Bronchiectasis is defined as the irreversible dilatation of the cartilage-containing airways or bronchi. Enlargement of the bronchi in acute illness, as can be seen in the setting of infectious pneumonia, is usually reversible and would, therefore, not qualify as bronchiectasis (Fig. 1). Care must be taken to distinguish this large airway dilatation from dilatation of the small airways (bronchioles) that do not contain cartilage.

MECHANISMS OF DEVELOPMENT

Bronchiectasis may result from one of three main mechanisms: bronchial wall injury, bronchial lumen obstruction, and traction from adjacent fibrosis. The latter two mechanisms are usually apparent on imaging, and are suggested by an endobronchial filling defect or adjacent interstitial lung disease. It is when the first group is encountered that the radiologist is faced with a differential diagnosis and a potential diagnostic conundrum. In this article, we aim to cover the CT appearance of bronchiectasis, potential pitfalls, and a diagnostic approach to help narrow the diverse spectrum of conditions that may cause bronchiectasis.

Many conditions may lead to bronchial wall injury and subsequent bronchiectasis. These include infection and recurrent infections, impaired host defense leading to infection, exaggerated immune response, congenital structural defects of the bronchial wall, and extrinsic insults damaging the airway wall (Table 1). These conditions share the common denominator of mucus plugging and superimposed bacterial colonization. The mucus plugging is either a result of abnormal mucus mucousity or abnormal mucus clearance. The toxins released by the bacteria and the cytokines and enzymes released by the surrounding inflammatory cells create a vicious cycle of progressive wall damage, mucus plugging, and increased bacterial proliferation.

Once bronchiectasis begins, therefore, it is sure to progress.

Airway obstruction is most commonly caused by an intraluminal lesion such as carcinoid tumor, inflammatory myofibroblastic tumor, or a fibrous stricture usually from prior granulomatous infection such as histoplasmosis or tuberculosis. The presence of bronchiectasis can be useful in the differential diagnosis of an endoluminal mass, as its presence usually implies a chronic component. Although it may be seen with squamous cell carcinomas arising from papillomas, the presence of bronchiectasis is more suggestive of a slowly growing less malignant lesion, such as a carcinoid tumor (Fig. 2). Distal atelectasis and/or postobstructive pneumonitis are common in these conditions. In the post lobectomy or post-lung transplant patient, granulation tissue at the suture line can occasionally result in an intraluminal occlusion and distal bronchiectasis. In these patients, immediate postoperative detection may allow for bronchoscopic removal of granulation tissue and avoid irreparable damage.

When bronchiectasis is from bronchial wall damage or bronchial obstruction, the bronchial wall becomes thickened because of infiltration by mononuclear cells and fibrosis. In cystic fibrosis (CF), an additional neutrophilic infiltration of the walls and airway lumen can be seen. This mural inflammatory process progressively destroys the elastin, muscle, and cartilage. This leads to airway dilatation.

The dilatation can be classified by its gross appearance as tubular (cylindric), varicose, or cystic (saccular). In the former, the bronchiectasis is manifest as parallel bronchial walls with failure of normal tapering and squared-off ends of the...
bronchus. As the process worsens, the bronchi become serpentine with a beaded appearance. This varicose bronchiectasis serves as an intermediate step before the development of grossly dilated, cystic airways. As the airway dilatation increases, there may be progressive collapse and fibrosis of the distal lung parenchyma.2

Traction bronchiectasis, as its name implies, is caused by retraction of mature fibrosis of the parenchyma around the bronchi. Such bronchiectasis follows the distribution of the underlying fibrosis. The traction bronchiectasis has an upper lobe distribution in cases of radiation fibrosis, sarcoidosis, and sequela of tuberculosis (Table 2) (Fig. 3). In cases of usual interstitial pneumonitis (UIP) (idiopathic pulmonary fibrosis) and fibrosing nonspecific interstitial pneumonitis (NSIP), the traction bronchiectasis tends to be mostly in the periphery and the lung bases (see Fig. 4).3

**CLINICAL PRESENTATION AND DIAGNOSIS**

Symptoms can be very nonspecific. Mild disease may manifest with a mild cough or minimal

