Pictorial Essay

Diffuse Abnormalities of the Trachea and Main Bronchi

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Abnormalities of the central airways may produce symptoms such as cough, dyspnea, wheezing, or stridor, but there is frequently a delay in diagnosis. If central airways abnormalities are clinically suspected, further evaluation of tracheobronchial lesions is best performed with CT. A variety of diseases may affect the airways, resulting in either focal or diffuse narrowing or enlargement. This pictorial essay focuses on diffuse abnormalities of the major airways. We recognize that the distinction of diffuse from focal airway abnormalities is somewhat arbitrary and is complicated by the fact that many diseases can cause either focal or diffuse airway narrowing. For purposes of this review, however, we will discuss only those diseases that most commonly result in long-segment or multifocal airway disease.

Diffuse Airway Narrowing

Diffuse narrowing of the trachea or main bronchi may result from relapsing polychondritis, ulcerative colitis, amyloidosis, sarcoidosis, Wegener's granulomatosis, tracheopathia osteochondroplastica, and various infections including papillomatosis. Occasionally, malignancies manifest with diffuse rather than focal airway narrowing.

Relapsing polychondritis is an autoimmune connective tissue disease characterized by polyarthritis, aortitis or arteritis, uveal inflam-

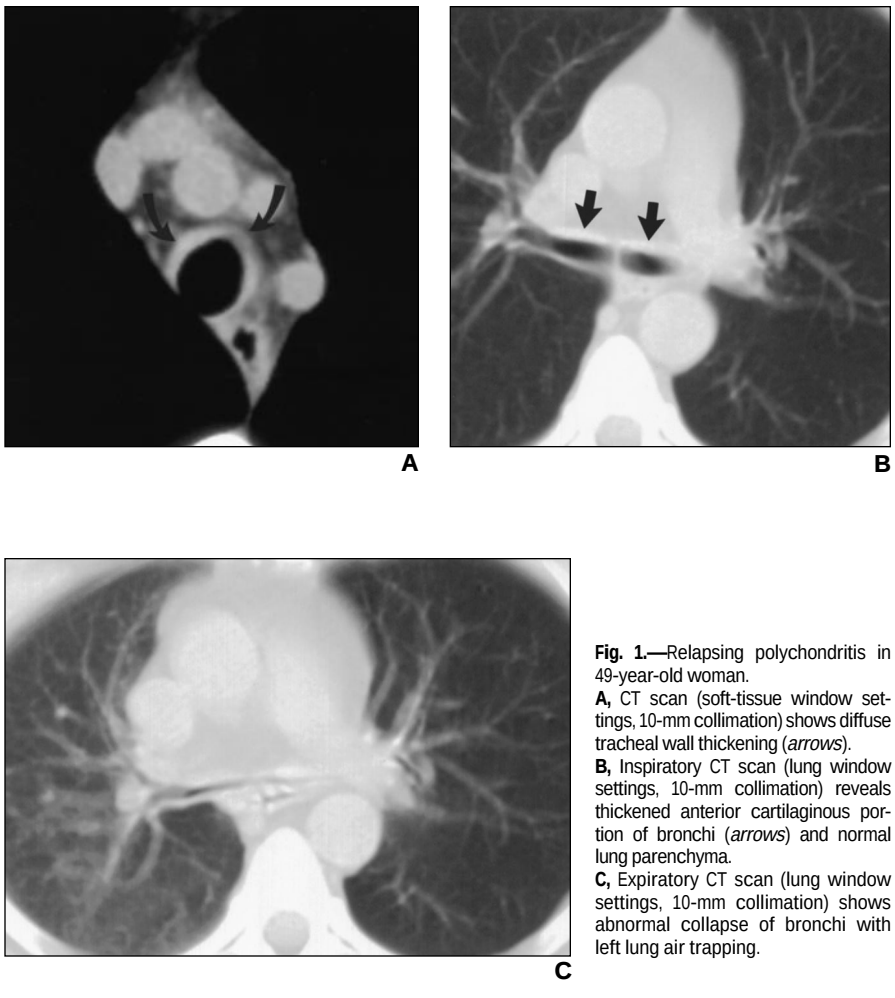

Fig. 1.—Relapsing polychondritis in 49-year-old woman.
A, CT scan (soft-tissue window settings, 10-mm collimation) shows diffuse tracheal wall thickening (*arrows*).
B, Inspiratory CT scan (lung window settings, 10-mm collimation) reveals thickened anterior cartilaginous portion of bronchi (*arrows*) and normal lung parenchyma.
C, Expiratory CT scan (lung window settings, 10-mm collimation) shows abnormal collapse of bronchi with left lung air trapping.

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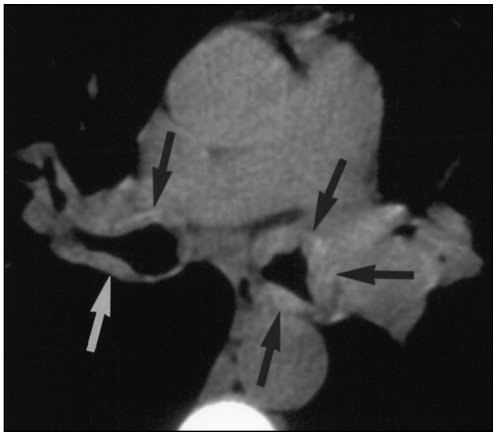

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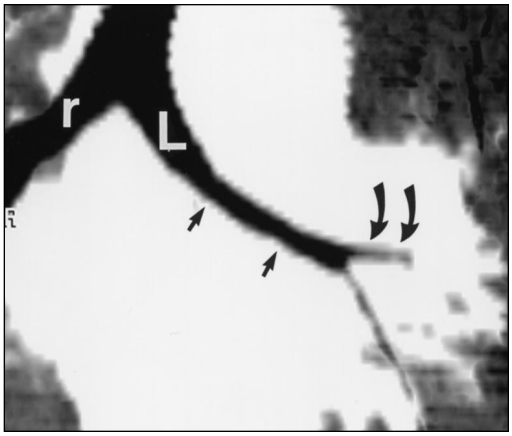

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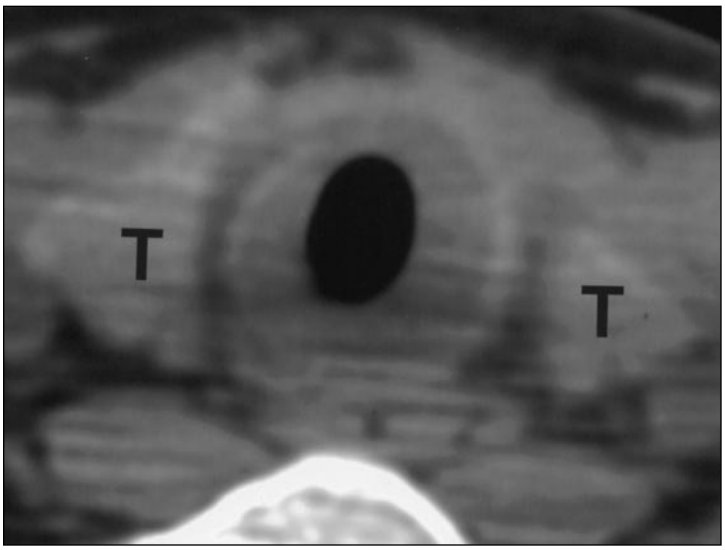
Fig. 2.—Amyloidosis in 27-year-old woman with progressive dyspnea. CT scan (1.5-mm collimation) obtained at level of left main bronchus depicts diffuse circumferential thickening of bronchial walls (*arrows*). Note high-attenuation regions in bronchial wall, likely representing calcification. Transbronchial biopsy revealed amyloidosis.

