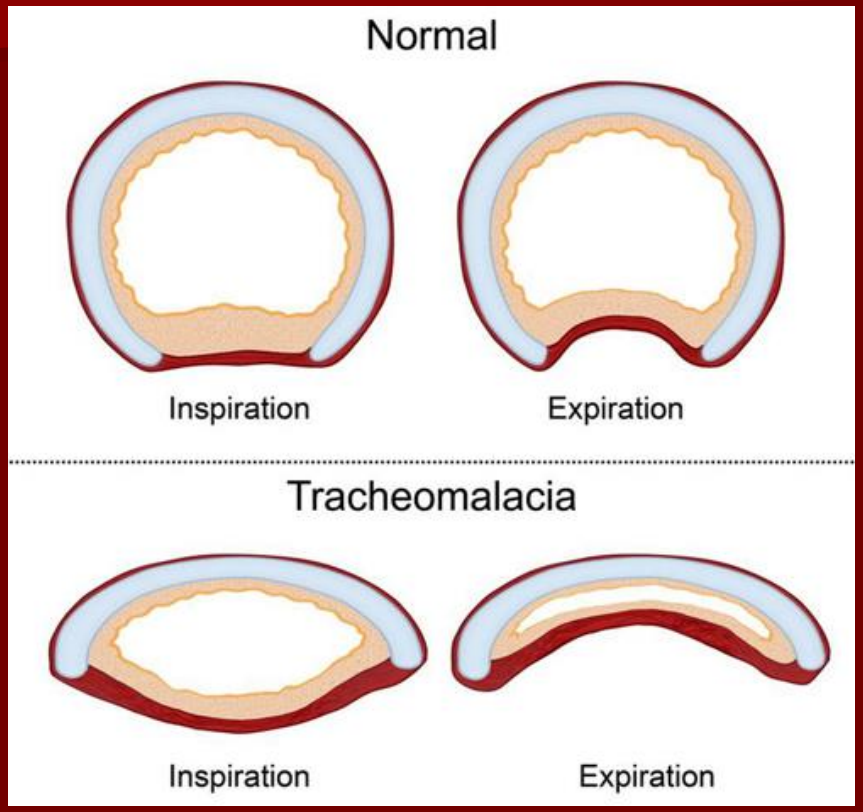
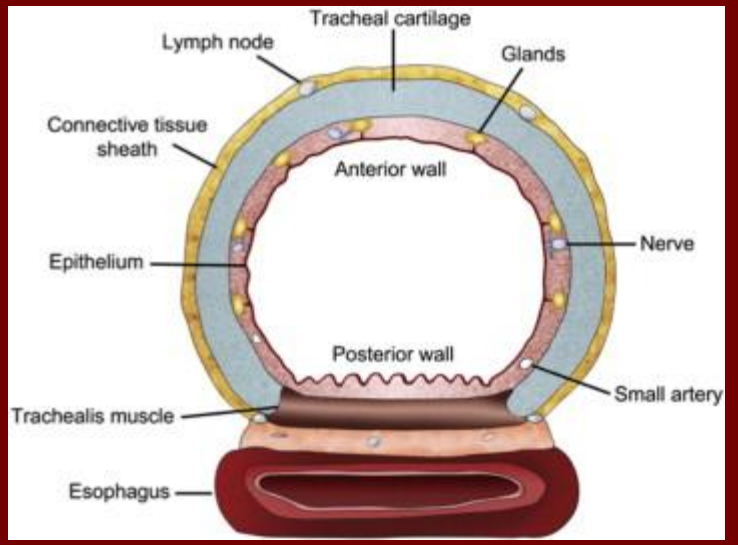
Tracheobronchomalacia

- Weakening of cartilage ± hypotonia of posterior membranous trachea with degeneration and atrophy of longitudinal elastic fibers
- Synonyms
  - Tracheomalacia, bronchomalacia
- Intractable cough, dyspnea, wheezing, recurrent respiratory infections
- Currently regarded as underdiagnosed condition
- Acquired form relatively common in adults, incidence increases with advancing age

