Introduction

- Rare congenital anomaly
- Incidence: congenital laryngeal anomaly occurs in 1 in 2000 live births, less than 0.3% of attributable to laryngeal cleft
  - boys: girls, with a ratio of 5:3
- possible autosomal dominant pattern of inheritance
- contributing factors: prematurity and hydramnios, but controversial
Embryology of larynx

- respiratory primordium develops from a diverticulum on foregut, at 25 days gestation
- tracheobronchial groove arise on either side and fuse in the midline and form tracheoesophageal septum
- The fusion is completed by the 6th week of gestation
Embryology of larynx

- Cricoid cartilage begins forming at 5 weeks of gestation and is derived from the 6th branchial arch. Chondrification should be complete by 6 weeks of gestation.

- Incomplete fusion of tracheoesophageal septum or cricoid cartilage will result in a laryngeal cleft / T-E fistula.
Classification of laryngeal cleft

- first patient with laryngeal cleft was described by Richter in 1792
- Pettersson reported the first surgical repair of type 1 laryngeal cleft in 1955
- Several classification systems
- Benjamin and Inglis (1989): 4 types
Classification of laryngeal cleft

- type 1: limited to the interarytenoid region, above level of true vocal folds
- type 2: cricoid lamina is partially involved, with extension below level of true vocal folds
- type 3: total cricoid cleft, extends completely through cricoid cartilage with or without further extension into cervical trachea
- type 4 extends into posterior wall of thoracic trachea and may extend as far as carina
Classification of laryngeal cleft
Classification of laryngeal cleft

(type 1)

(type 2)

(type 3)
Manifestations

- choking / aspiration while feeding
- regurgitation
- stridor
- recurrent pneumonia
- chronic cough
- cyanosis
Evaluation

- CXR might show pulmonary infiltrates secondary to aspiration
- modified barium swallow studies
- fiberoptic endoscopic evaluations of swallowing (FEES)
- flexible laryngoscopy: limited view of posterior glottic space
- intraoperative endoscopic-laryngoscopy under G.A
Evaluation--FEES

- dynamic view of swallowing that delineates timing and location of penetration and/or aspiration (valuable tools in assessing type 1 cleft)
- limitation: compliance of patient
Evaluation

- Laryngeal cleft may be obscured by redundant laryngeal and/or esophageal mucosa prolapsing into the cleft.
- Arytenoids must be parted with a probe to make the correct diagnosis.
Differential diagnosis

- newborn with feeding problems, repeated aspiration, and respiratory distress: detail evaluation
- complete prenatal and birth history (congenital infections, maternal drug use, hypoxia, or birth trauma)
- esophageal stricture, T-E fistula, cricopharyngeal spasm, neuromuscular abnormalities, laryngomalacia, GER, and vocal cord paralysis
Associated abnormalities

- gastrointestinal tract
- genitourinary tract
- cardiovascular
- Pallister-Hall syndrome (hypothalmic hamarblastoma, hypopituitarism, imperforate anus, and polydactyly)
- Opitz-Frias syndrome (cleft lip and palate, laryngeal clefts, cardiac anomalies, hypospadias, or defects of corpus collosum)
- VACTERL syndrome
Treatment

- Timing and approach for surgical repair depends on severity of symptoms, associated abnormalities, type of cleft
- In small cleft: conservative, positioning and thickened food to prevent aspiration, if failed, surgical repair
- Early diagnosis and surgical repair reduce irreversible pulmonary damage and other associated morbidities
Type 1 laryngeal cleft

- diagnosis can be missed, challenging
- presenting symptoms are usually non-specific (e.g. chronic cough and aspiration with feeds)
- difficult to visualize endoscopically

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Presenting symptoms of patients with type 1 laryngeal cleft</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptoms</td>
<td># of patients</td>
</tr>
<tr>
<td>Aspiration with thin liquids</td>
<td>18</td>
</tr>
<tr>
<td>Recurrent pneumonia</td>
<td>10</td>
</tr>
<tr>
<td>Chronic cough</td>
<td>7</td>
</tr>
<tr>
<td>Stridor</td>
<td>3</td>
</tr>
<tr>
<td>Hypoxia</td>
<td>2</td>
</tr>
</tbody>
</table>

Type 1 laryngeal cleft: establishing a functional diagnostic and management algorithm. 
**Type 1 laryngeal cleft**

- conservative measures first: anti-reflux therapy, thickened feeds
- surgical repair
  - Gelfoam injection
  - open approach and graft interposition
  - endoscopic approach
Treatment

- endoscopic approach
  - interrupted sutures used to close cleft by using suspension microlaryngoscopy under G.A with spontaneous breathing
Treatment

- Open approach
  - anterior approach:
    - exposes larynx through laryngofissure (surgical opening of larynx made through midline of thyroid cartilage)
    - excellent visualization of larynx defect
    - may cause laryngeal instability
  - lateral pharyngotomy approach
    - risk of injuring recurrent laryngeal n.
Treatment

- Open approach- interposition graft
  - tibial periosteum
  - mucosal margins dissected in 2 planes: posterior plane of pharyngoesophageal mucosa and anterior plane of laryngotraacheal mucosa
  - tibial periosteum is interposed as middle layer
Tibial periosteum sutured

Lateral fixation of tibial periosteum

Anterior plane of laryngeal mucosa sutured
Prognosis

- **mortality**: 46%
  

- **associated anomalies worsens the prognosis**

- **improvement in diagnostic methods and surgical techniques** → decrease in the perioperative mortality → 25%
  
Take home message

- rare congenital anomaly
- manifestations: recurrent aspiration pneumonias, stridor, choking on feeds or dysphonia
- 4 types of classification
- type 1: diagnosis can be missed; conservative therapy or endoscopic surgical repair
- open approach: revision surgery or more extensive clefts
- early diagnosis and surgical repair reduce irreversible pulmonary damage and other associated morbidities
References