Fig. 3.—Sarcoidosis in 30-year-old woman. Coronal minimum-intensity-projection image obtained from CT scan (3-mm collimation) reveals diffuse narrowing of left main bronchus (*straight arrows*) and its bifurcating branches, surrounded by conglomerate mediastinal and left hilar lymphadenopathy. Note occlusion of left upper lobe bronchus (*curved arrows*) by same process. L = left main bronchus, r = right main bronchus.



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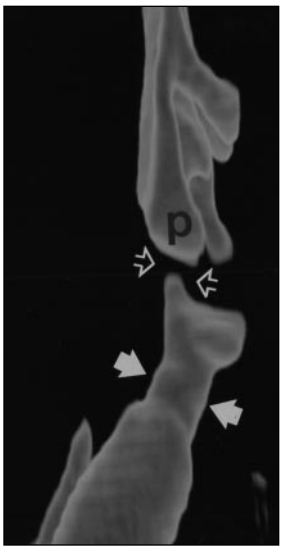

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Fig. 4.—Wegener's granulomatosis in 19-year-old man with dyspnea.
A, CT scan (3-mm collimation) obtained at level of thyroid gland (T) reveals significant tracheal narrowing with diffuse circumferential soft-tissue thickening.
B, Lateral shaded-surface–display image obtained from CT scan (3-mm collimation) reveals diffuse narrowing of subglottic trachea (*solid arrows*) that extends 1.5 cm inferiorly from vocal cords. Note apparent airway discontinuity due to adducted vocal cords (*open arrows*). p = pyriform sinus.

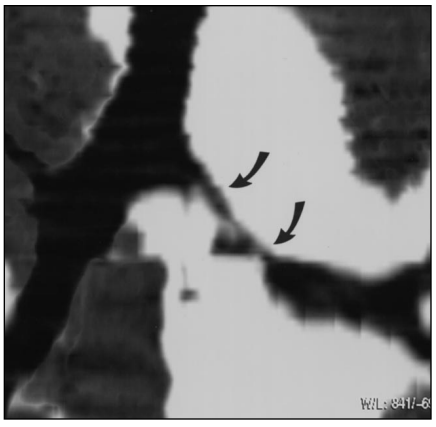

Fig. 5.—Wegener's granulomatosis in 33-year-old woman. Coronal minimum-intensity-projection image obtained from CT scan (3-mm collimation) delineates two focal strictures (*arrows*) in diffusely narrowed left main bronchus.

mation, and recurrent cartilage inflammation, particularly of the ears, nose, larynx, and trachea and mainstem bronchi (up to 70% of affected individuals) [1]. Recurrent pulmonary infection is the most common cause of morbidity and mortality in these patients [2, 3]. Radiologic manifestations include long-segment tracheobronchial strictures, mural thickening, and calcification (Fig. 1A). Expiratory collapse of the affected airway may be revealed on fluoroscopy or dynamic CT (Figs. 1B and 1C). Similar findings are rarely seen in patients with ulcerative colitis [4].

Tracheobronchial amyloidosis most commonly manifests as diffuse mural thickening and luminal narrowing. Less commonly, solitary or multiple mural nodules are seen [5]. Mural calcification may be evident on CT (Fig. 2).

Endobronchial granulomata in patients with sarcoidosis may rarely (1% of cases) result in significant airway narrowing and lobar atelectasis [6] (Fig. 3).

Wegener's granulomatosis produces airway abnormalities in 59% of patients as seen at bronchoscopy. Tracheal strictures affect 7% of patients [7]. CT depicts focal or diffuse wall thickening and airway narrowing (Figs. 4 and 5). The cartilaginous tracheal rings may calcify [2]. When evaluating tracheal stenosis due to Wegener's granulomatosis, the larynx should be included because focal stenosis from Wegener's granulomatosis most commonly affects the subglottic trachea.

