History of Laryngomalacia

- Congenital stridor – 1st described in 1853 by French physicians Rilliet and Barthez

- Congenital laryngeal obstruction – 1st described in 1897 by Sutherland and Lack

- “Laryngomalacia” – 1st coined by Chevalier Jackson in 1942
  - “-malakia” = Greek for softening of an organ
Anatomy of Larynx

- **Supraglottis** – tip of epiglottis to laryngeal ventricle
  - Includes epiglottis, aryepiglottic folds, false vocal cords, arytenoid cartilages

- **Glottis** – true vocal cords

- **Subglottis** – undersurface of true vocal cords to inferior border of cricoid
Muscles of the Larynx

- Respond to pressure sensors
- Actively dilate supraglottis during inspiration
- Prevent collapse of supraglottic larynx
Mechanics of Breathing

- Depression of diaphragm creates negative intrathoracic pressure

- Pressure transmitted to trachea and larynx

- Supraglottic collapse prevented by:
  - Laryngeal musculature
  - Bernoulli Principle
Stridor

- **Inspiratory:**
  - Supraglottic obstruction
  - High-pitched
- **Biphasic:**
  - Extrathoracic tracheal obstruction
  - Intermediate-pitched
- **Expiratory:**
  - Intrathoracic tracheal obstruction
  - Retractions
Differential Diagnosis

- **Supraglottis**
  - LARYNGOMALACIA
  - Laryngoecele/saccular cyst
  - Valleeular cyst
  - Lingual thyroid

- **Glottis**
  - Glottic web/atresia
  - Laryngeal cleft
  - Laryngeal stenosis
  - Laryngoecele
  - PapillomatosiS

- **Subglottis**
  - Subglottic stenosis
  - Subglottic cyst
  - Foreign body
  - Croup

- **Trachea**
  - Tracheomalacia
  - Tracheal stenosis/web
  - Vascular ring/sling
  - Tracheoesophageal fistula
  - Foreign body
Differential Diagnosis

Epiglottitis

Croup
Differential Diagnosis

Subglottic stenosis

Papillomatosis
Differential Diagnosis

Laryngocoele

Vallgugdial Cyst
Laryngomalacia

- Supraglottic airway obstruction due to
  - Flaccid laryngeal tissue
  - Narrowed laryngeal inlet
  - Inward collapse of supraglottic structures on inspiration

- Most common congenital laryngeal anomaly

- Most common cause of congenital stridor (60-75%)
Presentation of Laryngomalacia

- Intermittent inspiratory stridor
  - High-pitched

- Worsens with feeding, agitation, supine positioning
  - Must take breaks to breathe while feeding

- Normal cry/phonation

- Severity varies
  - Mild: may improve with crying
  - Moderate – severe: worsens with crying
Natural History

- Presents within 1st 2 weeks of life
- Symptoms may worsen, then peak at 6-8 months
- Median time to spontaneous resolution = 9 months of age
- 75% with no stridor at 18 months
- 85-90% – resolve by 2 years without sequelae
- 10-15% – complicated by life-threatening effects
Diagnosis - Endoscopy
Flexible Fiberoptic Laryngoscopy
Types of Laryngomalacia

- **Type 1** – Anterior prolapse of mucosa overlying arytenoid cartilages (57%)
- **Type 2** – Short aryepiglottic folds → tethering of supraglottic structures in close antero-posterior approximation (15%)
Types of Laryngomalacia

- Type 3 - Posterior collapse of epiglottis over glottis (13%)
- Combination of above types (15%)
Associated Pathology

• Isolated finding in otherwise healthy infant.
• Association with neurologic disorders
  • E.g. cerebral palsy
• 15-20% have a synchronous lesion
  • Mild subglottic stenosis
  • Tracheomalacia
Pathophysiology of Laryngomalacia

- **Neuromuscular hypotonia** (Thompson and Turner, 1900)
  - Poor muscular tone causing laryngeal collapse
  - Association with neurologic disorders
  - Dysfunction in sensorimotor integration of afferent reflexes, brainstem function and motor responses

- **Altered sensorimotor integrative function** (Thompson, 2010)
  - Intrinsic muscles of larynx not triggered to stent larynx open
  - Seen by lack of laryngeal adductor reflex
  - May be central/brainstem
  - Possibly related to damage from reflux
Pathophysiology of Laryngomalacia

