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8

Emphysematous and Cystic Lesions

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EMPHYSEMA

Classification

Based on the number of bronchi, bronchioles, and alveoli and on alveolar size and number, several categories of emphysema have been described in infants.[1] Congenital polyalveolar lobe is a lung lobe with the normal number of bronchial branchings but an increased number of alveoli (Fig. 8-1). In compensatory and obstructive emphysema, a lobe or lobes have normal bronchial branching and the normal number of enlarged alveoli. In congenital hypoplastic emphysema, a lobe has decreased numbers of bronchial and pulmonary artery branches and decreased numbers of markedly enlarged alveoli. The aerated lobe of bronchial atresia with a mucus plug has the normal peripheral bronchial number and normal number of markedly dilated alveoli.

Congenital Lobar Emphysema

Congenital lobar emphysema is caused by obstruction of a lobar bronchus with obstructive emphysema of the affected lobe (Color Fig. 8-1; Fig. 8-2). Abnormality or deficiency of bronchial cartilage, compression of bronchi by anomalous blood vessels or bronchogenic cysts, and obstruction of a bronchus by a mucus plug can be the cause. Instances of lobar emphysema of the left upper lobe from compression of a left tracheal bronchus by the aortic arch in Ellis-van Creveld syndrome and of the right upper lobe from compression of a right preeparterial bronchus by a right aortic arch are discussed in Chapter 7. The clinical picture can be that of severe acute respiratory distress from overdistention of the affected lobe, mediastinal shift, and collapse of the uninvolved ipsilateral lung, necessitating emergency surgery. The association with patent ductus arteriosus is unexplained. Rarely, the early radiologic picture is that of opacity, with a fluid-filled involved lobe. Lobar emphysema of the azygos lobe with superior caval syndrome has been reported.[2]

CYSTIC LUNG LESIONS

Several reviews consider the clinical and surgical aspects of bronchogenic cyst, congenital cystic adenomatoid malformation (CCAM), bronchial atresia, intralobar and extralobar pulmonary sequestration, or combinations of these disorders.[3–6] These reviews provide specific surgical and radiologic details.

Congenital Cystic Adenomatoid Malformation

The classification for CCAM originally proposed by Stocker and colleagues is used by most researchers (Figs. 8-3 and 8-4).[7] The type 1 lesion occurs in 50% of patients with CCAM. It is composed of large cysts, one of which may predominate (see Fig. 8-3*A*), and lined by ciliated or pseudostratified tall columnar epithelium. The usual clinical feature is respiratory distress, and differentiation from congenital lobar emphysema can be difficult.

The type 2 lesion represents 40% of CCAM cases. The lesion consists of multiple, evenly spaced, relatively small cysts (<1.2 cm in diameter; see Figs. 8-3*B* and 8-4) whose lining resembles that of terminal bronchioles. It was probably called congenital bronchiectasis in the older literature. Malformations of other organs are mostly associated with CCAM type 2. The clinical picture may be that of respiratory distress or one related to an associated malformation.

The type 3 lesion is the least common form of CCAM, representing only 10% of cases. The lesion is bulky, firm, and almost invariably causes mediastinal shift and early onset of respi-

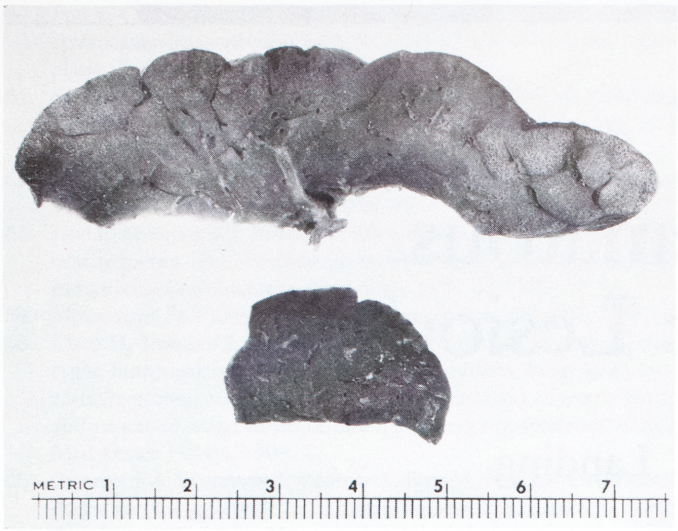


FIGURE 8-1. Congenital polyalveolar left upper lobe in a newborn infant (*upper, bisected specimen*) is compared with the left upper lobe of a normal control infant (*lower specimen*). Notice the prominence and size of the secondary lobules in the upper specimen. (Contributed by the editor.)