Tracheopathia osteochondroplastica is a rare idiopathic and usually asymptomatic disorder of older men; this disorder is characterized by multiple osteocartilaginous masses adjacent to

the tracheal rings of the inner anterolateral wall of the trachea. Tracheopathia osteochondroplastica is distinguished from tracheobronchial amyloidosis or relapsing polychondritis because it does not involve the posterior membranous portion of the trachea [1, 2]. Radiologically, focal tracheal thickening, calcification of the tracheal rings, multiple calcified tracheal nodules, and long-segment tracheal narrowing are typically seen (Fig. 6).

Tracheobronchial papillomatosis is caused by the human papillomavirus, usually acquired at birth from an infected mother. Dissemination of upper airway and laryngeal lesions occurs in 5% of patients and results in multiple nodules projecting into the airways. The papillomas are benign but may undergo malignant transformation to squamous cell carcinoma [8]. Chest radiography or CT may reveal intraluminal masses, parenchymal nodules (after distal airway dissemination), air-filled cysts (pneumatoceles), or thick-walled cavities (Fig. 7).

Other infections such as tuberculosis, coccidioidomycosis, histoplasmosis (Fig. 8), mucormycosis, and *Klebsiella rhinoscleromatis* can lead to airway narrowing that is more commonly focal than diffuse [7].

An important mimic of diffuse tracheal narrowing is the saber-sheath trachea deformity. This entity is a pathognomonic finding in patients with chronic obstructive pulmonary disease. The saber-sheath appearance is found when mechanical forces of hyperinflated lungs cause the coronal diameter of the intrathoracic trachea to narrow and the sagittal diameter to elongate so that the sagittal-to-coronal diame-

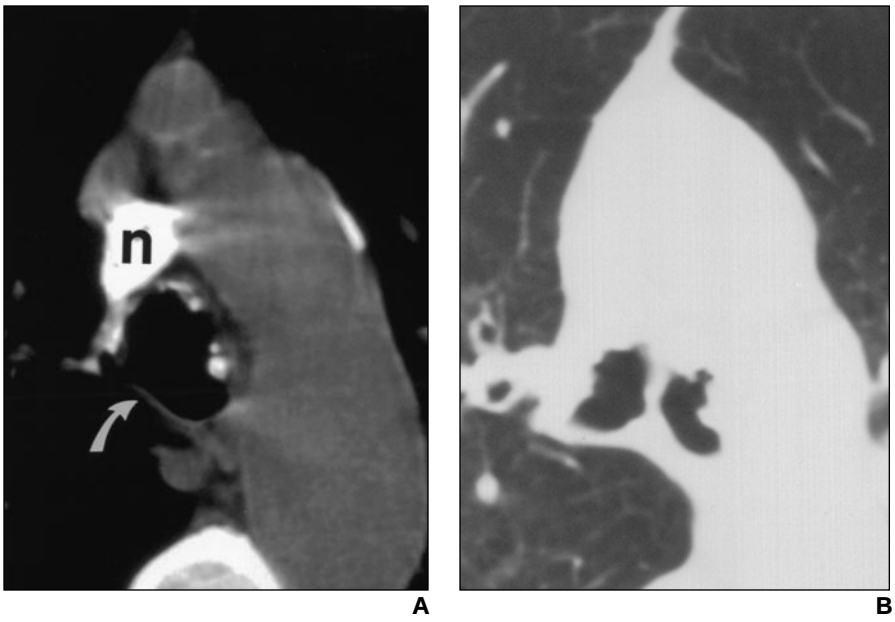

Fig. 6.—Tracheopathia osteochondroplastica in 75-year-old asymptomatic man. (Courtesy of CA Meyer, Indianapolis, IN)
A, CT scan (10-mm collimation) obtained at level of transverse aorta reveals multiple calcified nodules arising from inner anterolateral wall of trachea. Posterior membranous portion of trachea (*arrow*) is spared. Note lymph node (n) is calcified, which was likely caused by prior histoplasmosis.
B, CT scan (lung window settings) shows diffuse mural nodules involving only cartilaginous portion of bronchi resulting in mild luminal narrowing.

