left lobar bronchi branch dichotomously, giving rise to progressively smaller airways. Accompanying the branching airways is the double arterial supply to the lungs, derived from the pulmonary and bronchial arteries.

Progressive branching of the bronchi forms *bronchioles*, which are distinguished from bronchi by the lack of cartilage and submucosal glands within their walls. Further branching of bronchioles leads to the *terminal bronchioles*, which are less than 2 mm in diameter. The part of the lung distal to the terminal bronchiole is called the *acinus*; it is roughly spherical, with a diameter of about 7 mm. An acinus is composed of *respiratory bronchioles* (each of which gives off several alveoli from its sides), *alveolar ducts,* and *alveolar sacs*, the blind ends of the respiratory passages, whose walls are formed entirely of alveoli, which are the site of gas exchange (see Fig. 15-6). A cluster of three to five terminal bronchioles, each with its appended acinus, is referred to as the pulmonary *lobule*.

Except for the vocal cords, which are covered by stratified squamous epithelium, the entire respiratory tree, including the larynx, trachea, and bronchioles, is lined by pseudostratified, tall, columnar, ciliated epithelial cells. The bronchial mucosa also contains a population of neuroendocrine cells that have neurosecretory-type granules and can release a variety of factors, including serotonin, calcitonin, and gastrin-releasing peptide (bombesin). Numerous mucus-secreting goblet cells and submucosal glands are dispersed throughout the walls of the trachea and bronchi (but not the bronchioles).

The microscopic structure of the alveolar walls (or alveolar septa) consists of the following (Fig. 15-1):

- An intertwining network of *anastomosing capillaries* lined with endothelial cells
- *Basement membrane and surrounding interstitial tissue*, which separate the endothelial cells from the alveolar lining epithelial cells. In thin portions of the alveolar septum, the basement membranes of epithelium and endothelium are fused, whereas in thicker portions they are separated by an interstitial space *(pulmonary interstitium)* containing fine elastic fibers, small bundles of collagen, a few fibroblast-like interstitial cells, smooth muscle cells, mast cells, and rare lymphocytes and monocytes.
- *Alveolar epithelium*, a continuous layer of two cell types: flattened, platelike *type I pneumocytes*, covering 95% of the alveolar surface, and rounded *type II pneumocytes*. Type II cells synthesize *surfactant* (which forms a very thin layer over the alveolar cell membranes) and are involved in the repair of alveolar epithelium through their ability to give rise to type I cells.
- *Alveolar macrophages*, loosely attached to the epithelial cells or lying free within the alveolar spaces

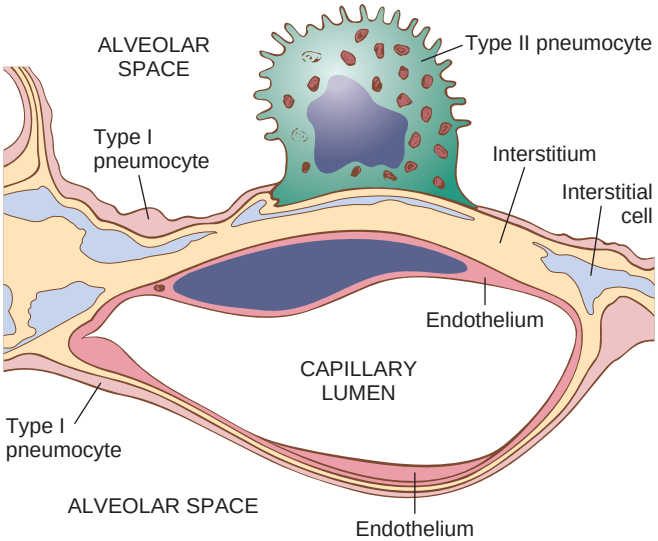

Figure 15-1 Microscopic structure of the alveolar wall. Note that the basement membrane *(yellow)* is thin on one side and widened where it is continuous with the interstitial space. Portions of interstitial cells are shown.

The alveolar walls are perforated by numerous *pores of Kohn*, which permit the passage of bacteria and exudate between adjacent alveoli (see Fig. 15-34*B*).

Congenital Anomalies

Developmental anomalies of the lung are rare; the more common of these include the following:

- *Pulmonary hypoplasia* is the defective development of both lungs (one may be more affected than the other) resulting in decreased weight, volume, and acini for body weight and gestational age. It is caused by abnormalities that compress the lung or impede normal lung expansion in utero, such as congenital diaphragmatic hernia and oligohydramnios. Severe hypoplasia is fatal in the early neonatal period.
- *Foregut cysts* arise from abnormal detachments of primitive foregut and are most often located in the hilum or middle mediastinum. Depending on the wall structure, these cysts are classified as bronchogenic (most common), esophageal, or enteric. A bronchogenic cyst is rarely connected to the tracheobronchial tree. Microscopically, the cyst is lined by ciliated pseudostratified columnar epithelium. The wall contains bronchial glands, cartilage, and smooth muscle. They usually present due to compression of nearby structures or are found incidentally.
- *Pulmonary sequestration* refers to a discrete area of lung tissue that (1) lacks any connection to the airway system and (2) has a abnormal blood supply arising from the aorta or its branches. *Extralobar sequestrations* are external to the lung and most commonly come to attention in infants as mass lesions. They may be associated with other congenital anomalies. *Intralobar sequestrations* occur within the lung. They usually present in older children, often due to recurrent localized infection or bronchiectasis.

Other less common congenital abnormalities include tracheal and bronchial anomalies (atresia, stenosis, tracheoesophageal fistula), vascular anomalies, congenital pulmonary airway malformation and congenital lobar overinflation (emphysema).

Atelectasis (Collapse)

Atelectasis refers either to incomplete expansion of the lungs (neonatal atelectasis) or to the collapse of previously