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Bronchiectasis caused by airway wall damage</th>
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<tbody>
<tr>
<td>Mechanism</td>
<td>Disease</td>
</tr>
<tr>
<td>Congenital structural defect</td>
<td>Mounier-Kuhn syndrome</td>
</tr>
<tr>
<td></td>
<td>William-Campbell syndrome</td>
</tr>
<tr>
<td>Infection</td>
<td>Pertussis (whooping cough)</td>
</tr>
<tr>
<td></td>
<td>Tuberculosis</td>
</tr>
<tr>
<td></td>
<td>Atypical mycobacterium</td>
</tr>
<tr>
<td>Impaired immune response</td>
<td>Cystic fibrosis</td>
</tr>
<tr>
<td>Abnormal mucociliary clearance</td>
<td>Primary ciliary dyskinesia</td>
</tr>
<tr>
<td>Decreased systemic immunity</td>
<td>Hypogammaglobulinemia</td>
</tr>
<tr>
<td></td>
<td>Lung and bone transplantation</td>
</tr>
<tr>
<td>Exaggerated immune response</td>
<td>Allergic bronchopulmonary aspergillosis</td>
</tr>
<tr>
<td></td>
<td>Inflammatory bowel disease</td>
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<tr>
<td></td>
<td>c-ANCA-positive vasculitis</td>
</tr>
<tr>
<td></td>
<td>Rheumatoid arthritis</td>
</tr>
<tr>
<td>Inhalational injury</td>
<td>Smoke and gaseous toxins</td>
</tr>
<tr>
<td></td>
<td>Chronic gastroesophageal reflux and aspiration</td>
</tr>
</tbody>
</table>
dyspnea. As it becomes more severe, patients may present with chronic cough, regular and copious sputum production, progressive dyspnea, and repeated pulmonary infections.\(^3\) Digital clubbing, anemia, and weight loss can develop because of chronic hypoxemia and hypercarbia in more severe disease. Severe sinusitis can be seen if the bronchiectasis is due to primary cilia dyskinesia, Kartagener syndrome, cystic fibrosis, and diffuse panbronchiolitis.\(^6\)

Hemoptysis, sometimes life-threatening, is caused by chronic airway inflammation and hypoxemia, which leads to bronchial arterial neovascularization.\(^7,8\) These enlarged bronchial arteries are quite fragile and may rupture with even minimal trauma. Pulmonary hypertension can ensue because of underlying hypoxic vasoconstriction and obstructive endarteritis.\(^9\) The bronchial arteries anastomose with pulmonary arterioles, leading to left-to-right shunting, and if severe enough, can contribute to the pulmonary hypertension.\(^9\)

The severity of airflow limitation is related to the extent and severity of the bronchiectasis. This can be seen in ventilation-perfusion mismatches and retained washout on the ventilation images of ventilation-perfusion scintigraphy. Decreased FEV1 (forced expiratory volume in 1 second) on spirometry can also be seen in the setting of bronchiectasis. Although useful in quantifying the ventilation impairment, spirometry has proven insensitive in detecting early structural damage.

**Table 2**

<table>
<thead>
<tr>
<th>Location</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal</td>
<td>Congenital bronchial atresia</td>
</tr>
<tr>
<td></td>
<td>Foreign body</td>
</tr>
<tr>
<td></td>
<td>Broncholithiasis</td>
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<tr>
<td></td>
<td>Endobronchial neoplasm</td>
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<tr>
<td>Diffuse</td>
<td></td>
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<tr>
<td>Upper lung</td>
<td>Cystic fibrosis</td>
</tr>
<tr>
<td></td>
<td>Sarcoidosis</td>
</tr>
<tr>
<td></td>
<td>Progressive massive fibrosis of pneumoconiosis</td>
</tr>
<tr>
<td></td>
<td>Radiation fibrosis</td>
</tr>
<tr>
<td>Central lung</td>
<td>Allergic bronchopulmonary aspergillosis</td>
</tr>
<tr>
<td></td>
<td>End-stage hypersensitivity pneumonitis (also upper lobes)</td>
</tr>
<tr>
<td></td>
<td>Mounier-Kuhn (also lower lobes if repeated infections)</td>
</tr>
<tr>
<td>Lower lung</td>
<td>Usual interstitial pneumonia (IPF)</td>
</tr>
<tr>
<td></td>
<td>Nonspecific interstitial pneumonitis</td>
</tr>
<tr>
<td></td>
<td>Hypogammaglobulinemia</td>
</tr>
<tr>
<td></td>
<td>Lung and bone transplantation</td>
</tr>
<tr>
<td></td>
<td>Chronic aspiration</td>
</tr>
<tr>
<td></td>
<td>Idiopathic</td>
</tr>
<tr>
<td>Right middle lobe and lingula</td>
<td>Atypical mycobacterial infection</td>
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<tr>
<td></td>
<td>Immotile cilia syndrome (PCD) (also lower lobes)</td>
</tr>
</tbody>
</table>
Because of the insensitivity of the other techniques and the high-spatial resolution of CT, high-resolution computed tomography (HRCT) has become the favored diagnostic tool for detection of bronchiectasis. Rapid disease progression has a poorer prognosis and has been shown to be associated with increased wall thickening, colonization by *Pseudomonas aeruginosa*, and high concentrations of proinflammatory markers in sputum or serum, such as neutrophilic elastase.

