Imaging of Tracheobronchomalacia: Case-based Review

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Relevant Financial Disclosures

None
Learning Objectives

- Normal Airway Anatomy
- Definition of Tracheobronchomalacia (TBM)
- Classification of TBM
- Clinical features of TBM
- Diagnosis and Assessment
  - Bronchoscopy
  - Radiological Imaging
- Causes of TBM
- Treatment of TBM
Normal Tracheal Airway Anatomy

• Tracheal Location:
  • Start: Inferior margin of the cricoid cartilage
  • End: Carina (then branches to the left and right main bronchi)

• Dimensions:
  • Length:
    • 10-12 cm including upper 2-4 cm extrathoracic segment\(^1\)
  • Cross-sectional dimensions (measured 2 cm above the aortic arch) on chest X-ray:\(^2\)
    • Coronal: 13-25 mm in men and 10-21 mm in women
    • Sagittal: 13-27 mm in men and 10-23 mm in women
Figure 1: Layers of the Trachea

- Mucosa
- Submucosa
- Horseshoe cartilage (anterolaterally) or trachealis smooth muscle and fibrous tissue (posteriorly)
- Adventitia

Image taken from Gray, 1918.
Figure 2: Tracheal Anatomical Variation during Respiration

- Normal intrathoracic trachea expands with inspiration and narrows with expiration\(^5\)
- End-inspiratory shape:
  - Round, oval, horseshoe, square, inverted pear shaped\(^6\)
- End-expiratory shape:
  - Flattening of the posterior membranous wall with variable degree of anterior bowing and collapsibility
  - Expected decrease in sagittal diameter of up to 30%\(^7\)

Inspiratory CT scan of 51-year-old male with chronic cough, demonstrates a normal oval shape of the tracheal lumen. Image taken from Boiselle, 2006.\(^4\)
Definition of Tracheobronchomalacia (TBM)

• Malacia = Softening
• Definition
  • Excessive airway collapsibility associated with reduction of cross-sectional area of the trachea and/or bronchi exceeding 50% on expiration
    • Some advocate 70% threshold to avoid overdiagnosis of asymptomatic patients
  • Trachea alone = Tracheomalacia (TM)
  • Trachea and Bronchi = Tracheobronchomalacia (TBM)
    • We will refer to TM and TBM collectively as TBM
• Pathophysiology:
  • Weakening of the walls of the trachea and bronchi
  • Histopathology:⑦
    • Atrophy of longitudinal elastic fibers
    • Fragmentation of tracheal cartilage
    • Increase in membranous tracheal diameter
Classification of TBM

- TBM can be classified based on:
  - Pathogenesis:
    - Primary/Congenital
    - Secondary/Acquired
  - Extent:
    - Diffuse or Focal/Segmental
  - Severity of Collapse
  - Location
  - Shape/morphology
  - Multiparametric Classification
# Adult Multiparametric Classification of TBM (1/2)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Extent</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Primary</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Idiopathic Mounier-Kuhn Syndrome/Tracheobronchomegaly</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Type 1 Tracheomalacia (persisting to adulthood)</strong></td>
<td></td>
</tr>
<tr>
<td>Ehler-Danlos syndrome, polychondritis, congenital syndromes (mucopolysaccharidoses), congenital anomalies (esophageal atresia, tracheoesophageal fistula)</td>
<td></td>
</tr>
<tr>
<td><strong>Acquired</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Iatrogenic</strong></td>
<td></td>
</tr>
<tr>
<td>Prolonged Intubation</td>
<td>Segmental</td>
</tr>
<tr>
<td>Prolonged Tracheostomy</td>
<td>Segmental</td>
</tr>
<tr>
<td>Post-surgical (tracheoesophageal fistula repair, post-thyroidectomy)</td>
<td>Segmental</td>
</tr>
<tr>
<td>Post-radiation</td>
<td>Segmental</td>
</tr>
<tr>
<td>Post-lung Transplantation</td>
<td>Segmental or Diffuse</td>
</tr>
</tbody>
</table>

