Review Article

Imaging of Pulmonary Sequestration

Richard E. Felker[1] and Ina L. D. Tonkin

American Journal of Roentgenology 1990;154:241-249.

Pulmonary sequestration is defined as a segment of lung parenchyma separated from the tracheobronchial tree and receiving its blood supply from a systemic artery rather than a pulmonary arterial branch. The arterial supply is usually from the aorta. Classically, it has been described in two forms: (1) intralobar sequestration (ILS), in which the sequestered part of lung lies within normal pulmonary visceral pleura, and (2) extralobar sequestration (ELS), in which the abnormal segment of lung is completely separate and enclosed in its own pleural investment.

The term *sequestration* was coined by Pryce [1] in 1946 to describe a disconnected bronchopulmonary mass or cyst with an anomalous systemic artery. Since this original description, terminology has become confusing as investigators have recognized many variants of sequestration not strictly meeting the original definition. The phrase *spectrum of pulmonary sequestration* was suggested by Sade et al. [2] in 1974 to describe this rather diverse group of pulmonary abnormalities that appear to be related and to have a common embryogenesis. Heitzman [3] expanded this concept further by including in the sequestration spectrum congenital malformations previously classified otherwise, such as lobar emphysema, congenital lung cysts, and even cystic adenomatoid malformation. The spectrum is now recognized as a continuum, with normal vessels supplying abnormal lung at one end and abnormal vessels supplying normal lung at the other end.

The sequestration spectrum includes all congenital lung anomalies in which there is abnormal connection of one or more of the four major components of lung tissue, namely, tracheobronchial airway, lung parenchyma, arterial supply, and/or venous drainage [4].

This article reviews the various abnormalities that may be seen in the pulmonary sequestration spectrum and discusses the imaging techniques that should be used for proper diagnosis.

Embryology

In order to appreciate the popular theories concerning the pathogenesis of sequestration, a brief review of the developmental stages of lung embryology is helpful.

The primitive foregut is surrounded by a vascular plexus that has anastomotic connections to the primitive ventral and dorsal aorta. The tracheobronchial component, entodermal in origin, meshes with the mesodermal vessels and connective tissue. As the primitive bronchus or so-called lung bud grows into the pulmonary plexus in a pseudopodlike fashion, the pulmonary artery, arising from the sixth branchial arch, similarly sends its branches into this foregut-mesodermal postbranchial pulmonary plexus. Normally the mesodermal vascular communications with the primitive aorta regress. Proponents of a congenital origin for sequestration have theorized persistence of one or rarely several of these communications as a plausible explanation for development of systemic supply to lung [5]. Others have presented evidence supporting theories for an acquired etiology [6].

Although many theories have been suggested for development of sequestration and its related malformations, no hypothesis has been universally accepted. The most widely accepted theories are summarized in Table 1.

Received July 5, 1989; accepted after revision September 11, 1989.
Presented at the annual meeting of the Cardiovascular and Interventional Radiological Society of Europe, Paris, July 1989.
[1] Both authors: Department of Radiology, The University of Tennessee, Memphis, 800 Madison, Memphis, TN 38163. Address reprint requests to R. E. Felker.

Intralobar Sequestration (ILS)

ILS is the most common form of classic pulmonary sequestration. On the basis of an extensive review of the literature from 1862 to 1978 by Savic et al. [13], ILS accounts for approximately 75% of all sequestrations. Most patients present with cough, sputum production, and recurrent pneumonia. Over 50% are symptomatic by the age of 20. A large majority of cases (98%) occur in the lower lobes, usually on the left side (Fig. 1A). All reported cases of ILS have been above the diaphragm. The arterial supply is usually from the descending thoracic aorta (73%) (Fig. 1B). According to Savic et al. [13], 21% are supplied by the upper abdominal aorta, or celiac or splenic arteries. In decreasing order of frequency, the intercostal, subclavian, internal thoracic, and pericardiophrenic arteries may be supplying arteries. Multiple supplying arteries are seen in about 16% of cases [13]. In 95% of cases the venous drainage is to the left atrium via the pulmonary veins so that there is a left-to-left shunt, a phenomenon that seldom occurs in any other condition (Fig. 1C). In less than 5% of cases venous drainage is to the systemic circulation via the azygos, hemiazygos, or intercostal veins or inferior or superior vena cava. In general, the anomalous pulmonary vein follows the artery: if the aberrant artery arises below or above the diaphragm, the vein drains also below or above the diaphragm [15].