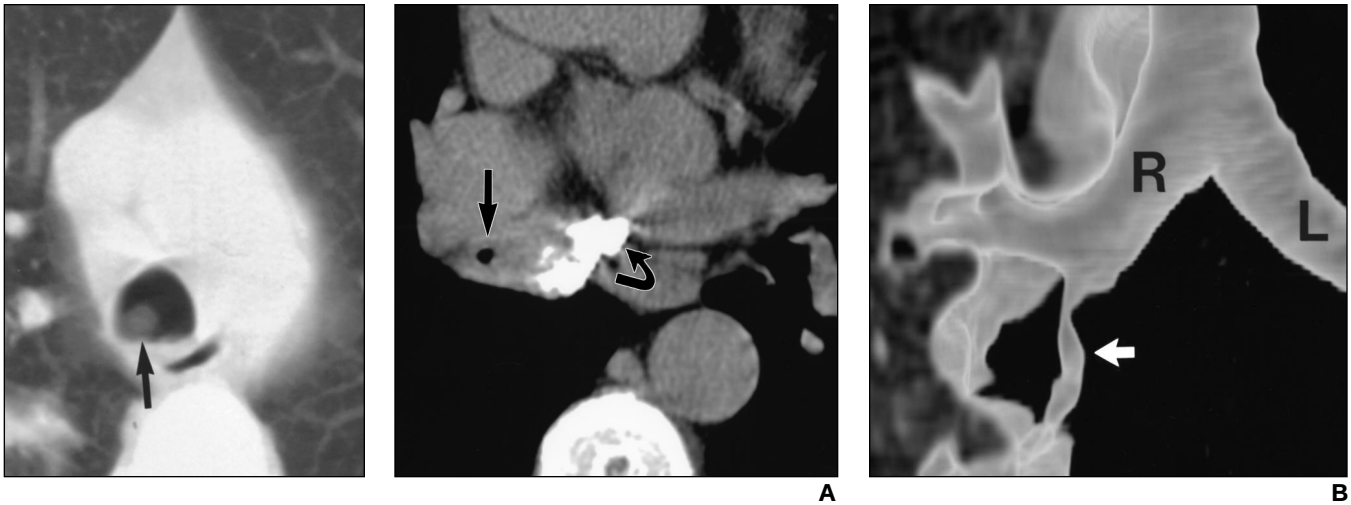

Fig. 7.—Tracheobronchial papillomatosis in 49-year-old woman with laryngeal papillomata. CT scan (10-mm collimation) reveals 6-mm intratracheal nodule consistent with tracheal papilloma (*arrow*). Tracheobronchial tree was diffusely studded with smaller papillomata.

Fig. 8.—Histoplasmosis in 45-year-old man with recurrent pneumonia in right lung.
A, CT scan (3-mm collimation) shows significant narrowing of bronchus intermedius (*straight arrow*), adjacent soft-tissue mass, and heavily calcified subcarinal lymph node (*curved arrow*).
B, Coronal oblique shaded-surface–display image obtained from CT scan (3-mm collimation) shows diffuse narrowing of bronchus intermedius (*arrow*). L = left main bronchus, R = right main bronchus.