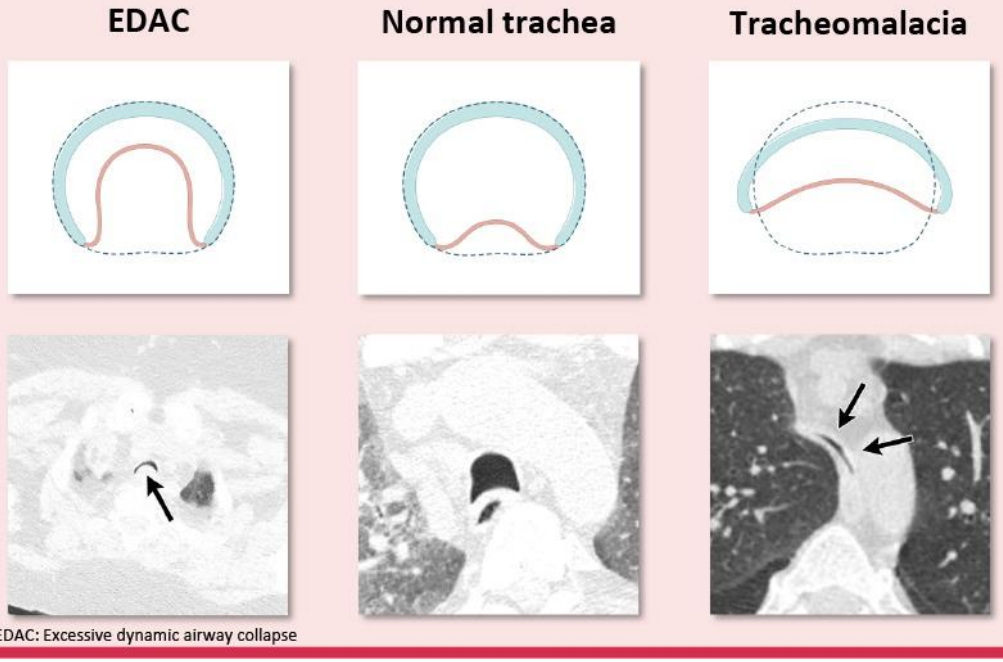
Imaging

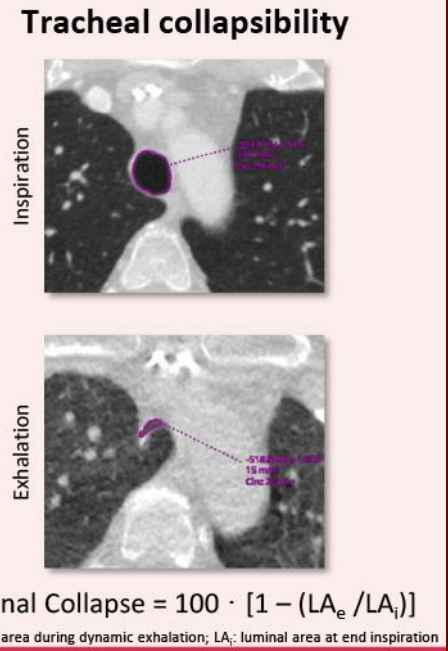
- **Fluoroscopy**
  - Diagnosis based upon > 50% decrease in airway lumen during expiration or coughing
- **CT**
  - Paired end-inspiratory/dynamic expiratory CT
  - Malacia defined as > 70% decrease in cross-sectional area with expiration
  - Most common finding during dynamic expiration: Tracheal collapse with anterior crescentic bowing of posterior membranous trachea (frown sign)
  - Coughing is most sensitive method for eliciting tracheal collapse on CT
  - May reveal source of chronic extrinsic tracheal compression (e.g., thyroid goiter, anomalous vessel)