TABLE 1: Theories Concerning the Origin of Sequestration and Its Variants

Reference	Theory
Pryce [1] (1946)	Traction by anomalous branch of aorta on segment of developing lung results in separation from normal lung
Smith [7] (1956)	Persistence of thoracic aortic arteries caused by insufficient pulmonary arterial supply; systemic blood pressure causes cystic degeneration of lung tissue
Boyden [8] (1958)	There is no causal relationship between nonfunctioning lung and systemic artery
Gebauer and Mason [9] (1959)	It is an acquired disease caused by a localized infectious process
Blesovsky [10] (1967)	Normal embryonic organizer control fails
Gerle et al. [11] (1968)	Accessory lung bud develops in embryo and either becomes incorporated into the normally developing lung (intralobar sequestration) or remains separate (extralobar sequestration); suggested term *congenital bronchopulmonary foregut malformation*
Moscarella and Wylie [12] (1968)	Intralobar sequestration is collection of bronchogenic cysts associated with a systemic artery
Stocker and Malczak [6] (1984)	It is an acquired disease that uses normally occurring pulmonary ligament arteries
Clements and Warner [4] (1987)	Theoretical "insult" disrupts normal connections producing "malinosculation" or faulty connection

Many believe that ILS is an acquired entity. This theory is supported by its rare association with other congenital anomalies and its usual occurrence in infants older than 2 months of age. In addition, there is the almost universal presence of chronic inflammation and fibrosis in ILS. Recent studies by Stocker [15] suggest that most intralobar sequestrations are acquired after birth. He proposes that small systemic arteries in the pulmonary ligament are parasitized to supply lung that has a compromised vascular supply due to bronchial obstruction in childhood [15]. Conversely, Felson [16] offers several reasons supporting a congenital origin for the intralobar type. He cites the presence of both ILS and ELS in the same patient, occasional bilaterality, the infrequency of ILS in the elderly, the association of ILS with other components of a venolobar syndrome, and the occurrence of ILS in twins as evidence for a congenital origin [16].

Extralobar Sequestration (ELS)

ELS accounts for 25% of all classic pulmonary sequestrations (Fig. 2) [13]. A majority of cases of ELS (61%) occur in the first 6 months of life, often presenting in the first day of life with dyspnea, cyanosis, and feeding difficulties. Rarely, ELS presents like ILS with recurrent pulmonary infection or, if a communication with the gastrointestinal tract is present, with gastrointestinal symptoms. Only 10% are asymptomatic [17].

More than 80% of cases receive arterial supply from the thoracic or abdominal aorta, with an additional 15% supplied from smaller arteries (such as splenic, gastric, subclavian, and intercostal). The remaining 5% obtain their blood supply from branches of the pulmonary artery or, rarely, from both systemic and pulmonary arteries. In a majority (>80%), venous drainage is via the systemic system (i.e., azygos, hemiazygos, and inferior vena cava). However, the venous drainage occasionally may be in continuity with the pulmonary veins. In addition, the venous drainage may be mixed with drainage to both the systemic and pulmonary circulations. An ELS receiving pulmonary arterial supply is more likely to have a pulmonary venous drainage [15]. Like ILS, most cases of ELS occur on the left side [18].

ELS is more often found between the lower lobe and the diaphragm but may be located in a variety of locations including within the substance of the diaphragm, in the mediastinum, within the lung, in the pleural or pericardial spaces, or in the retroperitoneum [17]. A majority (65%) of ELS cases have associated anomalies ranging from innocuous abnormalities such as accessory spleen to complex heart disease. Diaphragmatic hernia is the most common associated abnormality [15]. Because of its high prevalence in infants and its common association with extrapulmonary anomalies, ELS generally has been considered a congenital anomaly. Congestive heart failure and pulmonary overcirculation are well-recognized complications of ELS, particularly in infants [19]. Fluid imbalance may be reflected in both polyhydramnios during fetal life and anasarca in infants [15].

Variants of Sequestration

Once thought fundamentally significant, the distinction between ILS and ELS is now considered of secondary importance. A variety of components common to both classic forms may be present in any given patient. For instance, ELS and ILS may have venous drainage to both the left atrium and to the systemic circulation, combining features of both types. These may best be termed an ILS with mixed venous drainage or an ELS with mixed venous drainage (Fig. 3). Moreover, anomalous venous drainage can occur with any congenital lung malformation [21].

