and DISE in all 35 patients.

- AE had 50.0% sensitivity and 86.7% specificity for detecting BOT obstruction compared to DISE (Tables 1 & 2).
  - False negative rate = 28.6% (*N*=10).

	Value	95% CI
Sensitivity	50.0%	27.2-72.8%
Specificity	86.7%	59.5-98.3%
Positive Likelihood Ratio	3.75	0.96-14.65
Negative Likelihood Ratio	0.58	0.36-0.93
Prevalence	57.1%	39.4-73.7%
Positive Predictive Value	83.3%	51.6-97.9%
Negative Predictive Value	56.5%	34.5-76.8%

Table 2. Statistical output of AE findings compared to DISE.

- Sleep endoscopy demonstrated significantly more cases of BOT obstruction compared to awake endoscopy (McNemar's test, 2 tail, p=0.039).

Discussion & Conclusions

- Bedside awake endoscopy has low sensitivity for detecting BOT obstruction in infants with PRS compared to drug-induced sleep endoscopy.
- There is a substantial false negative rate with awake endoscopy.
- Awake endoscopy may not reliably rule out BOT obstruction in this susceptible population.
- Presumably, the upper airway is more patent during an awake exam due to the increased muscular tone of a crying infant.[5]
- Drug-induced sleep endoscopy may be indicated as an adjunct in high-risk infants with PRS in order to avoid missing the diagnosis of upper airway obstruction.

Contact

Jake J. Lee, BS
University of Pittsburgh School of Medicine
MD Candidate, Class of 2016
Email: lee.jake@medstudent.pitt.edu
Phone: 201-679-7758

References

1. Robin P. Glossoptosis due to atresia and hypotrophy of the mandible. *Am J Dis Child* 1934;48:541–7.
2. Anderson IC, Sedaghat AR, McGinley BM, *et al*. Prevalence and severity of obstructive sleep apnea and snoring in infants with Pierre Robin sequence. *Cleft Palate-Craniofacial J* 2011;48:614–8.
3. Daniel M, Bailey S, Walker K, *et al*. Airway, feeding and growth in infants with Robin sequence and sleep apnoea. *Int J Pediatr Otorhinolaryngol* 2013;77:499–503.
4. Côté A, Fanous A, Almajed A, *et al*. Pierre Robin sequence: review of diagnostic and treatment challenges. *Int J Pediatr Otorhinolaryngol* 2015;79:451-64.
5. Fishman G, Zemel M, DeRowe A, *et al*. Fiber-optic sleep endoscopy in children with persistent obstructive sleep apnea: inter-observer correlation and comparison with awake endoscopy. *Int J Pediatr Otorhinolaryngol* 2013;77:752-5.
6. Certal VF, Pratas R, Guimaraes L, *et al*. Awake examination versus DISE for surgical decision making in patients with OSA: A systematic review. *Laryngoscope* 2015; Epub ahead of print.
7. Yellon RF. Epiglottic and base-of-tongue prolapse in children: grading and management. *Laryngoscope* 2006;116:194-200.