ratory difficulty. The involved lung is replaced by evenly spaced, small cysts (see Fig. 8-3*C*).

In the series of 16 patients with CCAM reported by Beluffi and associates, 14 were male and 2 female.[8] In 10, the lesion was on the right, was on the left in 4 patients, and was bilateral in 1 patient; only in one was an entire lung affected. Fetal hydrops with ascites and polyhydramnios occurs frequently with CCAM, especially with types 2 and 3, which have significantly poorer prognoses than CCAM type 1.[9,10] Antenatal diagnosis of CCAM type 3 by ultrasonography has been reported, and some researchers have suggested seeking CCAM sonographically in cases of unexplained elevation of maternal serum α-fetoprotein levels.[11,12] Whether the relation is an effect of CCAM or secondary to fetal hydrops is not clear. CCAM type 1 can be misdiagnosed as congenital diaphragmatic hernia, and attention to the normal abdominal gas pattern is necessary to avoid this error.[13] Systemic arterial supply to CCAM from the aorta has been observed and may suggest a pathogenetic relation of CCAM type 1 with pulmonary sequestration.[14] A cartilaginous variant of CCAM appears to be a form of CCAM type 2.[15] Possible relations of congenital mesenchymal malformation and of rhabdomyomatous dysplasia of the lung are discussed in Chapter 7.

Bronchogenic and Neurenteric Cysts

Bronchogenic cysts (Fig. 8-5) may occur within the lung or in the mediastinum. Whether all such cysts with a ciliated epithelial lining are truly bronchogenic can be debated, because the fetal esophagus also has a ciliated epithelial lining. The cysts can cause dyspnea, stridor, and compression of bronchi with resultant atelectasis or obstructive (*i.e.*, lobar) emphysema. Neurenteric cysts are usually more posterior in the thorax than bronchogenic cysts, and vertebral anomalies are usually demonstrable. In addition to respiratory symptoms, they can produce sensory or motor deficits, including back pain and gait disturbance.[16,17]

Cysts in the lung septa due to chronic pulmonary interstitial emphysema are lined by granulation tissue containing foreign-body giant cells, which is the typical response to interstitial air and similar to that formerly seen when pneumothorax was used in the treatment of pulmonary tuberculosis.