Adapted from Carden, 2005 and Lee EY, 2009.\(^{5,9}\)
### Adult Multiparametric Classification of TBM (2/2)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Extent</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Acquired (continued)</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Chronic External Compression</strong></td>
<td></td>
</tr>
<tr>
<td>Tumors</td>
<td>Segmental</td>
</tr>
<tr>
<td>Enlarged Adjacent Structures</td>
<td>Segmental</td>
</tr>
<tr>
<td>Goiter, enlarged thymus, enlarged left atrium, aortic aneurysm</td>
<td></td>
</tr>
<tr>
<td>Vascular Rings and Anomalies</td>
<td>Segmental</td>
</tr>
<tr>
<td>Double aortic arch, abnormal origin of brachiocephalic artery, pulmonary sling, aberrant right subclavian, right aortic arch</td>
<td></td>
</tr>
<tr>
<td><strong>Infection/Abscess</strong></td>
<td>Segmental</td>
</tr>
<tr>
<td><strong>Chronic Inflammation</strong></td>
<td>Diffuse</td>
</tr>
<tr>
<td>Relapsing polychondritis</td>
<td></td>
</tr>
<tr>
<td><strong>Emphysema and Chronic Bronchitis</strong></td>
<td>Diffuse</td>
</tr>
<tr>
<td><strong>Chest Trauma</strong></td>
<td>Variable Extent</td>
</tr>
<tr>
<td><strong>Skeletal Abnormalities</strong></td>
<td>Variable Extent</td>
</tr>
<tr>
<td>Pectus, scoliosis</td>
<td></td>
</tr>
</tbody>
</table>

Adapted from Carden, 2005 and Lee EY, 2009.5,9
Symptoms of TBM

• Clinical manifestations: \(^5\)
  • Asymptomatic
  • Dyspnea, cough, sputum production, hemoptysis
  • Symptoms unmasked following exposure to stressors
    • Anesthesia
    • Infection
    • Pneumonia
  • Non-specific symptoms attributable to other obstructive airway diseases.

• TBM may go unrecognized for years and be detected incidentally on CT imaging of the chest\(^{10}\)
Diagnostic Modalities of TBM

• Gold standard
  • Bronchoscopy

• Role of CT
  • Dedicated CT has comparable accuracy to bronchoscopy\textsuperscript{11}
  • Non-invasive, but exposes patients to ionizing radiation
  • Can exclude other causes of symptoms
  • Can assess findings beyond the airways (such as an extrinsic compressive cause) and assess the full extent of disease

• Pulmonary function tests\textsuperscript{12}
  • Can show an obstructive pattern
  • Non-specific

• Chest X-ray\textsuperscript{12}
  • Non-sensitive
Dedicated CT Protocol for TBM

• Dual Phase Scan: Inspiratory and Expiratory Phases
  • 1) End-Inspiratory Acquisition
  • 2) Combined with one of:
    • End-Expiratory CT
      • Patient is asked to breath hold at end-expiration then images are acquired in the z-axis.
    • Dynamic Expiratory CT
      • Patient is asked to exhale during acquisition of the images in the z-axis.
    • Cine CT during coughing
      • Patient takes deep inspiration and then coughs repeatedly for 3 to 5 seconds as images acquired in cine mode, usually at the level of the lower trachea.
Dedicated CT Protocol for TBM

- Dynamic expiratory technique and the cine CT technique require greater patient cooperation and coaching
  - Reported to be more sensitive than an end-expiratory phase\textsuperscript{13}

- Multiplanar reformats (MPR) and 3-D reconstructions can be an adjunct for diagnosis and operative planning\textsuperscript{9}
  - Sagittal and coronal MPR
  - Volume rendering
  - Virtual bronchoscopy
Dedicated CT Protocol for TBM

• Inspiratory Phase:
  • Standard CT chest (contrast negative) technique
  • Standard coverage from thyroid gland to adrenal glands.

• Expiratory phase:
  • Low dose technique is sufficient with 120 or 100 kV and 40 to 80 mA
  • Coverage from cricoid cartilage to mid main bronchi for assessment of entire trachea and main bronchi
  • Radiation can be markedly reduced with larger detector width allowing greater z-axis coverage.
    • 64-row MDCT: Allows for 3.2-4.0 cm coverage
    • 320-row MDCT: Allows for up to 16 cm coverage

• Iodinated contrast material (IV or oral) unnecessary unless extrinsic mass compression suspected
Tracheal Morphology on CT

- **Inspiratory phase:**
  - Normal (most common)
  - Lunate Tracheal Configuration
    - Coronal to sagittal ratio of trachea > 1
    - Rare but specific
  - Saber Sheath Trachea
    - Sagittal to coronal ratio of trachea > 2
    - Non-specific (seen in many forms of obstructive lung disease)
  - Tracheobronchomegaly
    - Trachea, right main and left main bronchus greater than 3 standard deviations from normal (associated with collapsibility)

