

## Laryngomalacia 2020

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## Annual Meeting of American Pediatric Society, New York City, 1905

- “appears shortly after birth. The infant otherwise may be in apparent health, lying quietly in its crib or the arms of a nurse, a peculiar crowing or grunting is heard with inspiration. The sound may be scarcely audible, or at some distance, as across the room and of a loud sawing timbre. As a rule, the sound is heard only with inspiration, but may in severe cases be heard in expiration also. At times the grunting may become inaudible, but reappears at intervals.
- Autopsy: “epiglottis so disposed that its sides are rolled up and almost in contact, and the AE folds draw the epiglottis downward and backward, covering the superior opening of the glottis.”

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All perinatal stridor is from laryngomalacia. That's why it's also known as congenital laryngeal stridor

- True
- False

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## Laryngomalacia: Historical

- Stridor used to be a diagnosis.
  - Thymectomy, radiation, radical epiglottectomy
  - Tracheotomy
- Laryngeal diagnosis was inferential.
  - Flexible laryngoscopy not available/avoided
- 1980s: LGM ≠ congenital laryngeal stridor
- Realization: LGM is the most common of many causes of stridor.

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Child born 6 hours ago with inspiratory stridor. Least likely diagnosis

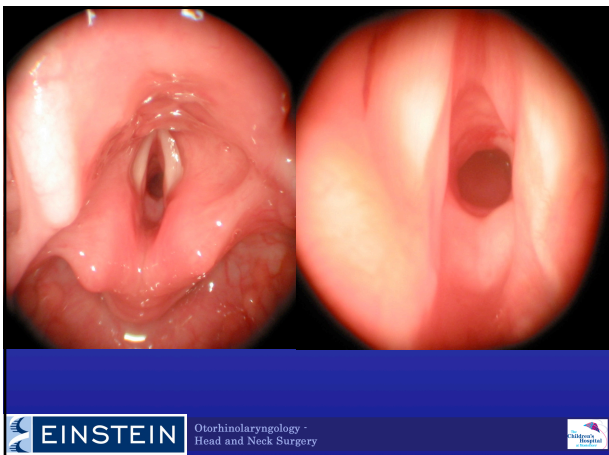
- Subglottic stenosis
- Subglottic hemangioma
- Bilateral vocal cord paralysis
- Laryngomalacia
- Tracheomalacia

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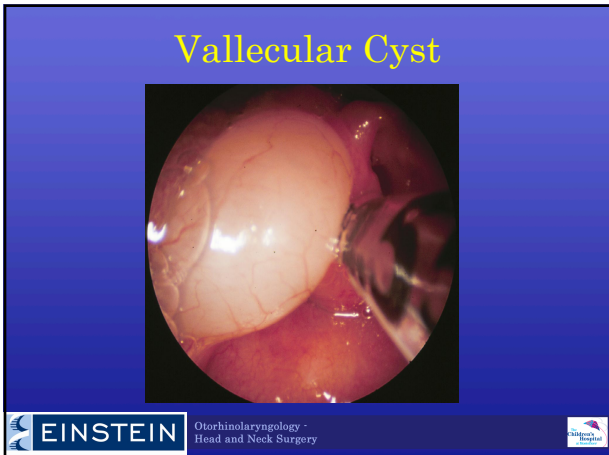
## Subglottic Hemangioma



