Current Concepts in Diagnosis and management of Laryngomalacia

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Overview

- Discuss Pathogenesis
- Clinical presentation
- Laryngomalacia and GERD
- Diagnosis
- Medical and surgical management
- Parental counseling
What is laryngomalacia?

- Laryngomalacia (LM) is the commonest congenital laryngeal anomaly of the newborn characterized by flaccid laryngeal tissue and inward collapse of the supraglottic structures leading to upper airway obstruction.

Etiopathogenesis

- Cartilage immaturity
- Anatomic abnormality
- Neuromuscular immaturity
Cartilage immaturity

- First proposed by Sutherland and Lack in the late 19th century
- Delayed development of the cartilaginous support of the larynx
- Theory has been disproved
  - No histological evidence of chondropathy
  - Incidence not different in premature infants
Anatomic abnormality

- LM is a result of the exaggeration of an infantile larynx (Iglauer 1922)

- May or may not be an important factor since stridor is not seen in all infants with ‘omega epiglottis’

Neuromuscular immaturity

- There is a high prevalence of neurologic disorders with LM
- Some believe that neuromuscular immaturity leads to laryngeal hypotonia and LM
- May be one of the several components of LM
Laryngomalacia and GERD

- 80-100% of infants with LM have GERD
- It is not clear whether GERD is a cause or an effect of LM
- **EMPIRIC REFLUX THERAPY**
  - choking,
  - frequent emesis,
  - regurgitation
  - or feeding difficulty
Proposed pathogenesis of GERD in LM

- Respiration against a fixed obstruction
- Large –ve intrathoracic pressure
- Reflux into esophagus and LPR
- Increased obstruction
- Increased prolapse
- Laryngeal edema
Proposed pathogenesis of GERD in LM

Disruption of effective vagal tone to LES

Relative decreased LES

GERD

This pathogenesis leads credence to “NEUROLOGICAL IMMATUREITY” theory of LM
Conditions that worsen LM

- Prematurity
- Neuromuscular disorders: higher incidence, increased severity
- Synchronous airway lesion
  - 20% incidence
  - *Tracheomalacia, subglottic stenosis, bronchomalacia, pharyngomalacia, vallecular cyst*
  - Potentiates GERD
  - Surgical failures

Clinical presentation

- Stridor is the hallmark of congenital LM
  - High pitched, inspiratory, worsens with agitation, crying, feeding or in the supine position

- Feeding symptoms
  - Choking, coughing, prolonged feeding time, recurrent emesis, dysphagia, weight loss

- GERD symptoms

- Complications
Complications of LM

- 10-20% of patients present with complications
- Life threatening airway obstruction
- Failure to thrive
- Cyanosis
- Sleep apnea
- Pulmonary hypertension, developmental delay and cardiac failure
Classification schemes

- Based on symptomatology/flexible laryngoscopy
  - Mild
  - Moderate
  - Severe

- Based on mechanism of collapse
  - Anterior: epiglottis
  - Posterior: large arytenoids
  - Laterally: AE folds
Classification scheme based on symptoms, flexible laryngoscopy

MILD

SEVERE
Classification scheme based on mechanism of LM
Diagnosis

- History
- Physical examination
- Flexible laryngoscopy
Direct laryngoscopy video

You may have to click or double-click to see the movie
Complementary studies

- Chest X-ray to r/o aspiration
- Esophagram
  - Extent and degree of reflux
  - r/o concomitant GI disorder
- pH study if Nissen’s surgery is necessary
- Sleep Study to document severity of apnea in severe LM and in surgical failures
Management

- Medical
  - Empiric reflux acid suppression
  - Feeding modifications
  - Posture repositioning

- Surgical
  - Supraglottoplasty
  - Epiglottopexy
  - Tracheostomy


Empiric reflux acid suppression

- 80-100% of patients with LM have GERD
- H2 receptor antagonist (RA) or Proton pump inhibitor (PPI)
  - H2RA: ranitidine 3mg/kg three times daily
  - PPI: 1mg/kg daily
- If symptoms worsen → 6mg/kg of ranitidine at night + 1mg/kg of PPI daily

Feeding modifications

- Pacing
- Thickening formula feeds
- Upright feeding position
- Small, frequent feeds
Evolving concepts in surgical management of LM

Variot was the first to suggest removal of excess of AE tissue as treatment of LM

1920 → Re-introduction of concept of removal of SG tissue for treatment of LM

→ Sporadic reports of endoscopic trimming, partial epiglottopexy, wedge resection but no definite technique