Normal
Inspiration
Expiration
Tracheomalacia
Inspiration
Expiration


Tracheal cartilage
Lymph node
Glands
Connective tissue sheath
Anterior wall
Nerve
Epithelium
Posterior wall
Small artery
Trachealis muscle
Esophagus

Tracheobronchomalacia and Excessive Dynamic Airway Collapse: Current Concepts and Future Directions





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RadioGraphics

Discussion

Tracheobronchomalacia (TBM) and excessive dynamic airway collapse (EDAC) often present with nonspecific symptoms of airway obstruction including dyspnea, sputum production, wheezing, cough, exercise intolerance, and difficulty weaning from ventilation. These diseases can go undiagnosed and may be misdiagnosed as asthma or chronic obstructive pulmonary disease. Accurate diagnosis is important for therapeutic and prognostic implications.

During expiration, positive pleural pressure causes the posterior tracheal membrane to move anteriorly into the tracheal lumen. In healthy individuals, luminal narrowing will be less than 50% and is termed dynamic airway collapse. In EDAC, the posterior tracheal membrane moves anteriorly, causing greater than 50% reduction in the cross-sectional area, though the anterior cartilage maintains its normal morphology, resulting in a shape like a frown. On inspiration for both dynamic airway collapse and EDAC, the trachea has a range of appearances and may be circular, oval, or horseshoe-like.

TBM is caused by abnormalities of the cartilage rings of the trachea, causing the anterior and lateral walls to deform on expiration. Three narrowed configurations have been described on axial imaging: saber shape, crescentic, and circumferential, each of which can be focal, segmental, or diffuse. Acquired causes include extrinsic compression from vascular structures or the thyroid, as well as cartilage injury from tracheostomy, long-term ventilation, smoking, chronic obstructive pulmonary disease, polychondritis, infection, or chronic inflammation. Congenital TBM is caused by disorders with impaired cartilage maturation such as Ehlers–Danlos syndrome. It may be associated with certain birth defects such as a tracheoesophageal fistula.

In order to diagnose TBM or EDAC, both inspiratory and expiratory images must be obtained, and the AP luminal diameter and/or cross-sectional area is compared. Some studies have found that dynamic expiratory sequences can better identify collapse because of a higher level of intrathoracic pressure during dynamic expiration compared with end expiration, theoretically causing more central airway collapse. As such, paired static inspiratory–expiratory images may underestimate the amount of tracheal collapse and result in more false-negative results.

Bronchoscopy with provocative techniques remains the gold standard for evaluation but is invasive, operator dependent, and not always readily available. CT evaluation can allow quantification of narrowing by using AP diameter and calculation of the luminal area.

