Tracheomalacia

R3 吳俊璟
Weakness of trachea

- Reduction of the longitudinal elastic fibers of the pars membranacea
- Impaired cartilage integrity, flaccidity of the supporting tracheal cartilage
- Reduced anterior-posterior airway caliber
- Histopathologic characteristics
  - The membranous trachea: larger
  - The ration of cartilage to muscle: reduced
# Major airway collapse classification

<table>
<thead>
<tr>
<th>Type</th>
<th>Presentation</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Congenital or intrinsic tracheal abnormalities that can be associated with a tracheoesophageal fistula</td>
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<tr>
<td>Type II</td>
<td>Extrinsic defects or anomalies, such as a vascular ring causing undue pressure on the trachea</td>
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<tr>
<td>Type III</td>
<td>Acquired tracheomalacia that occurs with prolonged intubation or chronic tracheal infections</td>
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Pediatric tracheomalacia

- Congenital TM
  - most common congenital anomaly of the trachea, more in premature
  - Most common associated disease → tracheoesophageal fistula → faulty division of the foregut into the trachea and esophagus
  - Primary TM is a self limiting disease

- Normal infants (idiopathic or primary TM proper)
- Prematurity
- Pulsatile collapse with normal innominate artery
- Congenital abnormalities of the cartilage
  - Dyschondroplasia/chondromalacia/chondrodysplasia
  - Polychondritis
  - Ehlers-Danlos syndrome
- Congenital syndromes associated with TM/TBM
  - Mucopolysaccharidosis
    - Hurler syndrome
    - Hunter syndrome
  - CHARGE syndrome
  - VATER anomaly
  - Trisomy 9
  - Trisomy 21
  - Atelosteogenesis type 1
  - Antley-Bixler syndrome
  - 11p13 deletion
  - 22q11 deletion
  - 18–22 translocation
  - Hallermann-Streiff syndrome
  - Pfeiffer syndrome
  - Blackfan-Diamond syndrome
  - Williams-Campbell syndrome
  - Kniest dysplasia
  - DiGeorge syndrome
  - Deletion of 12 q
  - Larsen syndrome and Larsen-like syndromes
  - Brachmann-de Lange syndrome
  - Camptomelic dysplasia
  - Pierre-Robin syndrome
- Congenital anomalies associated with TM/TBM
  - Tracheoesophageal fistula
  - EA with or without laryngeal cleft
  - Bronchopulmonary dysplasia
Pediatric tracheomalacia

- Acquired TM
  - More common
  - Male predominance
- Associated conditions
  - Cardiovascular: 20~58%
  - Bronchopulmonary: 52%
  - GE reflex: 50~78%
  - Neurologic impairment
  - Development delay
Incidence and nature history

- 1/1445 infants
- 30% in children ≤ 3 y/o undergoing bronchoscopy for respiratory distress.
- Primary TM is a self limiting disease
- Infants outgrow the condition by 2 y/o
- The mortality rate from severe TM → 80%
Symptoms

- **Mild**: respiratory difficulties associated with infectious process
- **Moderate**: stridor, wheezing, recurrent infection and cyanosis
- **Severe**: marked sputum retention, reflex apnea and cardiac arrest

### Table 2—Symptoms of Pediatric Tracheomalacia

<table>
<thead>
<tr>
<th>Symptom</th>
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<tbody>
<tr>
<td>Stridor</td>
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<tr>
<td>Barking cough</td>
</tr>
<tr>
<td>Respiratory distress</td>
</tr>
<tr>
<td>Wheeze</td>
</tr>
<tr>
<td>Anoxic spells</td>
</tr>
<tr>
<td>Cyanosis</td>
</tr>
<tr>
<td>Bradycardia</td>
</tr>
<tr>
<td>Tachyarrhythmias</td>
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<tr>
<td>Spontaneous hyperextension of the neck</td>
</tr>
<tr>
<td>Prolonged expiratory phase</td>
</tr>
<tr>
<td>Breathholding spells</td>
</tr>
<tr>
<td>Failure to thrive</td>
</tr>
<tr>
<td>Increased work of breathing</td>
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<tr>
<td>Sternal, substernal, and intercostal retractions</td>
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<tr>
<td>Recurrent pulmonary infections</td>
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<tr>
<td>Reflex apnea</td>
</tr>
<tr>
<td>Respiratory arrest</td>
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<tr>
<td>Cardiac arrest</td>
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</tbody>
</table>
Diagnosis (I)