- **Expiratory phase:**
  - Circumferential collapse
  - Anterior and lateral wall collapse
  - Anterior bowing of the posterior wall of the trachea
    - Frown sign configuration: Posterior wall is excessively bowed anteriorly closely apposing (≤ 8 mm) the anterior wall
      - Posterior wall of the trachea may be completely opposed to the anterior and lateral walls in most severe form
Reporting of TBM on CT

- No universally accepted classification of TBM or its severity
- Detailed descriptive reporting suggested:

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tracheal size in axial plane</td>
<td>AP diameter, width and area</td>
</tr>
<tr>
<td>Degree of collapse</td>
<td>Mild: 50-70%; Moderate: 70-90%; Severe: 90+%</td>
</tr>
<tr>
<td>Tracheal Morphology at end-inspiration and end-expiration</td>
<td>Saber Sheath, Frown Sign Configuration</td>
</tr>
<tr>
<td>Distribution of collapse</td>
<td>Anterolateral vs posterior collapse</td>
</tr>
<tr>
<td>Craniocaudal location and extent</td>
<td>Segmental vs diffuse</td>
</tr>
<tr>
<td></td>
<td>Upper, mid, or lower trachea</td>
</tr>
<tr>
<td></td>
<td>Trachea, bronchi, or both</td>
</tr>
<tr>
<td>Associated features</td>
<td>Tracheal wall thickness</td>
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<tr>
<td></td>
<td>Presence of wall calcifications</td>
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<tr>
<td></td>
<td>Tracheal wall diverticula</td>
</tr>
<tr>
<td>Extrinsic lesions</td>
<td>Chest deformity, congenital anomaly</td>
</tr>
</tbody>
</table>
Figure 3: 71 M with incidentally discovered moderate, diffuse TBM on PET CT

End-expiratory axial attenuation correction CT image shows marked anterior bowing of the posterior tracheal wall with a typical frown configuration during unintended expiration (A). This resolves on end-inspiratory CT (B). The reduction of cross-sectional area between the expiratory and inspiratory phase is 73.2%.
Figure 4: 53 M with chronic cough and diffuse TBM

Axial end-inspiratory CT image (A) at the level of the aortic arch shows that the patient has an abnormal **lunate tracheal** morphology with a cross-sectional area of 1.82 cm². Axial end-expiratory CT image at same level (B) shows collapse of the lateral tracheal walls and cross-sectional area of 0.66 cm² (area reduction of 63.7%).
Axial end-inspiratory CT (C) and end-expiratory CT (D) images at the level of the main bronchi show excessive narrowing of the bronchial lumens upon expiration, compatible with bronchomalacia.
Figure 5: 60 M with symptomatic acquired segmental TM of unknown etiology

End-Inspiration

(A) 3-D volume rendered frontal image

(B) 3-D volume rendered lateral image

End-Expiration

(C) 3-D volume rendered frontal image

(D) 3-D volume rendered lateral image

3-D volume rendered frontal (A) and lateral (B) end-inspiratory images in comparison to frontal (C) and lateral (D) end-expiratory images depict the segmental nature of TBM in this patient (blue arrows). The cause of TBM in this patient was unknown, however, the location suggests a post-intubation injury. The patient subsequently underwent tracheal resection and reconstruction (images not shown).
Causes of TBM: Post-intubation and Post-tracheostomy

• Most common causes of acquired TBM$^{14}$
• Associated with repeated or prolonged tube placement
• Tracheal stenosis location:$^{15}$
  • Endotracheal intubation: Inflatable cuff site
  • Tracheostomy: Stoma site or tip of the tracheostomy tube
• Malacia secondary to pressure necrosis, impairment of blood supply, chronic inflammation, and recurrent infections$^{16}$
• Characteristics:
  • Location is pathognomonic
  • Malacia is segmental, most commonly $\leq 3$ cm in length
  • Focal stenosis may be identified at end-inspiration radiograph or CT
Axial CT images at the level of the subglottic trachea at end-inspiration (A) and at end-expiration (B) show a reduction in cross-sectional area of 66% (from 1.72 to 0.58 cm²). There is also thickening of the lateral walls of the trachea due to inflammation and the development of granulation tissue. The morphology of the trachea in the expiratory phase results from the collapse of the lateral walls and anterior bowing of the posterior membrane of the trachea.
Figure 6: 53 M with symptomatic acquired segmental TM following intubation and tracheostomy