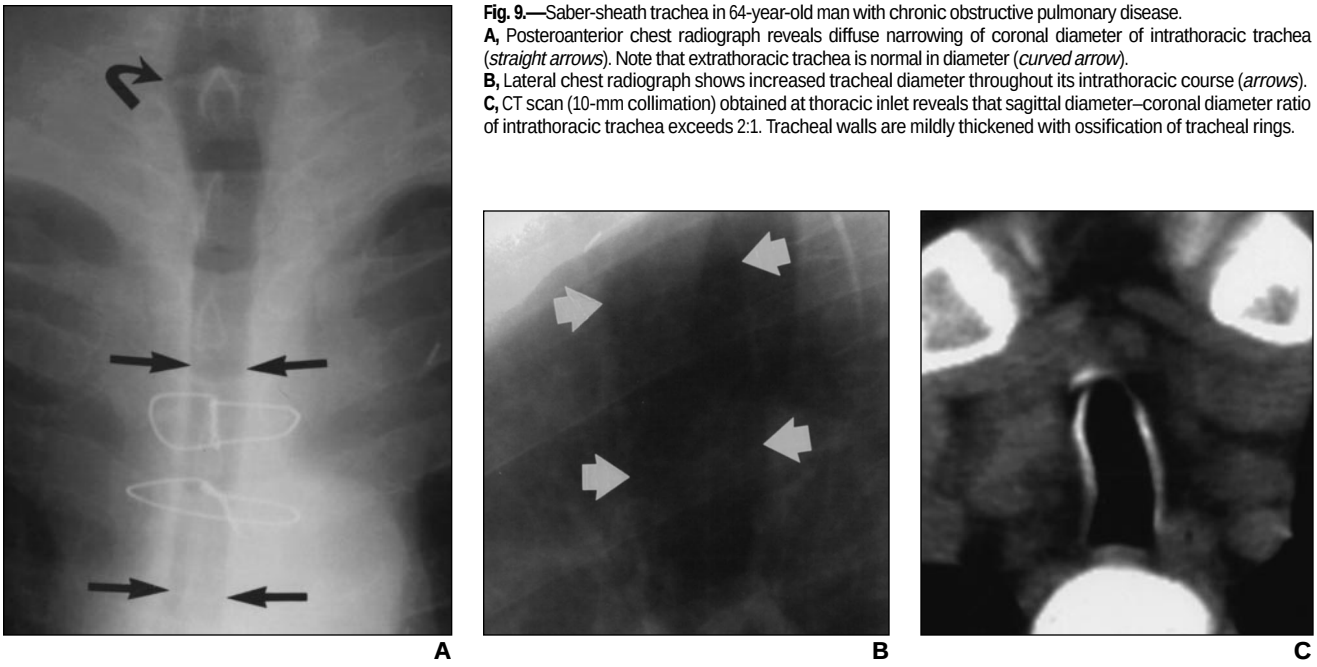

Fig. 9.—Saber-sheath trachea in 64-year-old man with chronic obstructive pulmonary disease.
A, Posteroanterior chest radiograph reveals diffuse narrowing of coronal diameter of intrathoracic trachea (*straight arrows*). Note that extrathoracic trachea is normal in diameter (*curved arrow*).
B, Lateral chest radiograph shows increased tracheal diameter throughout its intrathoracic course (*arrows*).
C, CT scan (10-mm collimation) obtained at thoracic inlet reveals that sagittal diameter–coronal diameter ratio of intrathoracic trachea exceeds 2:1. Tracheal walls are mildly thickened with ossification of tracheal rings.