Diagnosis is usually suspected by history and confirmed by spirometry and HRCT. Depending on the associated symptoms and age of presentation, the diagnostic workup may include a sweat electrolyte test, serum immunoglobulin levels, or serum *Aspergillus* antibody and precipitin, or even genetic testing. Occasionally electron microscopy of the ciliated cells may be used. Bronchoscopy is diagnostic and sometimes therapeutic. In localized bronchiectasis caused by a foreign body, an endobronchial neoplasm, bronchiolithiasis, and fibrotic stenosis, bronchoscopy may be used to remove the obstructing lesion. Although the bronchiectasis itself will not revert, the clearance of the airway will improve any postobstructive pneumonia and may prevent progression of the airway damage. Bronchoscopy and lavage is helpful in patients with more diffuse bronchiectasis who present with acute

**Fig. 3.** Upper lobe traction bronchiectasis from end-stage sarcoid. Transaxial image at the level of the carina (A) demonstrates tortuous, shortened bronchi amidst upper lobe fibrosis and volume loss. Upper lobe predominance of the bronchiectasis is better appreciated on coronal reconstruction (B).

**Fig. 4.** Lower lobe traction bronchiectasis from usual interstitial pneumonitis. Transaxial image at the level of the superior segmental bronchi (A) demonstrates traction bronchiectasis, within peripheral-dominant honeycombing in this 52-year-old man with idiopathic pulmonary fibrosis. Coronal reformatted image (B) shows that the basilar and peripheral bronchiectasis follows the distribution of the fibrosis.
exacerbation or sudden worsening of underlying symptoms. This is usually a sign of acute airway infection, commonly by *Pseudomonas* and *Staphylococcus*, and in such patients bronchoscopy is helpful in obtaining samples for culture and determining antibiotic sensitivity, when usual measures of sputum sampling are unsuccessful.  

**HIGH-RESOLUTION CT IMAGING TECHNIQUE**

Because of the nonspecific nature of symptoms associated with bronchiectasis, an accurate way of diagnosis is needed. HRCT provides the most accurate, least invasive technique. It enables the assessment of bronchial abnormalities to the level of the secondary pulmonary lobule level. Conventional HRCT is performed by acquiring 1.0- to 1.5-mm thick images, every 10 mm, reconstructed using a high spatial frequency algorithm. Images can be obtained in a spiral or sequential mode. Using conventional technique, visualization of bronchi 1 to 2 mm in diameter and vessels 0.1 to 0.2 mm in diameter may be achieved. The lungs are scanned twice, during suspended end-inspiration and suspended end-exhalation. The latter phase is performed to reveal subtle air trapping.

Multidetector computed tomography (MDCT) scanners allow fast, volumetric data acquisition, creating contiguous thin sections through the lungs with excellent z-axis spatial resolution. Similar scanning technique is used for all multidetector scanners that have 16 detector rows or higher, where a collimation of 0.50 to 1.25 mm is selected. The scans are performed in inspiration and reconstructed as contiguous 1-mm images or 1-mm-thick images with 10-mm reconstruction interval. Additional advantages are improved anatomic matching of airways with regions of air trapping during inspiration and expiration and providing the ability for postprocessing the axial images to create 3-dimensional (3D) assessment of the airways. These postprocessed multiplanar images have become known as volumetric HRCT.

In our center, we use a 16- or 64-row multidetector scanner and image the thorax without use of intravenous contrast during end-inspiration and end-exhalation. Inspiration images are acquired helically and reconstructed as 5 × 5-mm images and 1 × 10-mm images; 1 × 1-mm images are also created in case any multiplanar or 3D imaging is needed. Exhalation images are also obtained helically and reconstructed as 1 × 10 mm.

When radiation dose is a consideration (especially in younger patients), a low-dose technique of 30 mAs is used in the inspiratory phase. A growing number of authors have advocated the use of low-dose techniques for HRCT, but we have not routinely relied on these.