Demographics

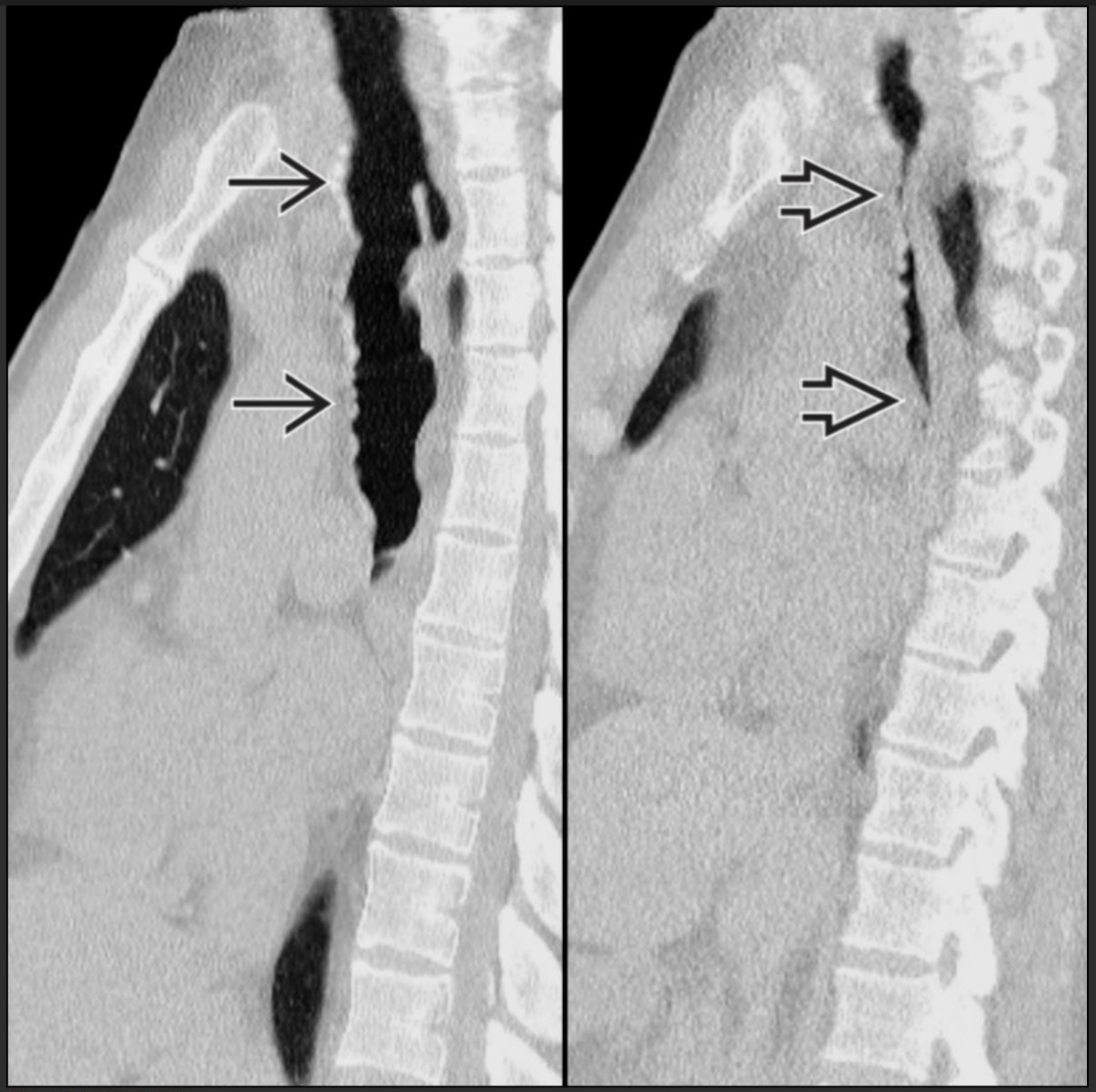
- Age
  - Adults: 2 cases/100,000 population
  - Bimodal age distribution
    - Children: 18 months to 3 years of age
    - Adults: 4th decade of life
- Sex
  - Children: M = F
  - Adults: M > F

CT

- Paired inspiratory-dynamic expiratory CT
  - Inspiratory CT: Comprehensive assessment of airway anatomy, including size, shape, wall thickness, and relationship to adjacent structures
  - Dynamic expiratory CT: Assessment of central airway collapse during 1 helical acquisition
  - Malacia: > 70% decrease in cross-sectional area with expiration
  - Most common finding during dynamic expiration: Tracheal collapse + anterior crescentic bowing of posterior membranous trachea (frown sign)
  - Multiplanar and 3D reformations (including virtual bronchoscopy) not required for diagnosis
    - May reveal craniocaudad extent and morphology of tracheal abnormality
- Cine mode CT during repeated coughing maneuvers
  - Coughing: Most sensitive method for eliciting tracheal collapse
  - Can be performed with electron beam or multidetector CT
  - Requires multiple acquisitions to cover central airways