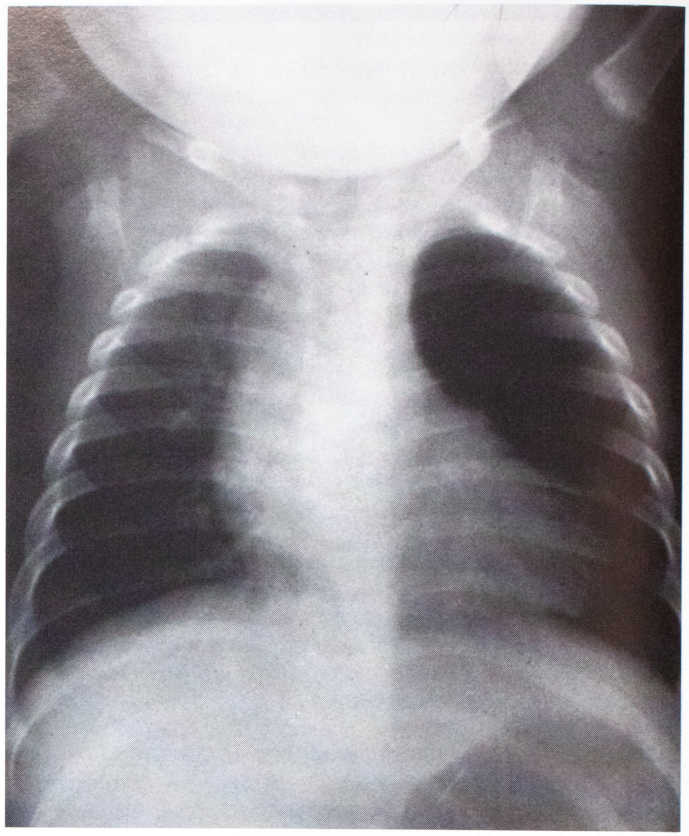


FIGURE 8-2. In the chest x-ray film of a patient with congenital lobar emphysema, the mediastinum is displaced toward the right side by a hyperinflated left upper lobe (see Color Fig. 8-2). (Courtesy of Daphne deMello, M.D., St. Louis, MO.)

Pulmonary Sequestration

Pulmonary sequestration is defined as an area of lung not connected to the bronchial tree by a normally located bronchus and having a systemic arterial supply. Sequestration occurs relatively frequently, with more than 500 cases reported.[18] The term "intralobar sequestration" is applied if the affected area is within the pleura of a lung, and extralobar sequestration is applied if it is not (Color Fig. 8-2; Fig. 8-6). Both types are more frequent on the left side, especially so for extralobar sequestration (*i.e.*, Rokitansky lobe). Approximately 10% of extralobar sequestrations are subdiaphragmatic and usually considered to be an abdominal mass.[19] An intralobar sequestration may appear radiologically as an opaque lung lesion. Despite the definition of no connection with the airway, they appear as air-filled cysts with significant frequency.

The clinical picture can be respiratory difficulty, but infection is common, and the clinical picture is then that of lung abscess. The arterial supply can be from a relatively large systemic artery arising from the subdiaphragmatic aorta, and care must be taken not to inadvertently sever the artery during surgery. Extralobar sequestration has been associated with massive pleural effusion.[20] In the older literature, because of the systemic artery supply to the affected region, patients who would now be placed in the category