**IMAGING FINDINGS**

The imaging findings on conventional radiography are based on visualizing the bronchial wall thickening. This results in ill-defined perihilar linear densities associated with indistinctness of the margins of the central pulmonary arteries. This appearance simulates interstitial pulmonary edema, but lacks the peripheral Kerley B lines. When the bronchus is seen on end, ill-defined ring shadows can be identified because of bronchial wall thickening. The presence of tram lines or parallel lines along the expected courses of the bronchi indicates more severe bronchiectasis, where the dilated bronchial lumen becomes visible on conventional radiography. Tram lines are best identified in the lower lobes, right middle lobe, and lingula (Fig. 5). Oftentimes, an abnormality is detected but the specific diagnosis of bronchiectasis is not.

Mucus plugging may appear as elongated opacities, which may be sometimes calcified. These tubular opacities can be confused for pulmonary vascular enlargement. In cystic bronchiectasis, air-fluid levels in thick-walled cysts are seen, associated with variable degrees of surrounding consolidations and atelectasis. Based on the cause and extent of bronchiectasis, the overall lung volume may be increased (Fig. 6). CT, especially HRCT, is quite reliable in diagnosing bronchiectasis. On CT a diagnosis of bronchiectasis is made when the internal luminal diameter of one or more bronchi exceeds the diameter of the adjacent artery. Other diagnostic criteria of bronchiectasis are the lack of normal tapering of a bronchus, a visible bronchus abutting the mediastinal pleura, or a visible bronchus within 1 cm of the pleura. Signs of bronchiectasis on CT include the signet ring sign, denoted by the artery simulating a jewel abutting the ring, the thick-walled dilated bronchus on a transaxial view, and the tram-track sign, from parallel, thickened walls of a dilated bronchus (Fig. 7).

In cylindrical bronchiectasis the luminal dilatation is uniform and the wall thickening is smooth. Varicose bronchiectasis denotes a more severe form of disease, where the luminal dilatation is characterized by alternating areas of luminal dilatation and constriction, creating a beaded appearance, and the wall thickening is irregular. In cystic bronchiectasis, the most severe form of bronchiectasis, a dilated, thick-walled bronchus terminates in a thick-walled cyst. Oftentimes, more than...
one type of bronchiectasis can be seen in the same patient (Fig. 8).

Smooth bronchial wall thickening is seen in all cases of bronchiectasis, except those caused by congenital cartilage deficiency (William-Campbell syndrome, Mounier-Kuhn syndrome) or in allergic bronchopulmonary aspergillosis (ABPA). Bronchial wall thickening alone is a nonspecific finding, as it is also seen in asthma and chronic bronchitis.\(^6\)

A potential pitfall in the diagnosis of bronchiectasis is the double image of a vessel created by cardiac or respiratory motion artifact simulating a dilated bronchus. This is most common in the lingula and left lower lobe where the effect of cardiac motion is most prominent (Fig. 9). Caution should be made when diagnosis of bronchial wall thickening is made on HRCT images that have been reconstructed with a very high frequency algorithm.

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**Fig. 5.** Diffuse bronchiectasis on conventional radiography. Diffuse bronchiectasis can obscure the perihilar vascular markings, similar to early interstitial pulmonary edema on a pulmonary artery (PA) radiograph (A). Identifying dilated bronchi on end (short arrow) and tram lines (long arrow) leads to the correct diagnosis of bronchiectasis. In this case of cystic fibrosis in a 42-year-old man, the findings are best seen on the lateral radiograph (B).

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**Fig. 6.** Cystic bronchiectasis on conventional radiography. Cystic changes are seen in both mid and lower lungs (arrows) on the PA chest radiograph (A) in this 61-year-old woman with chronic *Mycobacterium avium* intracellulare/complex (MAC) infection. The lateral (B) radiograph demonstrates an air-fluid level in cystic bronchiectasis of the right middle lobe (arrow).
Such algorithm causes the interstitium to appear very thickened and the prominence of the bronchial walls results in a “pseudo-bronchiectasis” appearance (Fig. 10).