The scimitar syndrome is a well-described variant of the sequestration spectrum. Synonyms include pulmonary venolobar syndrome and hypogenetic lung syndrome. The characteristic abnormality is that the anomalous vein, which has a scimitarlike appearance, drains to the inferior vena cava. The entire right lung drains to the inferior vena cava or its junction with the right atrium. Other features that may or may not be present include (1) hypoplasia of right lung with dextroposition of the heart, (2) anomalies of right lung lobation, (3) hypoplasia of the right pulmonary artery, and (4) anomalous systemic supply to the lung from the abdominal aorta or

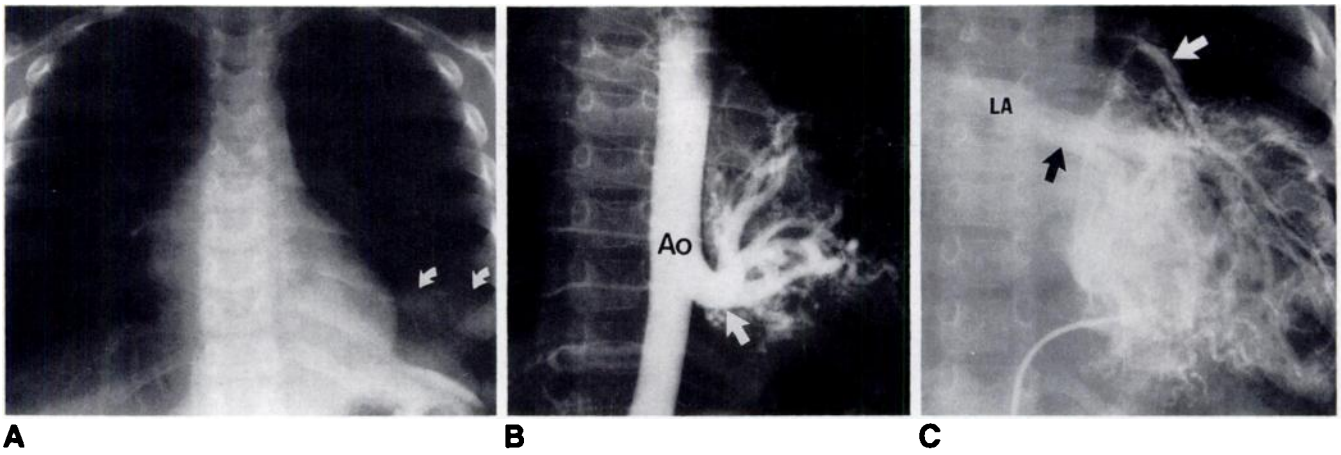


Fig. 1.—5-year-old boy with intralobar sequestration.
***A*, Chest radiograph after approximately 1 year of treatment for pneumonia shows opacity (*arrows*) in left lower lobe and retrocardiac region.**
***B*, Thoracic aortogram shows large supplying artery (*arrow*) to left lower lobe from descending aorta (Ao).**
***C*, Venous return is to left atrium (LA) via left lower lobe pulmonary vein (*black arrow*). There is also some reflux into left lower lobe pulmonary artery (*white arrow*). Intralobar sequestration was confirmed by surgical resection.**
(Reprinted with permission from Tonkin [14].)

