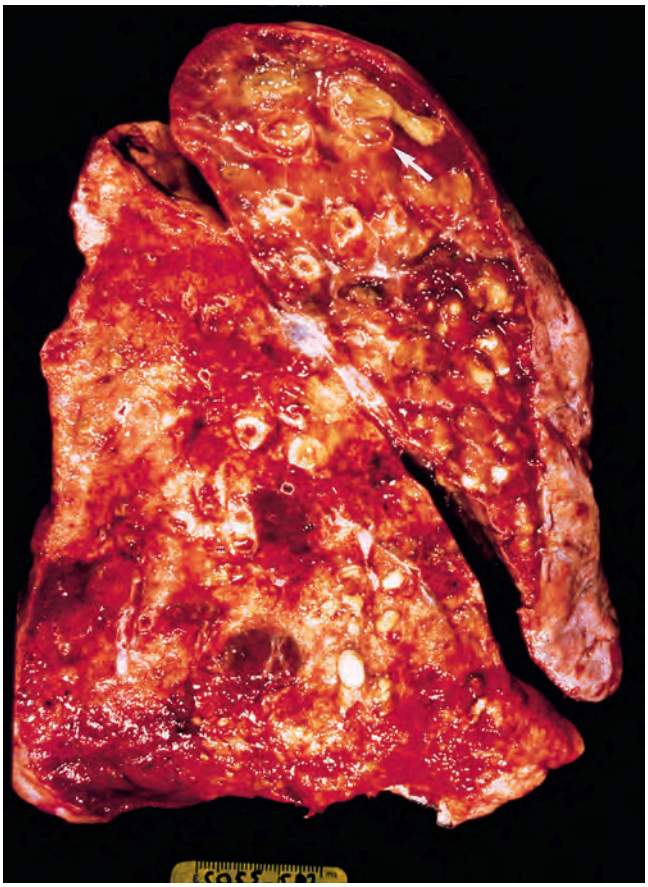

Figure 15-12 Bronchiectasis in a patient with cystic fibrosis, who underwent lung transplantation. Cut surface of lung shows markedly distended peripheral bronchi filled with mucopurulent secretions.

followed almost to the pleural surfaces. By contrast, in the normal lung, the bronchioles cannot be followed by eye beyond a point 2 to 3 cm from the pleural surfaces. On the cut surface of the lung, the dilated bronchi appear cystic and are filled with mucopurulent secretions (Fig. 15-12).

The histologic findings vary with the activity and chronicity of the disease. In the full-blown, active case there is an intense acute and chronic inflammatory exudation within the walls of the bronchi and bronchioles, associated with desquamation of the lining epithelium and extensive areas of ulceration. There may be pseudostratification of the columnar cells or squamous metaplasia of the remaining epithelium. In some instances necrosis destroys the bronchial or bronchiolar walls and forms a lung abscess. Fibrosis of the bronchial and bronchiolar walls and peribronchiolar fibrosis develop in the more chronic cases, leading to varying degrees of subtotal or total obliteration of bronchiolar lumens.

A large variety of bacteria can be found in the usual case of bronchiectasis. These include staphylococci, streptococci, pneumococci, enteric organisms, anaerobic and microaerophilic bacteria, and (particularly in children) *Haemophilus influenzae* and *Pseudomonas aeruginosa*. In allergic bronchopulmonary aspergillosis, a few fungal hyphae can be seen on special stains within the mucoinflammatory contents of the dilated segmental bronchi. In late stages the fungus may infiltrate the bronchial wall.

Clinical Course. Bronchiectasis causes severe, persistent cough; expectoration of foul smelling, sometimes bloody sputum; dyspnea and orthopnea in severe cases; and, on occasion, hemoptysis, which may be massive. Symptoms are often episodic and are precipitated by upper respiratory tract infections or the introduction of new pathogenic agents. Paroxysms of cough are particularly frequent when the patient rises in the morning, as the change in position causes collections of pus and secretions to drain into the bronchi. Obstructive respiratory insufficiency can lead to marked dyspnea and cyanosis. However, due to current treatment with better antibiotics and physical therapy, outcome has improved considerably and life expectancy has almost doubled. Hence cor pulmonale, brain abscesses, and amyloidosis are less frequent complications of bronchiectasis currently than in the past.

Chronic Diffuse Interstitial (Restrictive) Diseases

Restrictive lung disorders occur in two general conditions: (1) *chronic interstitial and infiltrative diseases*, such as pneumoconioses and interstitial fibrosis of unknown etiology; and (2) *chest wall disorders* (e.g., neuromuscular diseases such as poliomyelitis, severe obesity, pleural diseases, and kyphoscoliosis), which are not discussed here.

Chronic interstitial pulmonary diseases are a heterogeneous group of disorders characterized predominantly by inflammation and fibrosis of the pulmonary interstitium. Many of the entities are of unknown cause and pathogenesis, and some have an intra-alveolar as well as an interstitial component. There is frequent overlap in histologic features among the different conditions. These disorders account for about 15% of noninfectious diseases seen by pulmonary physicians.

In general, the clinical and pulmonary functional changes are those of *restrictive lung disease*. Patients have dyspnea, tachypnea, end-inspiratory crackles, and eventual cyanosis, without wheezing or other evidence of airway obstruction. The classic functional abnormalities are reductions in diffusion capacity, lung volume, and lung compliance. Chest radiographs show bilateral lesions that take the form of small nodules, irregular lines, or *ground-glass shadows*, all corresponding to areas of interstitial fibrosis. Eventually, secondary pulmonary hypertension and right-sided heart failure associated with cor pulmonale may result. Although the entities can often be distinguished in the early stages, the advanced forms are hard to differentiate because all result in scarring and gross destruction of the lung, often referred to as *end-stage lung* or *honeycomb lung*. Diffuse restrictive diseases are categorized based on histology and clinical features (Table 15-5).

Fibrosing Diseases

Idiopathic Pulmonary Fibrosis

Idiopathic pulmonary fibrosis (IPF) refers to a clinicopathologic syndrome marked by progressive interstitial pulmonary fibrosis and respiratory failure. In Europe the term *cryptogenic fibrosing alveolitis* is more popular. IPF has