Mucus plugging of the dilated bronchi and bronchioles appear as branching dense tubular structures, coursing parallel to, but thicker in diameter, than the adjacent artery and arteriole. These

Fig. 7. CT findings of diffuse bronchiectasis in a 27-year-old woman with cystic fibrosis. Transaxial images show the dilated bronchus and adjacent arteriole, seen along their short axis, creating the signet ring sign (arrow) (A). When the bronchi are visualized along their course, the lack of normal tapering and smooth bronchial wall thickening can be appreciated (B). In cystic fibrosis the fatty attenuation of the pancreas is indicative of the pancreatic insufficiency (C).

Fig. 8. CT findings of varicose and cystic bronchiectasis. CT images from the same patient as in Fig. 6 show a more severe form of bronchiectasis, where both varicose (long arrow) and cystic (short arrow) bronchiectasis is present (A). In varicose bronchiectasis, the bronchial lumen has a beaded appearance and nodular wall thickening is seen (arrowhead) (B).
plugged bronchi are accompanied by adjacent aggregates of nodules, in a “tree-in-bud” pattern, consistent with mucus-filled bronchiolectasis in the center of the secondary pulmonary lobule (Fig. 11).

Mosaic attenuation and air trapping are common associated findings. This is felt to be because of coexisting constrictive bronchiolitis in patients with bronchiectasis. The air trapping may be more pronounced when the bronchiectatic airway has a component of bronchomalacia (Fig. 12).

**BRONCHIECTASIS BASED ON ETIOLOGY**

A majority of bronchiectasis encountered in clinical practice will be postinfectious in origin. In fact, in two retrospective studies evaluating the etiology of bronchiectasis in large cohorts of patients with bronchiectasis, almost one third of the patients were found to have bronchiectasis from prior infection. Idiopathic bronchiectasis, where despite extensive workup the cause was not found, and bronchiectasis related to abnormal mucociliary clearance (cystic fibrosis and primary ciliary dyskinesia) each accounted for almost a quarter of the cases. ABPA and systemic immunodeficiency, due to congenital and acquired causes, were the next most common causes. Some of the more commonly encountered etiologies are presented in the following sections.

**Traction Bronchiectasis**

Traction bronchiectasis is common in UIP, NSIP, and sarcoid and end-stage hypersensitivity pneumonitis.

The mechanism of bronchiectasis in fibrotic lung is based on both physiology and mechanical forces involved in inspiration. Patients with widespread fibrosis require increased inspiratory work, leading to a more negative pleural pressure and therefore a greater transpulmonary pressure during inspiration. On the other hand, pulmonary fibrosis increases the elastic recoil of the lung, creating even further expansion of the bronchi during inspiration.

In our practice, we find it helpful to compare bronchiectasis to the adjacent fibrosis. When the bronchiectasis is out of proportion with the adjacent fibrosis, then NSIP secondary to collagen vascular disease may be considered. The explanation may stem from bronchial wall injury caused by collagen vascular disease–related inflammation or secondary to the high incidence of chronic aspiration in this subgroup of patients (Fig. 13).
Fig. 12. Bronchomalacia and bronchiectasis: a 68-year-old man with a reported history of asthma nonresponsive to steroid therapy. CT images show a normal caliber of the central bronchi (A), and marked collapse of their lumina when imaged during forced exhalation (B), consistent with bronchomalacia. The distal bronchi are diffusely dilated and have smooth wall thickening, consistent with mild bronchiectasis (C).

Fig. 11. Mucus plugging on CT. On transaxial images, the filling of bronchiectasis by mucus appears as tubular and branching opacities, with club-like, rounded ends (arrow) (A). The mucus-filled smaller branching bronchi and bronchioles appear as tree-in-bud opacities (arrowheads) (B). The mosaic attenuation of the surrounding lung is due to air trapping. In this case, the bronchiectasis was from cystic fibrosis.
Congenital Airway Wall Abnormality

Congenital defects of the cartilage, collagen, or other components of the bronchial wall lead to abnormal physiologic clearing of mucoid excretions, predisposing the bronchial epithelium to repeated infections and a vicious cycle of progressive bronchial dilatation. Structural wall defect is the common feature of Mounier-Kuhn disease or tracheobronchomegaly, William-Campbell syndrome, and congenital bronchial atresia.