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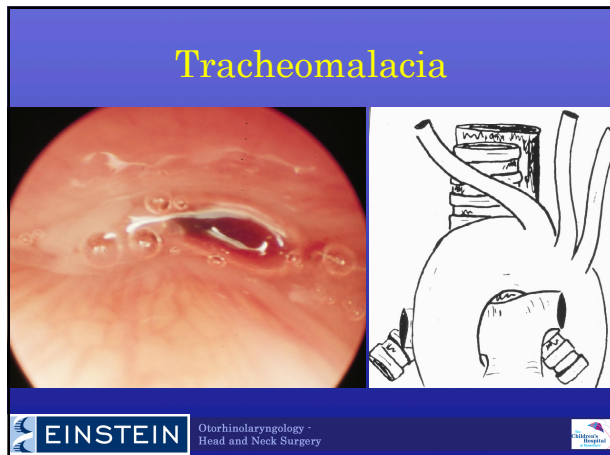
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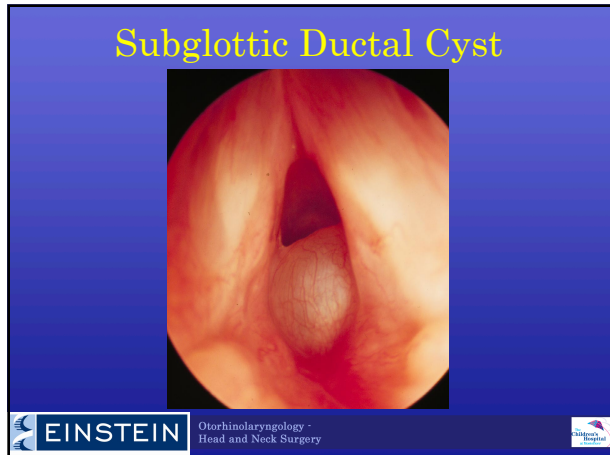
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If stridorous newborn weighs 10 pounds and was a difficult forceps delivery, your suspicion goes up for:

- Subglottic stenosis
- Subglottic hemangioma
- Vocal cord paralysis
- Laryngomalacia
- Tracheomalacia

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## Vocal Fold Paralysis



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Widespread pediatric flexible fiberoptic laryngoscopy can be attributed in part to:

- Improved optics
- The HIB vaccine
- Peds ORL subspecialty development
- All of the above

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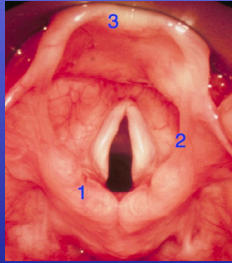
## Laryngomalacia ...

- Is most common cause of neonatal stridor
- Causes most stridor in children
- Should never be treated with tracheotomy
- Affects males and females equally
- All of the above

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## Laryngomalacia

- most common congenital laryngeal anomaly
- most common cause of stridor in infants
- high pitched inspiratory stridor
- presents hours to months after birth
- prolapse of supraglottic structures



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## Most laryngomalacia is caused by

- Unfavorable anatomy
- Prematurity
- Second airway lesion
- Sensorimotor abnormalities
- All of the above

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## Etiology

- Anatomic vs neurologic
  - Anatomic theory supported by response to surgery; acquired laryngomalacia is recognized anatomic problem
  - Majority of evidence supports neuro etiology for neonatal LGM.
    - Alters with state of awareness.
    - Resolution correlates with neuro development
- Abnormal laryngeal sensorimotor testing
  - Correlates with disease severity
  - Improves as symptoms resolve
    - Thompson DM Laryngoscope 2007
- LPR strongly associated in prospective studies
  - 64% Giannoni et al
  - 100% Matthews et al

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## Natural history

- Typically presents days after birth
- 2:1 M:F ratio
  - More even ratio among non-Caucasians
  - Higher rates in low birth weight and premies
    - Edmundson et al, IJPO 2011
- Stridor becomes more prominent over several months.
- Largely resolved by 12 MO
- Completely resolved by 18 MO

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## Laryngomalacia poses some risk of:

- Aspiration
- Poor growth
- Sleep apnea
- B and C
- All of above

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## Laryngomalacia Evaluation

- Apnea, cyanosis, general comfort
- Feeding
- Growth
- Other anomalies?
- Flexible laryngoscopy
  - State dependent
- Sleep study?