Etiology

- Increased compliance and excessive collapsibility of airways due to weak cartilaginous rings
- Primary tracheomalacia: Congenital weakness
  - Abnormal cartilaginous matrix (chondromalacia, mucopolysaccharidoses, such as Hurler syndrome)
  - Inadequate maturity of cartilage (e.g., premature infants)
  - Congenital tracheoesophageal fistula or esophageal atresia
  - Mounier-Kuhn syndrome (congenital tracheobronchomegaly)
- Secondary (acquired) tracheobronchomalacia
  - COPD (often correlates with severity of emphysema)
  - Prior (often prolonged) intubation with endotracheal or tracheostomy tube
  - Prior surgery (e.g., lung resection, lung transplantation)
  - Chronic inflammation (e.g., relapsing polychondritis, cystic fibrosis, chronic vaping)
  - Chronic extrinsic compression (e.g., thyroid mass, vascular ring, aneurysm)
  - Radiation therapy
  - Tracheoesophageal fistula
  - Idiopathic



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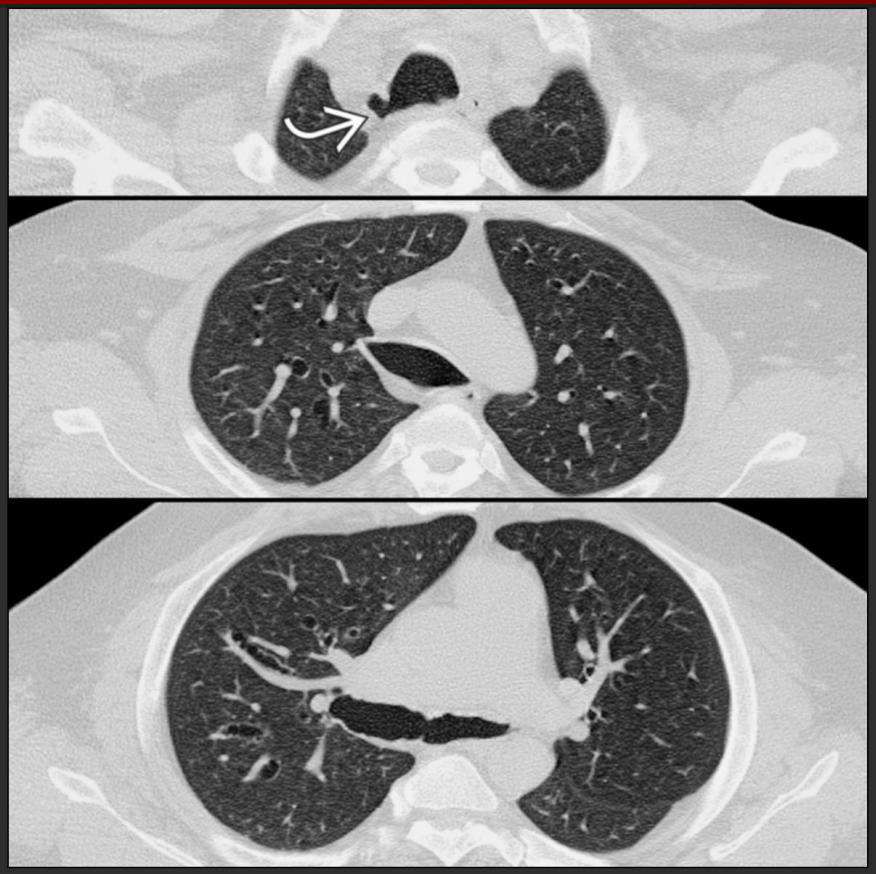
Composite inspiratory (left) and expiratory (right) sagittal NECT of the same patient shows a diffusely dilated trachea ⇨ that exhibits dramatic loss of luminal patency (collapse) during forced expiration ⇨, findings diagnostic of tracheomalacia.