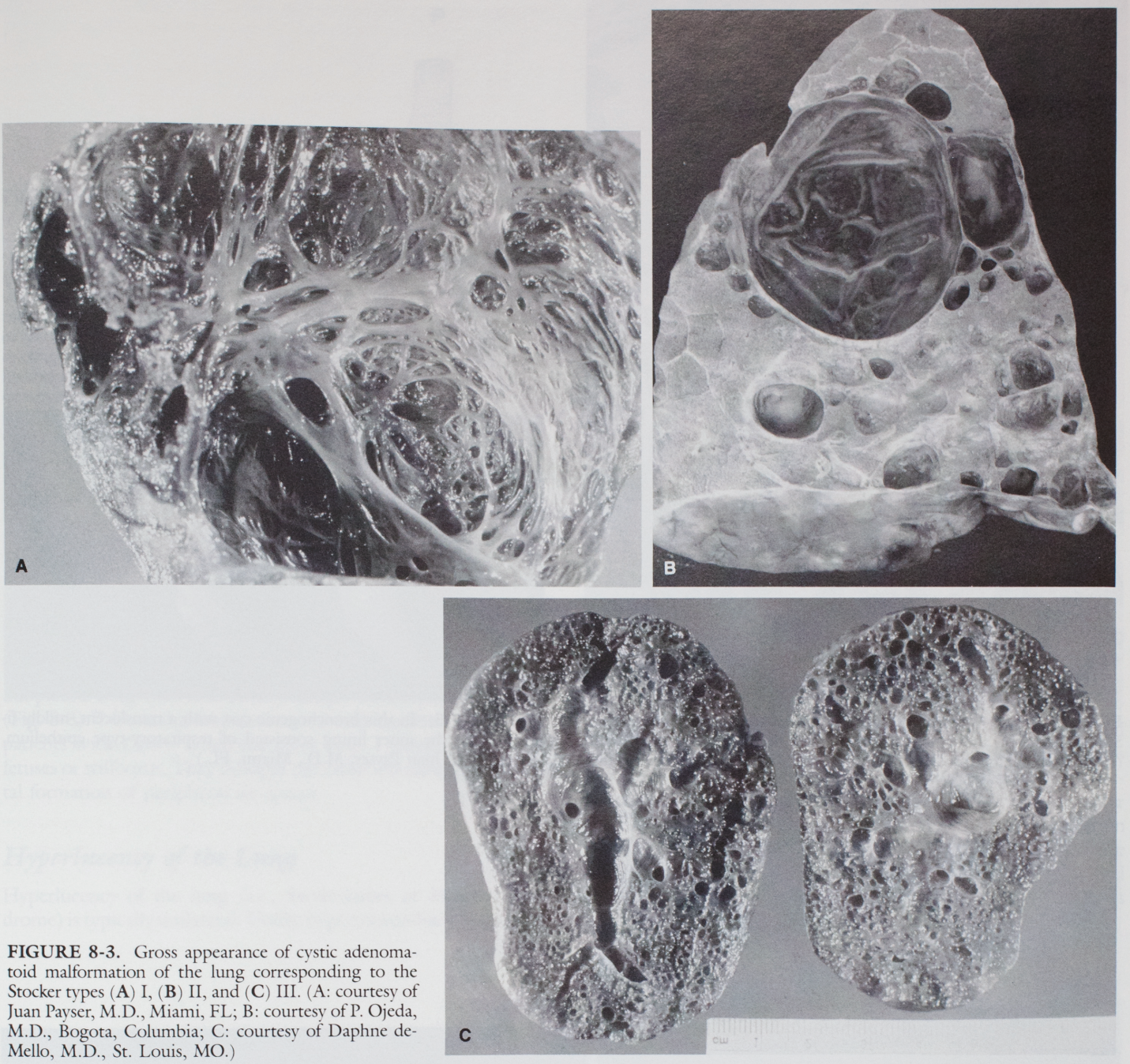


FIGURE 8-3. Gross appearance of cystic adenomatoid malformation of the lung corresponding to the Stocker types (**A**) I, (**B**) II, and (**C**) III. (A: courtesy of Juan Payser, M.D., Miami, FL; B: courtesy of P. Ojeda, M.D., Bogota, Columbia; C: courtesy of Daphne de-Mello, M.D., St. Louis, MO.)

of scimitar syndrome or esophageal origin of a bronchus were often considered in the category of pulmonary sequestration.

Figure 8-7 diagrams the possible mechanisms of intralobar and extralobar sequestration and of bronchopulmonary-foregut malformation (see Chap. 7). The theory is that an accessory tracheal bud from the foregut relatively close to the normal tracheal bud leads to intrapulmonary sequestration if the foregut connection later obliterates, but one arising from the foregut farther from the normal tracheal bud leads to extralobar sequestration. The possibility exists that bronchopulmonary-foregut malformation results from incomplete ingrowth on one side of the lateral ridges that normally fuse to form the tracheoesophageal septum. The reported associations of intralobar sequestration with bronchial isomerism and of extralobar sequestration with cystic adenomatoid malformation may convey pathogenetic information or may be coincidental in view of the frequency of pulmonary sequestration.[21,22]

Thilenius and colleagues analyzed the possible conditions within the broader sequestration spectrum, considering six anatomic variables: communication of the abnormal pulmonary tissue with the airway; systemic arterial supply to the lesion; abnormal venous drainage from the lesion to the right heart; esophageal communication with the abnormal area; defects of the diaphragm, which are especially frequent with left lower extralobar sequestration but are also seen with scimitar syndrome; and other lung abnormalities (*e.g.*, horseshoe lung, pulmonary hypoplasia).[23]

