DIFFUSE TRACHEAL ABNORMALITIES
AN OVERLOOKED DIAGNOSIS

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Learning Objectives

1. Illustrate the normal appearance of the trachea on CT and understand how it may change in different conditions.

2. Recognize specific tracheal wall abnormalities as a manifestation of certain nonmalignant diseases.

3. Provide a reasonable differential diagnosis based on the imaging findings and clinical correlation.
The trachea connects the larynx to the primary bronchi and is composed of 16–22 C-shaped anterior cartilaginous rings that offer support during expiration.

Its normal diameters are as follows:
- Coronal (transverse):
  - Men: 13 - 25 mm
  - Women: 10 - 21 mm
- Sagital (anteroposterior):
  - Men: 13 - 27 mm
  - Women: 10 - 23 mm

The anterior and lateral walls of the trachea are supported by bands of hyaline cartilage.

The posterior wall lacks cartilage and is supported by a thin band of smooth muscle (trachealis muscle).
The membrane of the trachea is represented by a thin (1 - 3 mm) soft-tissue stripe, bordered anterolaterally by mediastinal fat/lung and posteriorly by the esophagus.

Calcifications of the cartilage are a relatively common finding in older patients, particularly females. It tends to be diffuse, along the normal tracheal contour and nonfocal.

During expiration, there is usually an anterior bulging of the posterior (non-cartilaginous) membrane of the intra-thoracic trachea.

A more severe decrease in the anteroposterior diameter can be seen in pathologic conditions (e.g. tracheomalacia).

CT is the noninvasive modality of choice to evaluate the trachea and central airways. It allows the generation of multiplanar reformations, 3D images and virtual bronchoscopic images.
Diffuse Tracheal Abnormalities (overview)

- Change in luminal diameter (without wall thickening):
  - Increase in all diameters
  - Decreased coronal diameter (increased sagital)
  - Decreased sagital diameter (expiration)

- Wall thickening:
  - Sparing of the posterior membrane
  
  
  - YES
  - NO

- Normal Trachea

- Tracheo-bronchomegaly
- "Saber-sheath" (COPD)
- Tracheomalacia
- Tracheobronchopathia osteochondroplastica
- Polychondritis
- Amyloidosis
- Others
Tracheobronchomegaly (Mounier-Kuhn syndrome)

Rare congenital disorder. Most commonly affects men < 50 years old.

Enlargement of the trachea (> 3 cm) and bronchi due to an absence/atrophy of the elastic fibers and smooth muscle within their walls.

These changes lead to the build up of excess mucus in the bronchi, resulting in chronic productive cough and recurrent pulmonary infections.

Posterior wall diverticula are also common.
Pulmonary fibrosis (e.g. due to interstitial pneumonia, sarcoidosis or cystic fibrosis) can also lead to tracheobronchomegaly secondary to traction and subsequent dilatation.
Only the intrathoracic portion is affected - diffuse narrowing due to a marked decrease of the coronal (transverse) diameter. This change in morphology resembles the shape of a saber sword sheath.

Relatively common and virtually pathognomonic for chronic obstructive pulmonary disease (COPD).

It is believed to be due to repetitive cartilaginous injury from excessive coughing, and elevated intrathoracic pressure.

Other imaging features related to COPD/smoking can be often appreciated, such as:
- Emphysema (Fig. 1)
- Respiratory bronchiolitis (Fig. 2)
- Lung cancer (Fig. 3)
Tracheomalacia

May be congenital or **acquired**.

Ageing, COPD, prolonged intubation, chronic or recurrent infection, asthma, chronic extrinsic compression.
Tracheomalacia

May be congenital or acquired.

Weakness of the tracheobronchial walls and hypotonia of the myoelastic elements.

Leads to collapse of the intrathoracic trachea due to increased intrathoracic pressure during (forced) expiration.

Inspiration scans may be unremarkable or show a dilated trachea with posterior bowing of the non-cartilaginous wall (due to overcompliance).

On expiration, the posterior membranous trachea bows anteriorly (inverted U-shaped/crescent shaped air column on transverse CT).

Collapse > 50%
Tracheomalacia

May be congenital or **acquired**.

**Weakness** of the tracheobronchial walls and **hypotonia** of the myoelastic elements.

Leads to **collapse of the intrathoracic trachea** due to increased intrathoracic pressure during (forced) **expiration**.

**Inspiration** scans may be unremarkable or show a **dilated trachea with posterior bowing of the non-cartilaginous wall** (due to overcompliance).

“**Frown**” sign

Expiration
Tracheobronchopathia Osteochondroplastica

Rare idiopathic nonneoplastic condition. More typical in males, in the 5th - 6th decades.

Usually asymptomatic. May present with cough, dyspnea on exertion, wheezing or recurrent respiratory infection. Hemoptysis may also occur due to ulceration of a nodule or acute infection.

Characterized by the development of osseous and/or small cartilaginous nodules (1 - 8 mm) in the trachea (often protruding into its lumen) and bronchial walls. They may be either focal or diffuse, but more irregular than that seen with senile cartilage calcification.

These nodules arise from the cartilage, hence the sparing of the posterior (non-cartilaginous) membrane of the trachea - important distinction from other conditions such as amyloidosis.
Relapsing Polychondritis

Rare **multisystemic** autoimmune disease characterized by recurrent **inflammation of cartilaginous structures** in the body.

The upper airways are affected in greater than 50% of patients.

Both intra and extrathoracic portions of the trachea are usually involved.