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## Nonsurgical Treatment

- HOB elevation
- Thickened feedings
- Prone position (contradicts 'back to sleep')
- Anti-reflux medications
  - OTC topicals
  - H2 blocker
  - PPI
- Oxygen




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## IPOG LGM Consensus (2016)

- Treatment algorithm based on symptoms –
  - **Mild:** stridor only
    - observe
  - **Moderate:** stridor with coughing, choking, regurgitating, feeding difficulty
    - Acid suppression
    - Possible feeding therapy/swallow eval
  - **Severe:** apnea, cyanosis, FTT, pulm hypertension, cor pulmonale
    - Same as moderate with consideration of SGP




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8-week child with laryngomalacia not responsive to 3-weeks of Ranitidine, HOB elevation. Weight has decreased below 5<sup>th</sup> percentile






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## You advise:


- Proton pump inhibitor
- Barium swallow
- Sleep study
- Drug induced sleep endoscopy (DISE)
- DLB and supraglottoplasty




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Respiratory Statistics (Normal Values):			
Apnea Hypopnea Index / Respiratory Disturbance Index (<5/hr):	34.6	Obstructive Apnea Index (<1/hr):	21.0
Central Apnea Index (<1/hr):	1.8	Snore Intensity:	frequent, light
Hypopnea Index:	12.0	Minimum Oxygen Saturations (>92%)(%):	65
Baseline Oxygen Saturation (%):	100	Max ET/CO2 (<53 torr) (torr):	58
Baseline ET/CO2 (torr):	35		



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## Surgical indications


- Performed for approximately 15% of cases presenting to ORL
- Objective: cyanosis, sleep apnea, FTT, pectus deformity.
- Subjective: feeding difficulties, progressive stridor, general discomfort, sleep disordered breathing.

– Mancuso et al Arch OHNS 122: 302-6, 1996

All indications are relative.




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## Pectus Deformity Association

- elevated rates of pectus deformities
- deformity exists in 0.1-0.3% infants
- 137 LGM patients (2008-12)
  - 9 (6.6%) identified retrospectively (p=.001)
    - Virbalas et al, IJPO 2013
- seems to correlate with more severe disease.




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## All children with laryngomalacia should have:

- DLB
- DLB before SGP
- pH probe
- Modified barium swallow




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## Second lesion

- Described by Bluestone et al 1996
- May occur in up to 50-60%
  - Frequency correlates with LGM severity
    - Schroeder et al. Arch OHNS 2009;135:647-51.
    - Dickson et al. Ann ORL 2009;118:37-43.
- Relationship may be
  - Anatomic (Bernouille's law),
  - Chemical (edema from LPR), or
  - Neurologic (central tone)
    - Probably not a genetic association.
- Merits comprehensive endoscopy prior to surgery.
- A second lesion increases need for SGP 4.5X
  - Landry, Thompson IJPO 2012






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## Which type of laryngomalacia is most common?

- Type 1
- Type 2
- Type 3
- Type 1 and 2 combined




Type 1
Type 2
Type 3



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## Type 1 most common (n=48)

- Type 1 (57%)
- Type 2 (15%)
- Type 3 (12%)
- Types 1 and 2 (15%)
  - Olney et al, Laryngoscope 109: 1770-75, 1999.







Type 1
Type 2
Type 3





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## Supraglottoplasty (formerly called epiglottoplasty)

- Endoscopy first
- Laser
- Cold
- Microdebrider
- Avoid interarytenoid scar
- +/- divide folds
- Role for epiglottopexy/ectomy
- Tracheotomy

	Intra-operative	Post-operative
Type 1		
Type 2		
Type 3		



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## Preferred Anesthesia Method

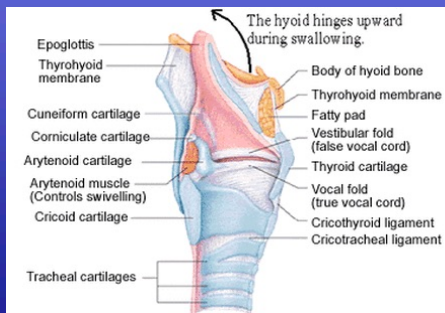
1. apneic technique
2. spontaneous ventilation
3. subglottic jet ventilation
4. supraglottic jet ventilation
5. endotracheal tube intubation