- Anatomic abnormalities
  - Flaccid laryngeal tissue
  - Narrow laryngeal opening
  - Histology: subepithelial edema, dilated lymphatics

- Gastro-esophageal reflux (GER)
  - Increased laryngeal edema
  - Altered sensation and functional denervation of larynx
  - Possibly caused by laryngomalacia
Laryngomalacia and Reflux

- Reflux → Laryngomalacia
  - Gastric acid exposure → edema of laryngeal tissue → prolapse of inflamed tissue

- Laryngomalacia → Reflux
  - Respiratory effort against fixed obstruction → increased negative intrathoracic pressures → reflux

OR

- Abnormal Anatomy → Neuromuscular Disorder → Reflux → Supraglottic Airway Obstruction → Altered Respiratory Mechanics → Edema → Reflux

"No, you back off! I was here before you!"
Laryngomalacia and Reflux

- Up to 80% of patients
- Control GERD prior to surgery
  - $H_2$ blockers, proton pump inhibitors
  - Nissen fundoplication +/- gastrostomy tube
- Symptoms of reflux
  - Regurgitation, emesis
  - Dysphagia
  - Feeding intolerance
  - Weight Loss
Gastroesophageal Reflux Disease

• Histological correlations

Gastroesophageal Reflux Disease

- Matthews et al. 1999
  - Increased prevalence in laryngomalacia
Gastroesophageal Reflux Disease

- Giannoni et al. 1998
  - Increased severity and complications

![Bar chart showing the number of infants tested with different grades of GER and laryngomalacia](chart.png)
## Other Contributing Factors

<table>
<thead>
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<th>Laryngomalacia</th>
<th>Uncomplicated/mild %</th>
<th>Complicated/severe</th>
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<td>18 (6/33)</td>
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</table>

Laryngomalacia and Aspiration

• 25-72% of patients with severe laryngomalacia also have aspiration
  • Clinical swallow exam
  • Video fluoroscopic swallow study (VFSS)
  • Fiberoptic endoscopic evaluation of swallowing (FEES)

• Symptoms of aspiration:
  • Coughing and choking with feeds
  • Cyanosis, apneic episodes, respiratory distress around meal times
Laryngomalacia and Aspiration

- Altered anatomy and neuromuscular reflexes $\rightarrow$ dysfunction of suck-swallow-breathe sequence

- Disruption of Laryngeal Adductor Reflex (LAR) $\rightarrow$ laryngeal penetration
  - Vagus nerve-mediated reflex
  - Closure of vocal cords and cessation of breathing as food passes into pharynx

- Rapid feeding $\rightarrow$ aspiration
  - Increased metabolic demands, weight loss, hunger
Laryngomalacia and Aspiration

- Altered anatomy and neuromuscular reflexes → dysfunction of suck-swallow-breathe sequence
- Disruption of Laryngeal Adductor Reflex (LAR) → laryngeal penetration
- Increased work of breathing + reflux + aspiration = FAILURE TO THRIVE
  - Closure of vocal cords and cessation of breathing as food passes into pharynx
- Rapid feeding → aspiration
  - Increased metabolic demands, weight loss, hunger
Additional Work-Up

- Assessment of swallowing
  - Barium swallow/ MBS/ FEES
  - Assessment for reflux and aspiration

- Airway Fluoroscopy
  - Dynamic study
  - Supplement to endoscopy to evaluate subglottis and trachea

- Chest X-Ray/ Neck X-Ray
  - Croup
  - Foreign body
  - Pneumonia

Aspiration Pneumonia

Esophageal Foreign Body
Additional Work-Up

- Sleep study
  - MCP/PSG
  - Evaluate sleep apnea
  - Must distinguish central versus obstructive apnea

- Echocardiogram
  - Check for cardiac origin of cyanosis
  - Preoperative clearance
  - Effects of OSA
Management of Laryngomalacia

• Sleep prone rather than supine
• Close observation of upper respiratory tract infections
• Feeding modifications
  • Pacing, thickened formula, upright feeding
• Reflux therapy
  • Feeding precautions
  • H₂ blockers
  • Proton pump inhibitors
• Follow growth curve
Indications for Surgery

- 10-31% of infants need surgery for laryngomalacia
- Not following expected course/ responding to medical therapy
- **Severe laryngomalacia**
  - Respiratory compromise
  - Feeding difficulty – reflux/aspiration
  - Weight loss/failure to thrive
  - Obstructive sleep apnea
Direct Laryngoscopy/Bronchoscopy