TRACHEOBRONCHOMEGALY (MOUNIER-KUHN DISEASE)

Tracheobronchomegaly is an uncommon disease that presents mostly in men, in the fourth and fifth decades. Although believed to be congenital, it may be associated with Ehlers-Danlos syndrome, Marfan syndrome, and generalized elastosis (cutis laxa). Pathological thinning of the muscle, cartilage, and elastic tissue of the airway walls is seen. This results in uniform dilatation of the tracheal and bronchial lumina and increased distensibility of the tracheal and bronchial walls. This tracheobronchomalacia leads to recurrent infections in the dependent lungs.6

The disease involves the entire trachea and bronchi of first to fourth order. On imaging, a tracheal diameter exceeding 3 cm in both coronal and sagittal planes and central bronchiectasis is seen without associated airway wall thickening. More distal bronchiectasis, bronchial wall thickening, occasionally fibrosis, and cystic changes in the lower lobes are common because of sequelae of repeated pneumonia.22,23 The net effect is a progression of the bronchiectasis and lung disease, which, in turn, may result in increased tracheobronchomegaly (Fig. 14).

CONGENITAL BRONCHIAL ATRESIA OR MUCOCELE

A focal area of bronchiectasis surrounded by lucent lung is typical of congenital bronchial atresia or congenital mucocele. This condition is characterized by congenital focal obliteration of the lumen of a segmental bronchus, resulting in focal bronchiectasis and air trapping more distally. The dilated airway is commonly filled by inspissated mucus, which may occasionally calcify. Congenital bronchial atresia is usually focal, and is commonly discovered incidentally, as an ovoid, tubular, or branching density on a chest radiograph. It may be confused with a pulmonary nodule. CT reveals its bronchial, branching nature, and the presence of surrounding and more distal hyperexpanded and hyperlucent lung parenchyma, due to the associated air trapping (Fig. 15).

Conversely, acquired mucocele is caused by focal scarring of a segmental bronchus because of prior granulomatous infection or from an endobronchial lesion. It should be differentiated from congenital bronchial atresia by the absence of air trapping of the distal lung parenchyma.24,25 The presence of an acquired mucocele should prompt further interrogation to exclude the possibility of an endobronchial neoplasm (Fig. 16).

WILLIAMS-CAMPBELL SYNDROME

Williams-Campbell syndrome is a rare disease in which the cartilage of the fourth-, fifth-, and
**Fig. 14.** Mounier-Kuhn Syndrome. Transaxial CT images show tracheomegaly (A), and basilar-predominant varicos and cystic bronchiectasis (B) seen in Mounier-Kuhn syndrome. The coronal reconstructed image demonstrates the typical corrugated appearance of the tracheal wall (C).

**Fig. 15.** Congenital bronchial atresia: a 60-year-old woman with pleuritic chest pain. CT with contrast (A) shows a nodular density in the left lower lobe with surrounding hyperlucency of the lung parenchyma, due to focal air trapping. Coronal reconstruction (B) better demonstrates the tubular nature of that structure, which connects to a dilated bronchus. These features (mucocele with surrounding air trapping) are diagnostic for this condition.
sixth-generation bronchi is defective. The disease may involve the lung focally or diffusely. The congenital form presents in childhood and is commonly associated with congenital heart disease, polysplenia, bronchial isomerism, and situs inversus. The acquired form is likely a sequela of prior adenovirus (measles and pertussis) infections. CT imaging shows cystic bronchiectasis distal to the third-generation bronchi, and inspiratory-expiratory CT imaging reveals ballooning on inspiration and collapse on exhalation.

Acquired Wall Abnormalities

Chronic or past infections, inhalational injury, and cellular infiltration in the setting of graft versus host disease lead to inflammation of the bronchial wall, resulting in structural damage of the bronchial wall. This leads to irreversible bronchial dilatation and increased mucus production, leading to a vicious cycle of inflammation and wall damage.

ATYPICAL OR NONTUBERCULOSIS MYCOBACTERIA

Atypical or nontuberculosis mycobacterial pulmonary infection was previously considered to occur only in adults with chronic lung disease, such as CF, lung cancer, or emphysema; adults with impaired immunity, especially acquired immunodeficiency syndrome (AIDS), or those with thoracic skeletal abnormalities. These organisms, however, especially mycobacterium avium intracellulare-complex (MAC), are increasingly being recognized as the cause of chronic lung infection in adults with normal immunity and no underlying lung disease, especially older women. In immunocompetent people, MAC has three different forms: a fibrocavitary form, a nodular bronchiectatic form, and hypersensitivity pneumonitis.