Sagittal CT images at the level of the subglottic trachea at end-expiration (C) and coronal oblique CT image at end expiration (D) show the marked decreased in the subglottic sagittal (arrow) and coronal (arrowhead) diameter of the trachea indicating the segmental/focal extent of malacia.
Causes of TBM: Lung Transplant

- TBM is one of many airway complications following lung transplant
  - Incidence post-transplant complications: 5-15\%\textsuperscript{17}
  - Other complications:
    - Bronchial dehiscence, stenosis, fistula
    - Airway infection
- Cause of TBM is multifactorial\textsuperscript{18}
  - Poor Perfusion
  - Immunosuppression
  - Rejection
  - Inadequate surgical technique
  - Infection
  - Inadequate organ preservation
Causes of TBM: Lung Transplant

- TBM usually seen within 4 months of transplantation\textsuperscript{18}
- Patterns of malacia: \textsuperscript{18}
  - Segmental bronchomalacia
    - Restricted to the site of bronchial anastomosis
      (segmental bronchomalacia)
  - Diffuse bronchomalacia
    - Involvement the bronchi beyond the anastomosis
  - Diffuse TBM
    - Involvement of the entire airway
Causes of TBM: Relapsing Polychondritis

• **Relapsing Polychondritis:**
  • Characterized by recurrent episodes of inflammation cartilage and perichondrium that may affect the trachea

• **CT Findings:**
  • Collapse of cartilage and narrowing of tracheal lumen (fixed versus respiratory variation)
  • Wall thickening and calcification of the anterior and lateral tracheal walls
  • Collapse of anterior and lateral walls
  • Sparing of the posterior wall
Treatment of TBM

• Asymptomatic
  • No treatment

• Symptomatic
  • Continuous positive airway pressure (CPAP)
  • Tracheostomy
  • Stenting
  • Surgical repair

• Management of symptomatic patients depends on pattern of disease: 9
  • Focal: Stenting, resection and reconstruction
  • Diffuse disease: Tracheoplasty
    • Greater benefit in patients with anterior bowing than circumferential collapse
    • In diffuse TBM secondary to COPD, optimization of treatment regimen and supportive therapy should be attempted first
Figure 7: 45 F with type IV Ehler-Danlos syndrome presenting with recurrent cough, dyspnea and pulmonary infections

Axial CT images at the level of the aortic arch shows a horseshoe configuration trachea on end-inspiration (A). Upon end-expiration (B), there evidence of TBM with apposition of the anterior and posterior walls of the trachea.
Figure 7: 45 F with type IV Ehler-Danlos syndrome presenting with recurrent cough, dyspnea and pulmonary infections

Axial CT images at the level of the main bronchi on end-inspiration (C) and end-expiration (D). End-expiration shows apposition of the anterior and posterior walls of the right bronchus (and, to a lesser extent, of the left bronchus).
Figure 7: 45 F with type IV Ehler-Danlos syndrome presenting with recurrent cough, dyspnea and pulmonary infections.

Posteroanterior (E) and lateral (F) radiographs of the chest show a synthetic custom Y-stent extending from the trachea to the main bronchi. The patient continued to experience repeated pulmonary infections.
Figure 7: 45F with type IV Ehler-Danlos syndrome presenting with recurrent cough, dyspnea and pulmonary infections

Patient subsequently underwent tracheobronchoplasty using AlloDerm mesh of the trachea and both main bronchi. Post-tracheobronchoplasty end-inspiratory CT images displayed with lung (G) and soft tissue (H) windows show expected thickening of the posterior wall of trachea due to the mesh (arrow).
Figure 7: 45F with type IV Ehler-Danlos syndrome presenting with recurrent cough, dyspnea and pulmonary infections.

Post-operative end-expiratory CT image (I) at the same level as images (A) and (B). Although there is still anterior bowing of the posterior wall of the trachea on end-expiration, the reduction in cross-sectional area is dramatically improved. The patient was able to enjoy excellent quality of life following the procedure.
Summary

We have discussed:

- Normal airway anatomy
- Definition of tracheobronchomalacia (TBM)
- Classification of TBM
- Clinical features of TBM
- Diagnosis and assessment of TBM
  - Bronchoscopy
  - Radiological imaging
- Causes of TBM
- Treatment of TBM

For further questions, please contact Dr. Karl Sayegh at sayeghkarl@gmail.com.
References