- Symptoms that do not correlate with degree of laryngomalacia noted on FFL
- Evaluate for synchronous airway lesions (12-27%)
- Evaluate for surgical intervention
Direct Laryngoscopy/Bronchoscopy

- Normal Vocal Cords
- Normal Subglottis
- Curled Epiglottis
- Arytenoid Prolapse
Surgery for Laryngomalacia

- Tracheotomy – standard of care for severe laryngomalacia for ~100 years
- 1889, Variot – suggested excision of aryepiglottic folds for relief of obstruction
- 1922, Iglauer – resect part of epiglottis
- 1928, Hasslinger – performed 3 endoscopic resections of aryepiglottic folds
- Mid-1980s – more interest in endoscopic surgery → current standard of care:
Supraglottoplasty

- Procedure tailored to site/mechanism of obstruction
Supraglottoplasty

Type 1
- Excise redundant arytenoid tissue

Type 2
- Divide shortened aryepiglottic folds

Type 3
- Pexy posteriorly displaced epiglottis to base of tongue
Intraoperative Images

Preoperative

Postoperative
Intraoperative Images
Benefits of Supraglottoplasty

- Well tolerated procedure
- High success rate
  - 69-94% with resolution of airway and feeding symptoms
  - Improvement of reflux, aspiration and sleep apnea
- Low failure rate
  - 1-3% need tracheotomy
  - Typically patients with associated neurologic disorder or syndrome
  - Discoordinate pharyngolaryngomalacia

Nissen fundoplication may also help failures!
Postoperative Improvements in Reflux

- Hadfield et al. 2003
- Significant Decrease in Reflux Index after Supraglottoplasty
Postoperative Improvements Sleep Apnea

• Marked improvement in Respiratory Disturbance Index after supraglottoplasty
Controversy on Aspiration

- **Traditional Thoughts** – Rastatter, Hollinger, et al.
  - Few patients with preoperative aspiration – 10/39 (26%)
  - Some improvement of aspiration with surgery – 2/10 (20%)
  - *Supraglottoplasty causes postoperative aspiration* – 13/29 (45%)
    - Regardless of technique, cold knife v. CO₂ laser

- **New Views** – Richter, Thompson et al.
  - High rate of preoperative aspiration with severe laryngomalacia – 36/44 (72%)
  - *Supraglottoplasty leads to resolution of aspiration* – 31/36 (86%)
    - May not improve aspiration in patients with medical comorbidities
  - *Supraglottoplasty does not cause aspiration* in patients without preoperative aspiration – 0/14 (0%)
Complications of Supraglottoplasty

- Laryngeal webs, granulation tissue
  - Preserve interarytenoid tissue to avoid web
- Supraglottic stenosis (4%)
- Transient dysphagia (10-15%)
- Aspiration
  - Newly diagnosed and persistent from preoperative aspiration
Take Home Points on Laryngomalacia

- Congenital inspiratory stridor due to supraglottic collapse
  - 80-90% resolve by 1-2 years of age with conservative management
- Strong association with gastroesophageal reflux
- Aspiration and sleep apnea are common complications
- More severe symptoms warrant surgical intervention
Thank you
History of the Supraglottoplasty

- 1984, Lane – otologic instruments to trim tips of arytenoid processes and incise aryepiglottic folds
  - Resolution of pectus excavatum from airway obstruction
- 1985, Seid – CO₂ laser division of aryepiglottic folds
History of Supraglottoplasty

- 1987, Zalzal – endoscopic epiglottoplasty
- 1995, Kelly and Gray – unilateral division of aryepiglottic fold, unilateral resection of redundant tissue
  - 94% success rate, no complications
- 2001, Loke – simple endoscopic division of aryepiglottic folds
  - 90% success rate, no complications
- 2005, Zalzal – microdebrider-assisted
Schematic of Supraglottoplasty
Supraglottoplasty

- Supraglottoplasty = aryepiglottoplasty
  - CO\textsubscript{2} laser, laryngeal microscissors or microdebrider

- Different procedures based on what portion of collapsing supraglottis is to be removed
  - Tissue overlying arytenoids
  - Aryepiglottic folds
  - Posterior portion of epiglottis