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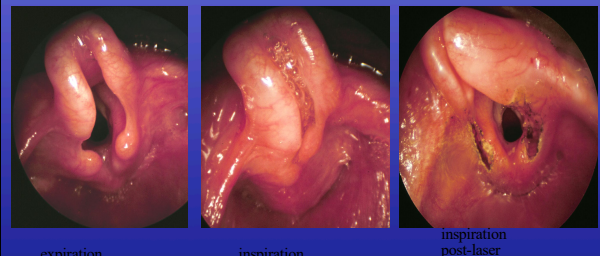
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## Laryngeal cartilages



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## Laryngomalacia: Intraoperative



expiration

inspiration

inspiration post-laser

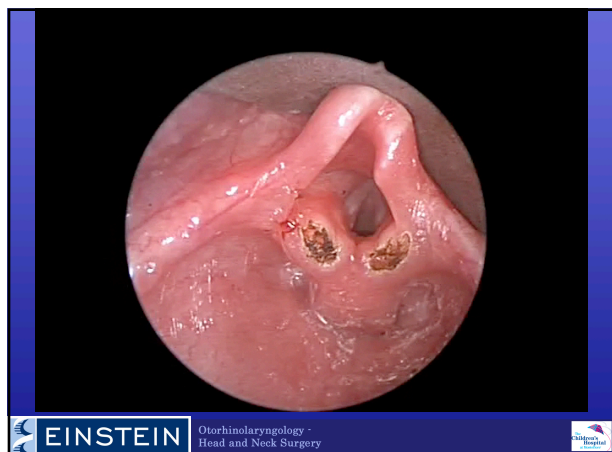
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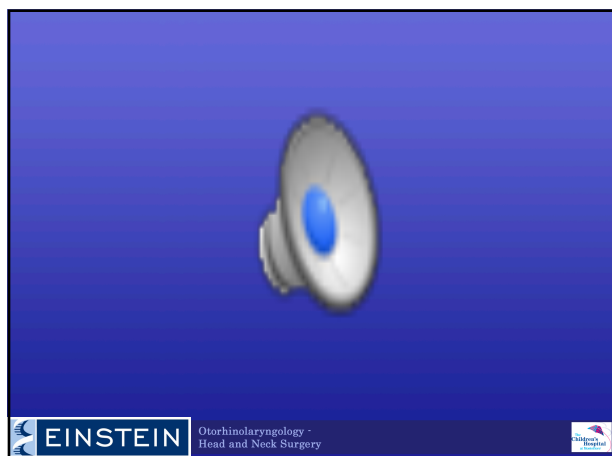


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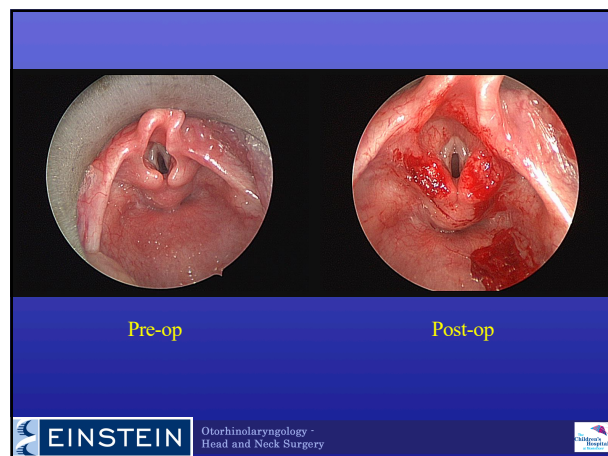
### Microdebrider-assisted Supraglottoplasty

- Alternative to laser or microlaryngeal instrumentation.
- Controlled trimming of AE folds
- 5/5 pts had resolution of stridor without complications.
  - Zalzal and Collins. IJPO 69: 305-9, 2005.

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### Appropriate post op care involves

- Intubation
- PICU care
- NPO
- Anti-reflux medication

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### Outcomes

- Higher rates of aspiration vs normal infants
  - Related to neuro impairment, not surgery
- 36 supraglottoplasties at CHAM 2008-14
  - 89% symptoms relieved after 1 procedure
    - 2 revisions, 2 tracheotomies
  - Mean postop stay 1.7 days for outpatients
    - 18.4 days for inpatients
- Poor outcomes and prolonged stay correlates with co-morbidity.