The fibrocavitary form, similar to postprimary tuberculosis, involves the apices and upper lobes and causes traction bronchiectasis in the affected lung. It occurs mostly in older men with emphysema. The nodular bronchiectasis form represents a slowly progressive disease, often resistant to treatment and more common in older women. On CT and HRCT imaging it appears as multiple clusters of centrilobular micronodules, in a branching or tree-in-bud pattern, aggregating around air or mucus-filled cylindric bronchiectasis and bronchiolectasis. Multifocal consolidations and cavities can occur. There is associated mosaic attenuation and air trapping. The disease has a predilection for the right middle lobe, upper lobes, and lingula, but involvement of other lobes may also be seen. Scarring and traction bronchiectasis of the right middle lobe and lingula are indicative of long-standing disease.

Mucociliary Clearance Abnormalities

The ciliary ladder is responsible for effective clearing of mucoid excretions of the airway epithelium. Abnormalities in the consistency of mucus in

**Fig. 17.** Mycobacterium avium intracellulare/complex (MAC) infection: a 68-year-old Chinese woman who developed mild cough and intermittent hemoptysis after a trip to China, which did not respond to routine antibiotic therapy. CT shows extensive micronodules, mostly in the right middle lobe, right lower lobe, and lingula, which aggregate in a tree-in-bud pattern around the mildly dilated subsegmental bronchi (arrow). Sputum specimen was positive for MAC.
cystic fibrosis, and abnormalities of the structure and function of the cilia of the airway epithelium, as seen in primary ciliary dyskinesia or immotile ciliary syndrome, leads to ineffective mucus clearance and secondary colonization of the airway lumina by bacteria. This chronic infection and repeated bouts of pneumonia lead to bronchiectasis. The bronchiectasis of cystic fibrosis is typically worse in the upper lobes, as opposed to primary and acquired ciliary dyskinesia, such as Young syndrome where bronchiectasis is associated with azospermia, where the bronchiectasis is worse in the dependant or lower lungs.

**CYSTIC FIBROSIS**

CF is an autosomal recessive trait and occurs in approximately 1 in 3000 live births in the United States and Europe. It is caused by a mutation in the CF transmembrane conductance regulator (CFTR). This results in failed secretion of chloride through the CFTR and associated ion channels, leading to dehydration of the endobronchial secretions. This thickened mucus cannot be efficiently cleared by the mucociliary system, leading to obstructed airways and bacterial infection. Colonization and recurrent infection with
Staphylococcus aureus, Haemophilus influenza, and Pseudomonas aeruginosa is common, leading to progression of airway destruction. A poor prognosis is made when atypical mycobacteria or Burkholderia cepacia colonize the dilated airways.6

Although CF is usually diagnosed in childhood, the heterogeneity of severity of disease leads to patients with milder disease, first diagnosed in adulthood. As genetic testing improves and awareness of CF increases, milder forms are increasingly being detected later in adulthood. In fact, at our institution, the oldest first-time diagnosis of CF was in a 72-year-old woman with mild bronchiectasis. Sweat chloride test greater than 40 mmol/L indicates the presence of disease.

CT findings include diffuse cylindric, varicose, or even cystic bronchiectasis, bronchial wall thickening. Extensive mucus plugging of the dilated bronchi and bronchioles manifests as centrilobular nodules and branching densities (see Fig. 7).1 In early or mild forms of CF, these findings may be confined to the right upper lobe.6 In adults with long-standing diffuse disease, the findings are widespread throughout the lungs and the upper lobes may be completely scarred and collapsed with associated traction bronchiectasis. This lobar scarring is due to chronicity of disease where the bronchiectasis has been most severe and present longest. Despite the upper lobe volume loss, the lungs remain hyperinflated.

Typically patients with CF have pancreatic insufficiency and on CT the pancreas has a homogeneous fat attenuation. However, in cases of a milder mutation, which leads to an initial diagnosis later in life, the pancreatic insufficiency is commonly absent (Fig. 19).31

Fig. 20. Kartagener syndrome. Transaxial CT of chest (A) in a 12-year-old boy with Kartagener Syndrome shows dextrocardia and bronchiectasis of the lower lobes and scarring of the left middle lobe caused by more severe bronchiectasis and repeated pneumonia. Nonenhanced CT of the paranasal sinuses (B) show marked mucosal thickening and opacification of the maxillary sinuses. The transaxial nonenhanced CT image of the upper abdomen (C) demonstrates the situs inversus associated with this syndrome.
Primary ciliary dyskinesia (PCD) or immotile ciliary syndrome is caused by a defect in structure and function of the airway cilia. This leads to impaired mucociliary clearance.\(^3\) It is a genetically heterogeneous, autosomal recessive trait with a prevalence of approximately 1 in 15,000 to 1 in 30,000 of live births. Patients present with recurrent infections of the lungs, sinuses, and middle ear. Similar to CF, PCD causes progressive bronchiectasis. Thoracoabdominal asymmetry occurs in approximately 50% of patients. Kartagener syndrome or triad is present in half of the PCD patients. This triad consists of situs inversus, bronchiectasis, and sinusitis (Fig. 20).\(^3\)\(^,\)\(^3\)\(^4\)