- History and physical examination
- Radiography:
  - Sensitivity: 62%
  - Inspiratory and expiratory views
  - Airway Fluoroscopy
  - Barium Swallow
- Bronchoscopy
  - General anesthesia
  - Decrease 50% diameter
Diagnosis (II)

- **CT scan**
  - High sensitivity
  - Dynamic CT
  - Multidetector helical CT
  - Virtual bronchoscopy

- **MRI scan**
  - Evaluate extrinsic compression
  - Vascular compression syndromes
Treatment (I)

- Conservative therapy
  - Treat respiratory infection
  - Humidified oxygen therapy
  - Pulmonary physiotherapy

- Tracheostomy and mechanical ventilation
  - 12%~62%
  - Elongated tracheostomy tube

- Continuous positive airway pressure (CPAP)
Surgery

- Indication: recurrent pneumonia, intermittent respiratory obstruction, inability to extube the airway, dying spell

- Aortopexy

- External splinting with autologous and prosthetic materials
Treatment (III)-internal tracheal stent

- **Silicone prothesis**
  - Montgomery, 1965
  - Granulation
  - Easy to remove

- **Metal stent**
  - Minimal thickness
  - Easy deployment
  - Granulation
  - Migration
  - Difficult removal
  - Halitosis
Adult TM

- Isolated tracheal enlargement, Mounier-Kuhn, 1932
- Acquired tracheal enlargement, Lemoine, 1949
- Classification by macroscopic finding:
  - Lateral wall narrow → saber-sheath type, fissure shape
  - Anterioposterior wall narrow → crescent type, scabbard shape
  - Circumferential narrowing, combination type
Classification of adult tracheomalacia

- **Primary (congenital)**
  - Polychondritis
  - Idiopathic (Mounier-Kuhn syndrome)

- **Secondary (acquired)**
  - Posttraumatic (postintubation, posttracheotomy, external chest trauma, post-lung transplantation)
  - Emphysema
  - Chronic bronchitis
  - Chronic inflammation (relapsing polychondritis)
  - Chronic external compression of trachea (malignancy, benign tumors, cysts, abscesses, aortic aneurysm)
  - Vascular rings (diagnosed in childhood)
Incidence and symptoms

- Majority of adult TM is acquired, in men > 40 y/o
- 1% in patient with respiratory disease for bronchoscope.
- TM is progressive in some patient
- Symptoms: asymptomatic until infection, dyspnea, cough, sputum production, hemoptysis, wheezing, stridor, syncope
Diagnosis

- **Bronchoscope:**
  - Mild: 50~75%
  - Moderate: >75%
  - Severe: posterior wall touches the anterior wall

- **CT**
- **MRI**
- **Pulmonary function study**
  - not diagnostic
  - Decreased FEV1, low peak flow rate with repeat decrease in flow
Treatment

- Supportive, treat underlying disease
- CPAP
- Tracheostomy
- Stent
  - Metal: flexible bronchoscope,
  - Silicone: rigid bronchoscope, GA
  - External stenting
- Surgery
  - Surgical support ➔ bone graft to posterior wall
  - Resection and reconstruction
  - Surgical placation of posterior wall with mesh
TM Diagnosed by CT or by Bronchoscopy

/ /  
| Asymptomatic | Symptoms c/w TM |
|/ | |
|/ | |
| No Further Intervention | Bronchoscopy and CT if not Previously Performed |
| | |
| / /  
| Evaluate and Treat Co-morbid Pulmonary Conditions |
| | |
| / /  
| Symptoms Controlled | Persistent Symptoms |
| | |
|/ | |
|/ | |
| No Further Intervention | Evaluation of Baseline Functional Status (PFTs, SMWT, QOL) |
| | |
| / /  
| Stenting Trial (Silicone) |
| | |
| / /  
| Symptoms Unchanged Or Worse | Symptoms Improved |
| | |
|/ | |
|/ | |
| Remove Stent | Surgical Candidate? |
| | |
|/ | |
|/ | |
| No Further Intervention | No | Yes |
| | |
| | | |
| | | |
| | | |
| Long-term Stenting | Surgery vs. Stenting |

PFTs, pulmonary function tests
SMWT, six minute walk test
QOL, quality of life survey
References

References

- Masters IB, Chang AB, Patterson L, Wainwright C, Buntain H, Dean BW, Francis PW. Series of laryngomalacia, tracheomalacia, and bronchomalacia disorders and their associations with other conditions in children. Pediatr Pulmonol. 2002 Sep;34(3):189-95