1980 → Endoscopic techniques revisited and defined

Current → Endoscopic supraglottoplasty

Epiglottopexy
# Indications for surgery

<table>
<thead>
<tr>
<th>Absolute indications</th>
<th>Relative indications</th>
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<tbody>
<tr>
<td>Cor pulmonale</td>
<td>Aspiration</td>
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<tr>
<td>Pulmonary hypertension</td>
<td>Difficult-to-feed child who has failed medical intervention</td>
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<tr>
<td>Hypoxia</td>
<td>Weight loss with feeding difficulty</td>
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<tr>
<td>Apnea</td>
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<td>Recurrent cyanosis</td>
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<td>Failure to thrive</td>
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<td>Pectus excavatum</td>
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<td><strong>Stridor with respiratory compromise</strong></td>
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<td>Stridor with significant retractions</td>
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Contraindications

- Relatively uncommon
- Proceed with caution
  - Patients with comorbidities
  - Patients with multiple levels of airway obstruction
- Postpone surgery till resolution of URI
- WEIGHT AND AGE ARE NOT CI TO SURGERY
Pre-operative counseling

- Overnight hospitalization in the ICU
- The stridor will improve, but NOT DISAPPEAR
- Expect feeding improvement
- Reflux precautions and medications to be continued
- Risk of revision surgery
Anesthetic considerations

- Spontaneous breathing analgesia
- ETT in the nasopharynx, mouth
- Spray (1% lidocaine <2 years, 2% ≥ 2yrs)
- 0.5 mg/kg of decadron
- Intubation only for unstable patients or patients with poor pulmonary reserve

GOOD COMMUNICATION WITH THE ANESTHESIOLOGISTS
Surgical Set-up

- Rigid bronchoscopy
  - Visualize subglottis, trachea and bronchi
  - R/O synchronous airway lesion
- Assess VC mobility if not assessed previously
What is Supraglottoplasty

- It is a surgery designed to treat LM that aims to **trim the aryepiglottic folds** and **remove soft tissue**, overriding the **arytenoids**.
Surgical Steps of Supraglottoplasty

1. Pharyngoeopiglottic fold
2. Arytenoids

Extent of AE fold dissection
AE fold trimming with forceps and scissors
Surgical steps, contd...
Removal of redundant arytenoid mucosa

- Achieve hemostasis using Afrin pledgets
- Laser precautions

CO2 laser to remove redundant soft tissue over both arytenoids
Preserve inter-arytenoid mucosa
How much supra-arytenoid mucosa is to be removed?

- **Suction test**


Pre and Post op results

Pre-op

Post-op
Instrumentation

- Cold instruments
- CO2 laser
- Microdebrider
Post-operative care

- Intubation versus immediate extubation
- Feeding may be started when infant is awake
- One or two doses of post-operative steroids
- Aggressive empiric reflux therapy
- Follow-up in 2-4 weeks
- Monitor airway symptoms, apneic spells and feeding adequacy
Complications after surgery

- 8%, relatively uncommon
- Increases with multiple comorbidities
- Site-specific complications include bleeding, infection, web formation, granulation tissue
- Technical complications include supraglottic stenosis – difficult to treat, so best is prevention
Epiglottopexy

- Indicated if the primary level of obstruction is a retroflexed epiglottis
- Commonly seen in infants with global delay, hypotonia & neurological disorders
- Tell parents that tracheostomy may be necessary
- Main risks are aspiration, supraglottic stenosis
**Epiglottopexy: Surgical technique**

- Suspension of the patient
- Mucosa of the epiglottis is denuded with CO2 laser (1-10W) under microscopic guidance
- Additionally the epiglottis can be secured to the tongue base with 4.0 vicryl

Indications for tracheotomy

- Presence of > 3 comorbidities
- Severe sleep apnea
- Worsening symptoms after revision supraglottoplasty
Proposed algorithm for the treatment of mild and moderate laryngomalacia

Mild LM

1m FU + FL

2m FU + FL

3m FU + FL

FU @3m till resolution

Moderate LM

Acid suppression

Feeding modification

Symp worsen, persist

Complications

Surgery
Proposed algorithm for treatment of severe LM

- Severe LM
  - Maximum acid suppression and SGP
    - FU 2-4 weeks post op
      - FU as recommended for mild/moderate LM
    - Symptoms worsen
      - Consider PSG
        - Consider tracheotomy
      - Revision SGP
        - Symptoms worsen
        - pH study and Nissen’s fundoplication
So what did we learn?

- LM is the commonest congenital anomaly of the newborn larynx.
- 80-90% of patients have a benign course
- High pitched inspiratory stridor is the hallmark clinical presentation
- Feeding difficulties and GERD are seen in 80-100% of patients with LM
- History, PE and Flexible laryngoscopy aid diagnosis
Learning pearls contd...

- Identifying patients who will benefit most from surgery is of paramount importance

- “Less is More” when performing surgery on the infant larynx

- Strict FU and reflux therapy
QUESTIONS?