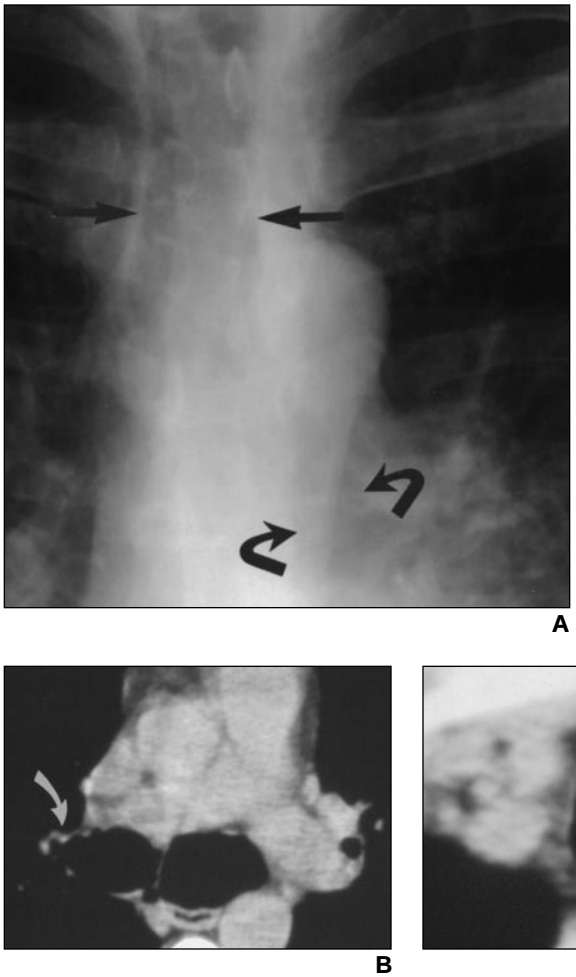

Fig. 10.—Mounier-Kuhn's syndrome in 42-year-old man who presented with recurrent pneumonia.
A, Posteroanterior chest radiograph shows marked tracheal (*straight arrows*) and bronchial enlargement (*curved arrows*).
B, CT scan (8-mm collimation) reveals marked enlargement of main bronchi with corrugated appearance of anterior bronchial wall from mucosal prolapse through tracheal rings (*arrow*).
C, CT scan (8-mm collimation) confirms marked enlargement of trachea (T), measuring 3.9 × 3.5 cm in diameter.

ter ratio exceeds 2:1. The extrathoracic trachea remains normal in configuration. CT may also reveal mild intrathoracic tracheal wall thickening, frequently with ossification of the tracheal rings [2, 7] (Fig. 9).

Diffuse Airway Enlargement (Tracheobronchomegaly)

Tracheobronchomegaly is defined as a transverse and sagittal tracheal diameter exceeding 25 and 27 mm, respectively, or the left and right mainstem bronchi exceeding 18 or 21 mm in diameter, respectively, in men. The respective figures for women are 21, 23, 17.4, and 19.8 mm [2]. Conditions that can result in tracheobronchomegaly include congenital disorders such as Mounier-Kuhn's and Ehlers-Danlos syndromes; diseases that result in severe upper lobe fibrosis such as sarcoidosis and cystic fibrosis; and inflammatory disorders of the airways such as allergic bronchopulmonary aspergillosis.

Mounier-Kuhn's syndrome is thought to result from a congenital deficiency in the internal elastic membrane of the trachea and central bronchi. The disease is diagnosed in most patients during the fourth or fifth decade of life; men are more commonly affected than women. Patients usually present with signs and symptoms related to recurrent infection and bronchiectasis. Radiologic findings of an irregular air column reflect the "corrugated" effect that is pro-

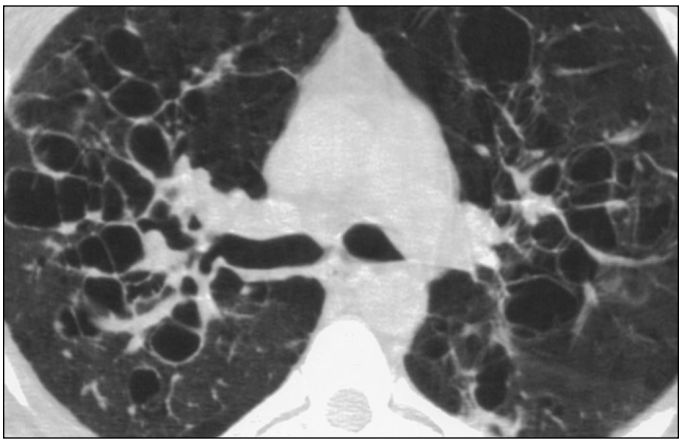

Fig. 11.—Williams-Campbell syndrome in 27-year-old man with recurrent infection. Thin-section CT scan (1.5-mm collimation) shows varicoid and cystic central bronchiectasis associated with normal-caliber main bronchi.