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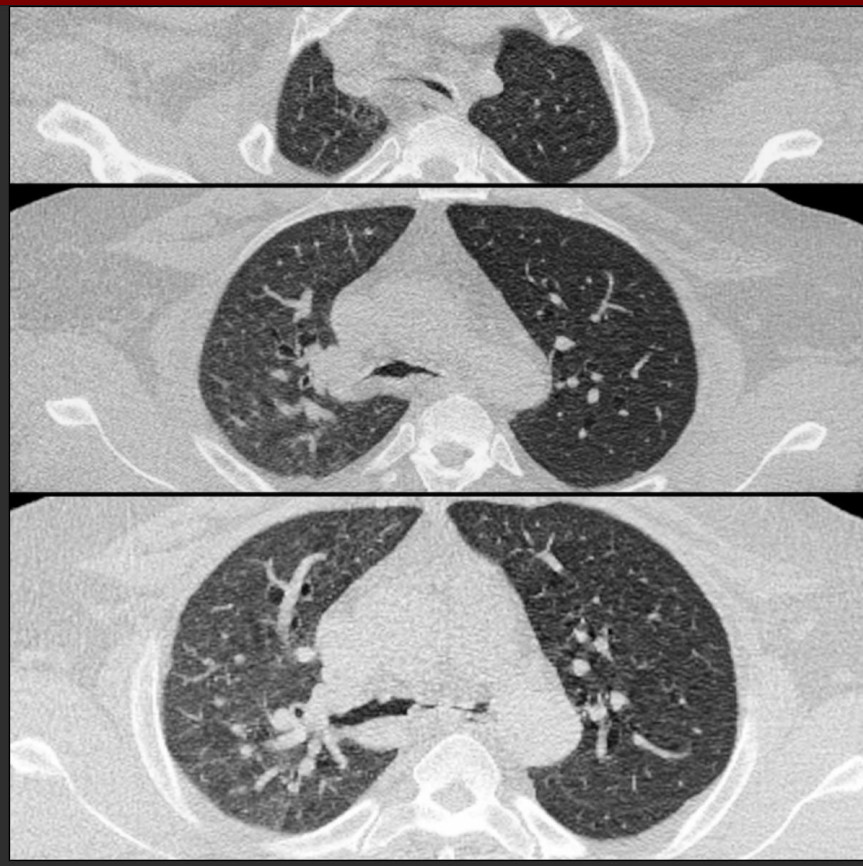
Composite image with axial NECT (lung window) of the same patient obtained at full end-inspiration shows significantly dysmorphic dilated trachea and mainstem bronchi, as well as a tracheal diverticulum ↗.



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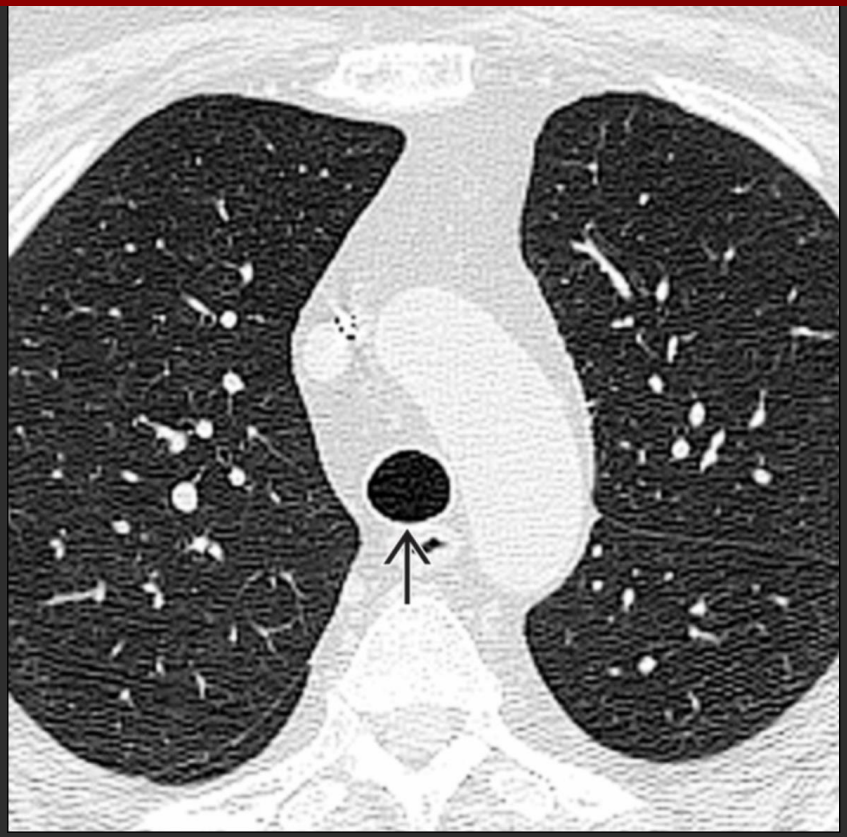
Composite image with axial NECT of the same patient obtained during forced expiration shows severe tracheal and central bronchial airway collapse, confirming the diagnosis of tracheobronchomalacia.