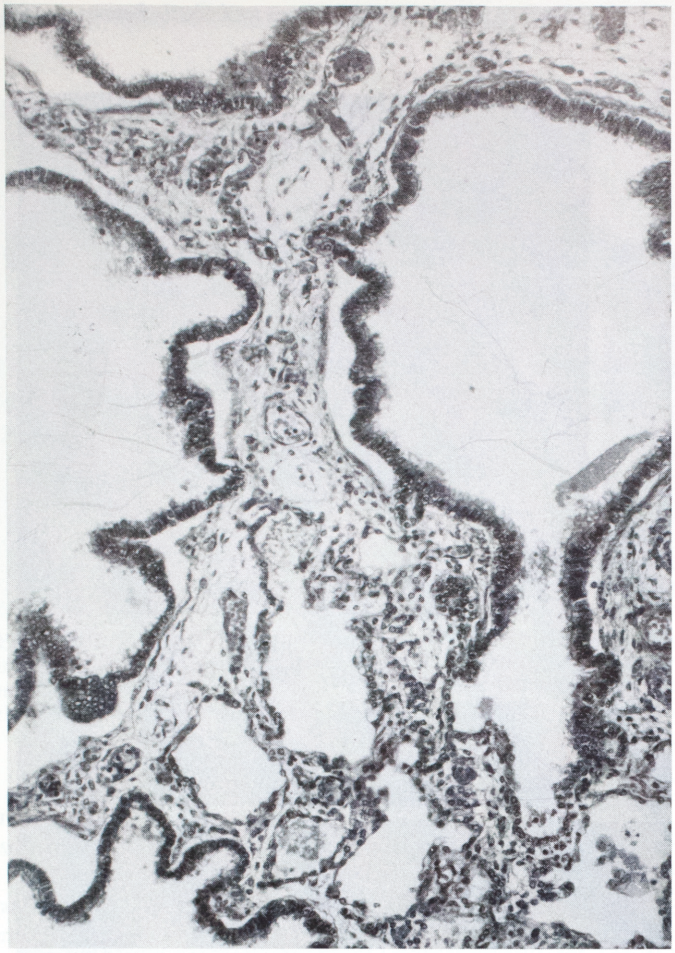


FIGURE 8-4. Microscopic view of a cystic adenomatoid malformation, Stocker type II, shows cystic spaces lined by respiratory epithelium. (H & E stain; intermediate magnification; contributed by the editor.)

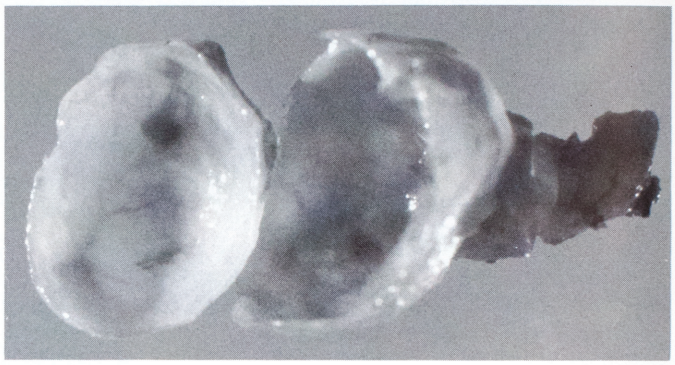
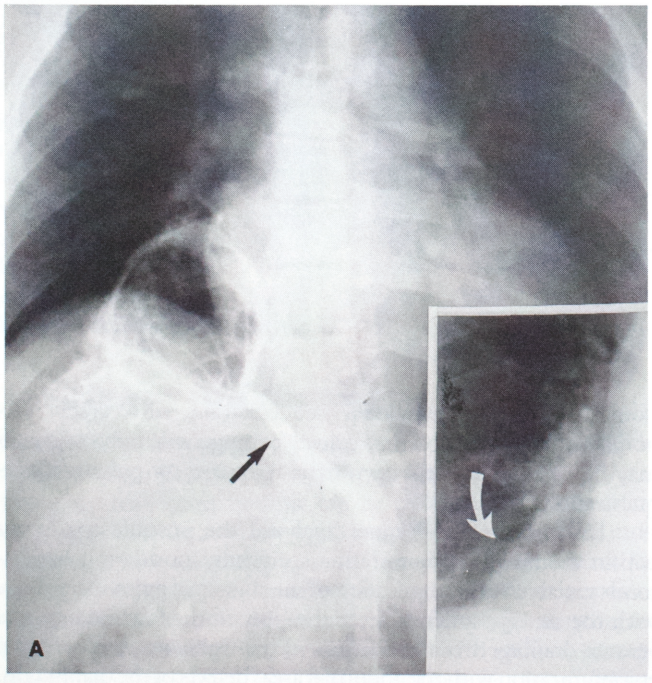


FIGURE 8-5. In this bronchogenic cyst with a translucent, mildly fibrotic wall, the inner lining consisted of respiratory-type epithelium. (Courtesy of Juan Payser, M.D., Miami, FL.)