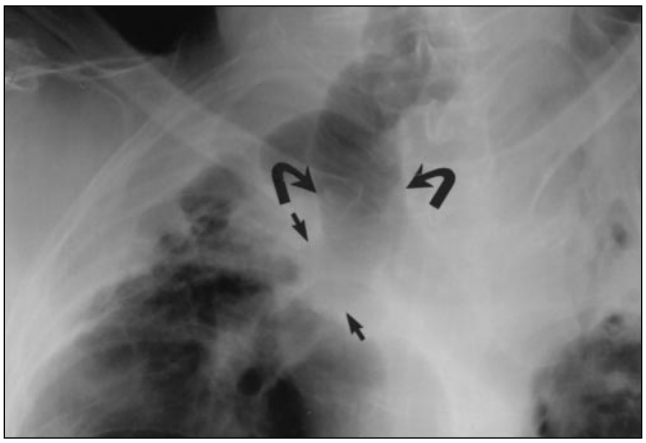

Fig. 12.—Traction tracheomegaly in 63-year-old man with prior history of tuberculosis. Posteroanterior chest radiograph shows severe upper lobe fibrosis resulting in displacement and dilatation of trachea (*curved arrows*) and main bronchi (*straight arrows*).

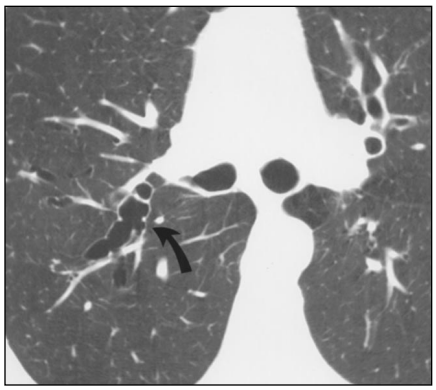

Fig. 13.—Allergic bronchopulmonary aspergillosis in 41-year-old man with severe asthma. Thin-section CT scan (1.5-mm collimation) reveals central varicoid bronchiectasis (*arrow*).

duced when redundant mucosa prolapses through the tracheal rings (Fig. 10). Tracheobronchial diverticula and central bronchiectasis also occur, and CT scans reveal these abnormalities better than chest radiographs. Expiratory studies may reveal collapse of the major airways [2, 7]. Mounier-Kuhn's syndrome may be confused with Williams-Campbell syndrome, a rare form of congenital cystic bronchiectasis that results from a deficiency of cartilage in the fourth- to sixth-order bronchi (Fig. 11). Although both diseases can result in significant bronchiectasis, patients with Williams-Campbell syndrome have normal-caliber trachea and main bronchi [2].

Pulmonary fibrosis in the upper lobes causes retraction of the tracheobronchial walls, leading to tracheomegaly (Fig. 12). Chronic airway inflammation or infection, most commonly caused by smoking, chronic bronchitis, emphysema, and cystic fibrosis [7], may result in tracheobronchomalacia; patients with tracheobronchomalacia present with a diffusely flaccid and dilated airway.

Allergic bronchopulmonary aspergillosis and cystic fibrosis, like Mounier-Kuhn's syndrome, may produce predominant enlargement of the central bronchi or central bronchiectasis. Allergic bronchopulmonary aspergillosis results from airway colonization by *Aspergillus* species and the subsequent immunologic response to aspergillus antigens. It usually occurs in patients with chronic asthma or cystic fibrosis. The diagnosis is suggested by radiologic visualization of central, round, or varicoid bronchiectasis; large mucoid impactions; and fleeting peripheral air-space opacities [2, 9] (Fig. 13).

Summary

Airway lesions are frequently overlooked clinically and radiographically, but if they are suspected, the differential diagnosis for these diseases is limited. The increased use of chest CT to delineate lung abnormalities may lead to the incidental detection of more tracheobronchial abnormalities. Familiarity with the appearances of more typical airway abnormalities should improve diagnosis and patient care.

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