Common CT findings include:
- **smooth** anterolateral wall hyperdense **thickening** sparing the posterior membrane;
- luminal narrowing;
- downstream lung parenchymal ventilation abnormalities;
- **calcifications** may occur within the thickened wall.

Most patients will develop symptoms related to stenosis of the tracheal lumen (tenderness, hoarseness, dyspnea and strydor/wheeze).
Amyloidosis

Extracellular deposition of insoluble proteinaceous (amyloid) fibrils. Various subtypes (including primary and secondary). Common **multisystemic involvement**, including the tracheobronchial tree in rare cases.

Deposits are **submucosal** and generally involve the entire trachea. Bronchi may also be affected.

There is usually a **concentric thickening** of the tracheal wall, **WITHOUT sparing of the posterior membrane**.

**Calcifications** may also occur and are usually concentric.
Granulomatosis with Polyangiitis (Wegener's)

Idiopathic necrotizing granulomatous vasculitis that affects multiple organs, namely the kidneys and the lungs/respiratory tract and sinuses.

The trachea is involved in 15-25% of patients. Predilection for the subglotic trachea and distal trachea/proximal bronchi.

Main CT features include circumferential thickening and narrowing of the tracheal/bronchial lumen.

Mucosal ulcerations may also occur.

Positive c-ANCA (PR3) in 90% of cases. Levels correlate with disease activity.

Sing/symptoms may include:
- cough;
- haemoptysis;
- sinusitis;
- epistaxis;
- proteinuria and haematuria.
Granulomatosis with Polyangiitis (Wegener's)

**Pulmonary manifestations**
- Nodules + cavitations
- Peripheral wedge-shaped consolidations + surrounding ground-glass opacities

**Upper respiratory tract manifestations**
- Sinonasal mucosal thickening + nasal septum perforation
Upper tracheal and laryngeal involvement is more common than distal central airway involvement.

Idiopathic systemic disease characterized by the formation of noncaseating hyalinizing granulomas.

However, large airway involvement is rare - may result from:

- extrinsic compression by adjacent lymphadenopathy;
- infiltration of the airway walls with noncaseating granulomas.

Upper tracheal and laryngeal involvement is more common than distal central airway involvement.

Calcification and circumferential thickening of the tracheal-bronchial tree.
Pulmonary and mediastinal involvement in 90% of cases.

- Perilymphatic nodularity
- Galaxy sign
- Mediastinal and hylar lymphadenopathy
- Fibro cystic changes

Cases:
- Frank Gaillard rID: 6545
- Bruno Di Muzio rID: 58806
- Mohammadtaghi Niknejad rID: 61590
- Bruno Di Muzio rID: 63669
<table>
<thead>
<tr>
<th>Disease</th>
<th>CT appearance</th>
<th>Tracheal narrowing</th>
<th>Wall thickening</th>
<th>Wall calcification</th>
<th>Useful differentiators</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tracheomegaly (Mounier Kuhn Syndrome)</td>
<td>Tracheal diameter &gt; 3 cm; main bronchial diameter &gt; 2.4 cm; scalloping of tracheal wall and diverticula.</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Only diffuse tracheal abnormality resulting in both anteroposterior and transverse dilatation.</td>
</tr>
<tr>
<td>Tracheomalacia</td>
<td>Severe expiratory collapse of the tracheal lumen; lunate-shaped trachea.</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Tracheal collapse; history of predisposing condition or trauma.</td>
</tr>
</tbody>
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## Summary (2)

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<tr>
<td>Saber-sheath trachea</td>
<td>Decreased coronal diameter with concomitant increase in sagittal diameter</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Characteristic bullet-shaped trachea on axial CT; history or imaging findings suggesting COPD</td>
</tr>
<tr>
<td>Tracheobroncopathia osteochondroplastica</td>
<td>Calcified or ossified nodules in the cartilaginous trachea with sparing of the posterior membrane</td>
<td>Yes</td>
<td>Yes</td>
<td>Always</td>
<td>Nodular calcified tracheal wall thickening sparing the posterior tracheal membrane</td>
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## Summary (3)

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<tr>
<td><strong>Amyloidosis</strong></td>
<td>Calcified or ossified nodular or concentric tracheal wall thickening</td>
<td>Yes</td>
<td>Yes</td>
<td>Often present</td>
<td>Concentric calcified or ossified nodular or concentric wall thickening without posterior sparing; may involve the larynx and upper trachea</td>
</tr>
<tr>
<td><strong>Granulomatosis with Polyangiitis</strong></td>
<td>Circumferential tracheal wall thickening or cartilage defects; subglottic region most often involved</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Subglottic narrowing when focal; history of sinus or renal disease, pulmonary cavitary nodules, or pulmonary hemorrhage</td>
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<tbody>
<tr>
<td>Sarcoidosis</td>
<td>Secondary tracheal narrowing from lymphadenopathy or primary tracheal narrowing from tracheal noncaseating granulomas</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Hilar and mediastinal lymphadenopathy; perilymphatic pulmonary nodules.</td>
</tr>
</tbody>
</table>

Take Home Points

- When reading a CT scan of the chest, the tracheobronchial tree should not be overlooked, with special attention to wall thickness and contour.

- The differential diagnosis for diffuse tracheal abnormalities is relatively short and in most cases includes diseases with multysystemic involvement - look for other organ systems!

- Tracheobronchopatia osteocondroplastica and relapsing polychondritis are the only two diseases sparing the posterior membrane of the trachea.

- CT may be the only method required for the diagnosis of diffuse tracheal abnormalities. However, in some cases, it also serves as a guide for bronchoscopic evaluation and transbronchial biopsy.
References