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## Supraglottoplasty- Failures and Complications

- Isolated laryngomalacia- 102 pts
- Additional congenital anomalies (ACA)-34 pts
- All failures 8.8% ACA group
- Complication rate 7.4%
  - 3.6% supraglottic stenosis
    - Denoyelle F. Arch OHNS 129: 1077, 2003.

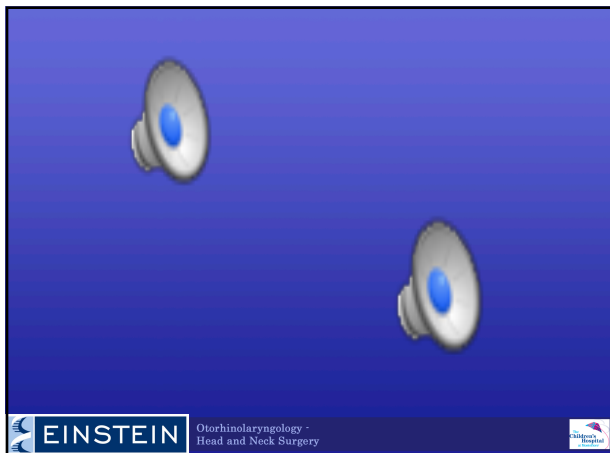
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## Supraglottoplasty failures occur ...

- In < 10% of attempted cases.
- Occasionally from excessive surgery around posterior commissure.
- Most often among children with other congenital anomalies.
- At a higher rate when associated with pharyngolaryngomalacia.

•OHNS 129: 1077, 2003.

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## Do we do this surgery enough?

- Balancing risk of hypoxia and UAO vs potential anesthetic neurotoxicity
- In 1989 Hollinger predicted supraglottoplasty will be performed not just for “life-threatening obstruction” but also “to improve quality of life”
  - Laryngoscope 99: 136-42, 1989
- Comparison to T&A for OSA
  - Is supraglottoplasty underutilized?
  - White D, Laryngoscope 2012

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## Type 3 Laryngomalacia



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If this patient has moderate sleep apnea and feeding difficulty, and you exclude a second lesion, you advise:

- Partial epiglottectomy
- Epiglottis-tongue adhesion
- Corniculate vaporization
- Oxygen and thickened feedings

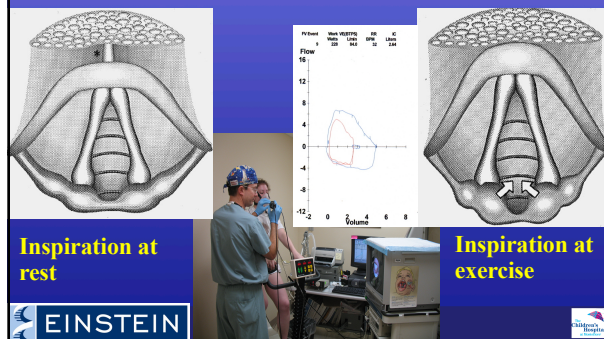
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## Late Onset Laryngomalacia

- Late onset LGM may cause dysphagia in toddlers, OSA in children, and exercise intolerance in teenagers.
  - Richter GT et al, Arch Otolaryngol Head Neck Surg. 2008 Jan;134:75-80.
- post-ictal, post head injury
- **Increasingly recognized:** “Occult” LGM as cause for persistent OSA after T&A

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## Exercise Induced Laryngomalacia



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## All of the following are true about LGM except

- Most cases are caused by abnormal sensorimotor integration.
- Type 3 LGM is the most common.
- Surgical success rates are approximately 90%.
- Roughly 50% of children have a second airway lesion.

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## Take home points Laryngomalacia

- The most common cause of pediatric and neonatal stridor
- Must consider other and second causes
- Most cases managed non surgically
- Most surgery very successful
- Keep in mind comorbidities and atypical presentations
- Surgery: balance how often and how much

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