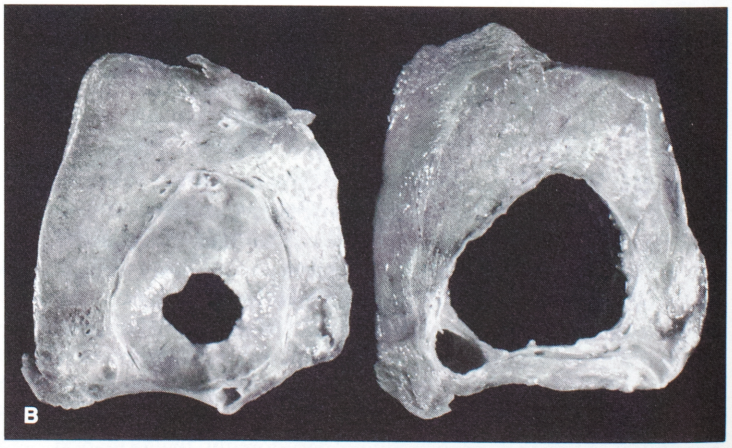


FIGURE 8-6. (**A**) An angiogram in a 26-year-old woman shows the systemic arterial supply to an intralobar sequestration of the right lower lobe (*arrow*). The lesion has an air-fluid level that is consistent with suppuration (*inset*). (**B**) The resected specimen is cavitary and infected. Notice the systemic artery at the base of the sequestration. (Contributed by the editor.)