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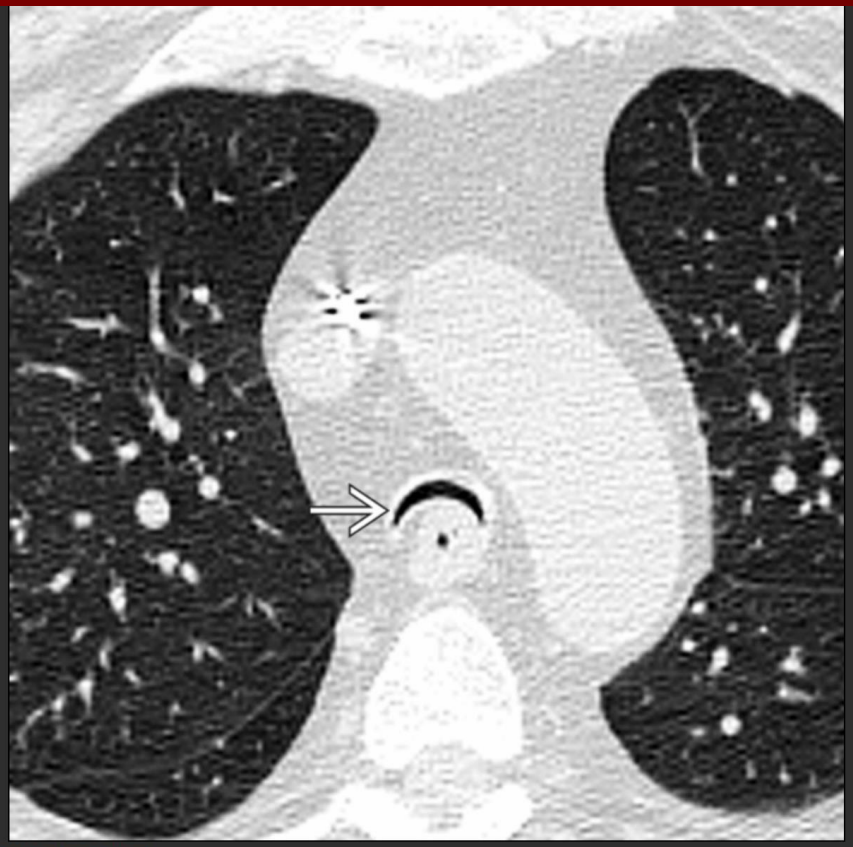
Axial NECT obtained during full inspiration shows a normal tracheal diameter. The posterior wall of the trachea (composed mainly of the trachealis muscle) bows outward ⇨, which indicates that this is an inspiratory image.



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Axial NECT of the same patient obtained during full expiration shows severe tracheal narrowing → in a frown sign configuration, which is highly suggestive of tracheomalacia. Inspiratory CT is insensitive for the detection of tracheomalacia.