PCD tends to be diagnosed relatively late, because of its nonspecific presenting symptoms in children. Clinical suspicion based on a focused history leads to diagnosis. The current diagnostic test of choice is electron microscopic analysis of respiratory cilia in samples of nasal or airway mucosa. This reveals defects in the outer or inner dynein arms of the cilia.\(^3\)

The CT findings of PCD are bronchiectasis of variable severity, associated with tree-in-bud nodules and branching densities because of mucus plugging, and lobar scarring and air trapping (Fig. 21). Bronchiectasis in PCD is predominantly in the lingual and middle and lower lobes of the lung. Isolated upper lobe involvement and isolated peripheral bronchiectasis is very rare.

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**Fig. 21.** Primary ciliary dyskinesia: a 37-year-old woman with primary ciliary dyskinesia and chronic pseudomonas aeruginosa infection being evaluated for bilateral lung transplantation. HRCT of chest (A) shows mixed tubular, varicose, and cystic bronchiectasis. Minimal intensity reformatted images in axial (B) and sagittal (C) planes and volume-rendered image (D) demonstrate the extent of bronchiectasis and the associated mosaic attenuation.
Hyper-Immune Response

Inflammatory bowel disease, rheumatoid arthritis, Sjogren disease, antineutrophilic cytoplasmic antibody (c-ANCA)–positive vasculitis (Wegener disease), and allergic bronchopulmonary aspergillosis all can lead to bronchiectasis, possibly because of inflammation of the airway wall in the setting of a hyperimmune response to internal or external antigens. The chronic inflammation damages the bronchial walls, leading to bronchiectasis.

Allergic Bronchopulmonary Aspergillosis

Allergic bronchopulmonary aspergillosis (ABPA) is due to a hypersensitivity reaction to Aspergillus fumigatus antigens, leading to development of bronchocentric granulomata in the bronchi and bronchioles, associated with mucus impaction. It is most commonly seen in patients with atopic rhinitis, asthma, or CF. Patients present with wheezing, fever, and pleuritic chest pain and may cough up brown mucus plugs. Clinical diagnosis could be made by detecting an elevated serum IgE level, eosinophilia on peripheral blood smears, or positive skin reaction to Aspergillus antigen. Mycelia can occasionally be identified in the exopored mucus plugs.

Chest radiographs and CT show migratory pneumonitis early in the disease. This usually involves the upper lobes. Central and upper lobe bronchiectasis, varicose or cystic subtypes, is best detected by CT, which also helps monitor progression and response to treatment. In more chronic disease, inspissated mucus in the dilated central bronchi created the appearance of the classic finger-in-glove appearance on the chest radiographs (Fig. 22). Atelectasis or hyperinflation of the lung distal to the impacted bronchi can occur.

SUMMARY

Bronchiectasis, or the irreversible dilatation of bronchi, can present with a host of nonspecific...
clinical symptoms, including hemoptysis, cough, and hypoxia. The radiologist, then, can play an important role in its detection and characterization. Bronchiectasis must be differentiated from motion artifact and transient bronchial dilatation in acute lung disease. When diagnosed, a logical approach may allow for proper triage of the patient to prevent progression of disease.

The radiologic approach usually begins with CT, which is fast and accurate. The diagnostic approach should be based on the mechanisms of development of bronchiectasis (bronchial wall damage, endobronchial obstruction, and traction) and the location. Once an endobronchial lesion or adjacent fibrosis is excluded, location of the abnormality can be used to help narrow the differential diagnosis. When the bronchiectasis is upper lobe predominant, CF should first be considered but occasionally MAC infection may present with this finding. When the bronchiectasis is mid-upper lobe, then ABPA or chronic hypersensitivity pneumonitis might lead the list of diagnoses. Lower lobe bronchiectasis is usually the sequela of recurrent infection and conditions that predispose to recurrent infections, including Mounier-Kuhn, hypogammaglobulinemia, PCD, and recurrent infections. By using this approach, the radiologist can remain an integral part of the pulmonary team.

REFERENCES