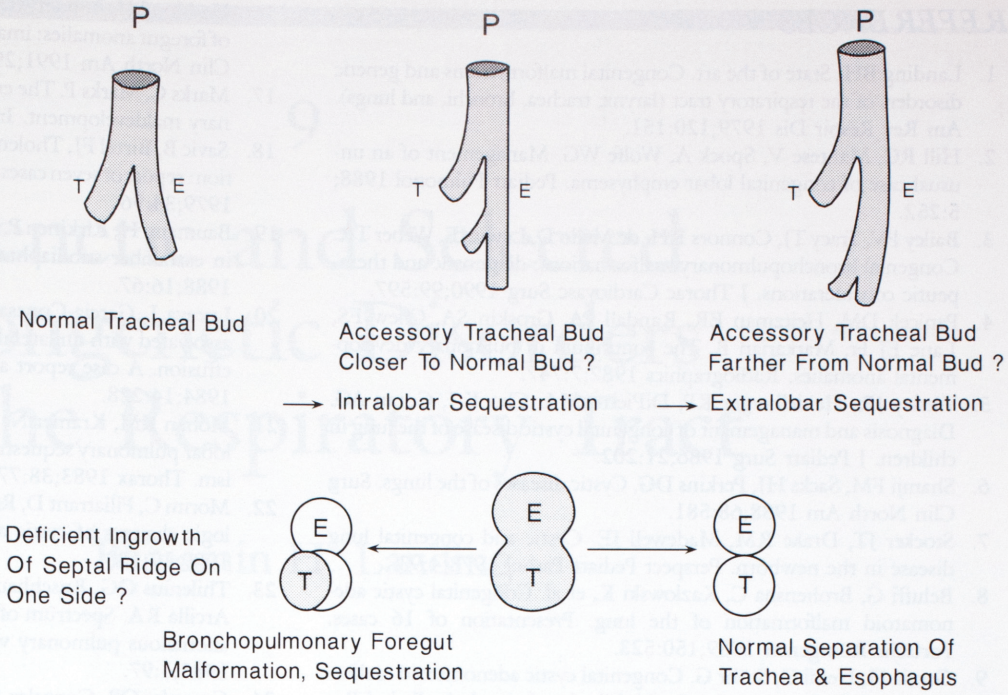


FIGURE 8-7. There are various types of pulmonary sequestration. The accessory tracheal bud caudal to the main tracheal bud gives rise to *intralobar* sequestration, if it is closer to the main bud, and to *extralobar* sequestration if it is farther from the main bud. The overlapping of patterns are forms of sequestration or congenital bronchopulmonary-foregut malformation. The strict definition of sequestration implies that the stem of the accessory tracheal bud regressed after its original formation. Bronchopulmonary-foregut malformation could be the result of incomplete ingrowth of one of the lateral ridges that normally fuse to form the tracheoesophageal septum. (E, esophagus; P, pharynx; T, trachea.)

Peripheral Cystic Lung Lesion of Down Syndrome

Subpleural lung cysts in Down syndrome were reported in 1984 (Fig. 8-8).[24] Gonzalez and associates found the lesion in 20% of patients with Down syndrome older than 1 month but not in fetuses or stillborns. They thought the cause was deficient postnatal formation of peripheral air spaces.

Hyperlucency of the Lung

Hyperlucency of the lung (*i.e.*, Swyer-James or Macleod syndrome) is typically unilateral. Unlike hypertransradiant lungs with obstructive or compensatory emphysema, the affected lung is not enlarged. Hyperlucency is the result of deficient pulmonary artery blood flow because of congenital absence of an artery, pulmonary artery thrombosis or embolism, or obliterative bronchiolitis or alveolitis (*i.e.*, chronic obstructive pseudoemphysema) after a viral respiratory tract infection or inhalation of pulmonary irritants (*e.g.*, chlorine, hydrocarbons).[25–28] Factitious pulmonary translucency from abnormalities of the pectoralis muscles or the shoulder girdle must be differentiated from the effects of prior radiation therapy for mediastinal or pulmonary disease.[29] Among three children with homocystinuria and nephrotic syndrome who had pulmonary artery thrombosis, one was diagnosed clinically as having Swyer-James syndrome.[30]

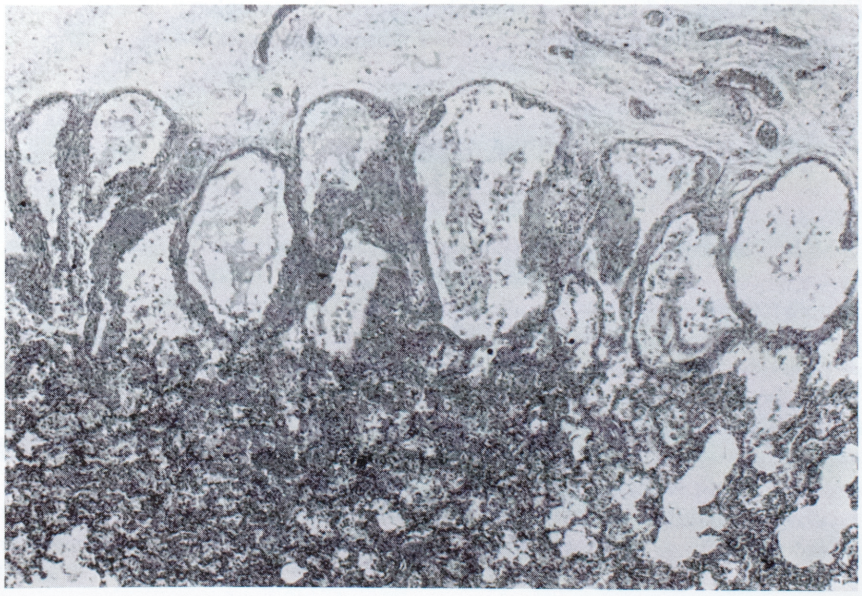


FIGURE 8-8. The subpleural location of pulmonary cysts in this microscopic view of Down syndrome. (H & E stain; low magnification.)

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