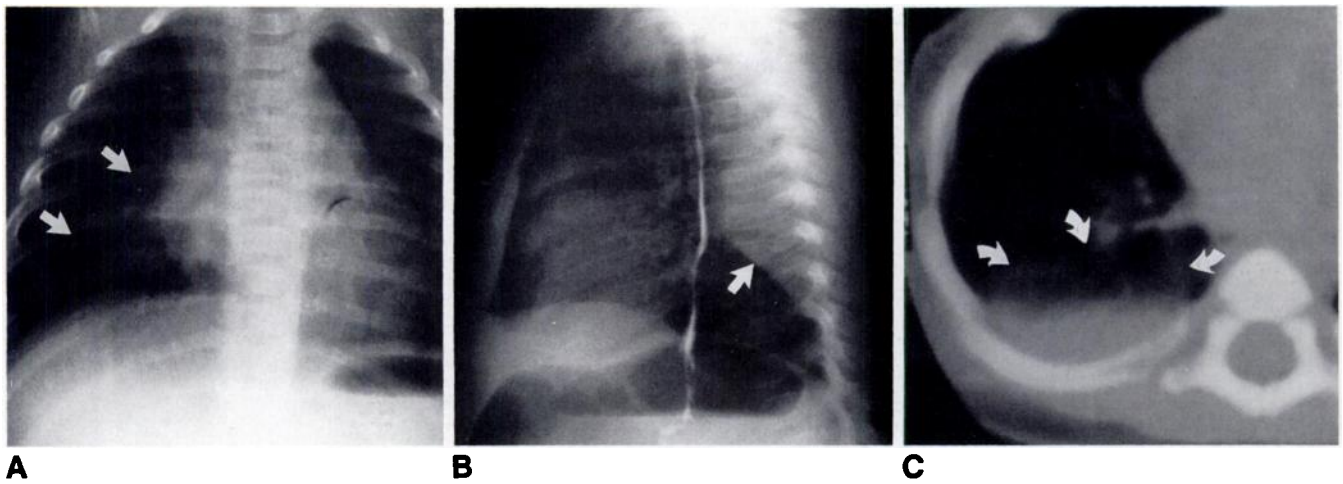


Fig. 2.—3-month-old girl with extralobar sequestration.
***A*, Posteroanterior chest radiograph in infant with mild respiratory distress shows unusual opacity in right mid lung (*arrows*).**
***B*, On lateral projection, opacity is posterior (*arrow*). There was no indentation on esophagus nor esophageal distension on this view and others (not shown).**
***C*, Unenhanced CT scan shows mixed air-filled and solid mass in posterior aspect of right lung (*arrows*). At surgery, supplying artery from thoracic aorta was identified. Venous drainage was to systemic system via azygos vein. Pathology confirmed mediastinal extralobar sequestration.**

its main branches [22]. A classic case of scimitar syndrome is shown in Figure 4.

Forty-eight percent of cases of scimitar syndrome are associated with an aberrant systemic artery. Asymptomatic cases are often detected on routine chest radiographs. The patients with a more severe degree of lung hypoplasia or associated congenital heart disease are more likely to become symptomatic in infancy or childhood.

Horseshoe lung, a rare congenital anomaly, frequently is associated with the typical characteristics of the scimitar syndrome. In this condition an isthmus of pulmonary tissue extends from the right lung base across the midline behind the pericardium and fuses with the base of the left lung [23]. Pathologically the parenchyma of the isthmus is normal [24].

Other pulmonary anomalies that may demonstrate some overlapping characteristics of pulmonary sequestration include cystic adenomatoid malformation, pulmonary arteriovenous fistula, and systemic arterial supply to normal lung.

Cystic adenomatoid malformation is primarily an abnormality of the pulmonary parenchyma in which there is overgrowth of bronchioles [25]. The vascular supply is usually normal; however, occasionally there is an aberrant systemic artery [26]. Also, cystic adenomatoid malformation may arise in an otherwise classic pulmonary sequestration [4]. For these reasons it is thought to be closely related to pulmonary sequestration and its variants in the sequestration spectrum.

Pulmonary arteriovenous fistula or pulmonary arteriovenous malformation consists of abnormal connection of a pulmonary artery to a pulmonary vein [14]. The pulmonary parenchyma itself is normal. It is mentioned only because of the increasing evidence that it has a common embryogenesis with sequestration and its related anomalies.

Systemic arterial supply to normal lung is an uncommon but well-recognized variant of the sequestration spectrum [27]. The arterial supply of a normal segment of pulmonary parenchyma is supplied by an aberrant vessel usually arising

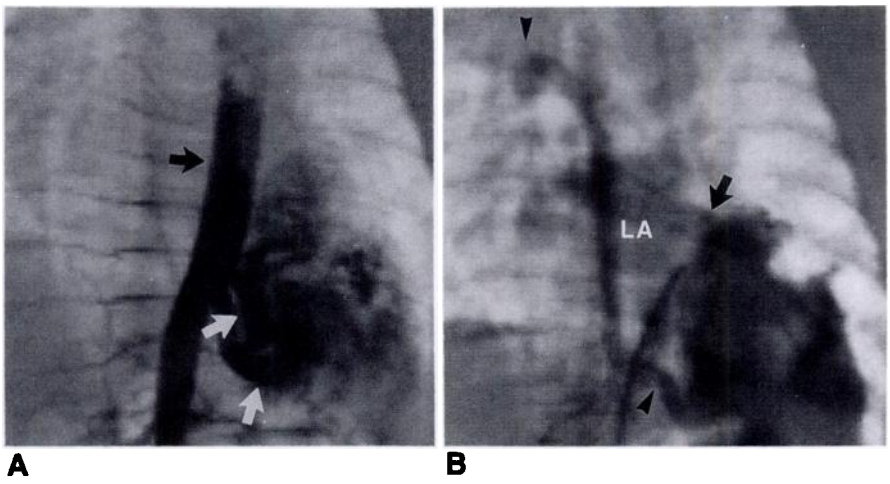


Fig. 3.—Intralobar sequestration with mixed venous drainage in 2-year-old boy with congestive heart failure.

***A*, Chest radiograph (*not shown*) showed opacity in left lower lobe and signs of congestive heart failure. Digital subtraction angiogram shows two supplying arteries (*white arrows*) to sequestered posterior basilar segment of left lower lobe from aorta (*black arrow*).**

***B*, Levophase of angiogram shows mixed venous drainage to left lower lobe pulmonary vein (*arrow*) emptying into left atrium (LA) and azygos vein (*arrowheads*), which empties into superior vena cava.**

(Reprinted with permission from Tonkin [20].)

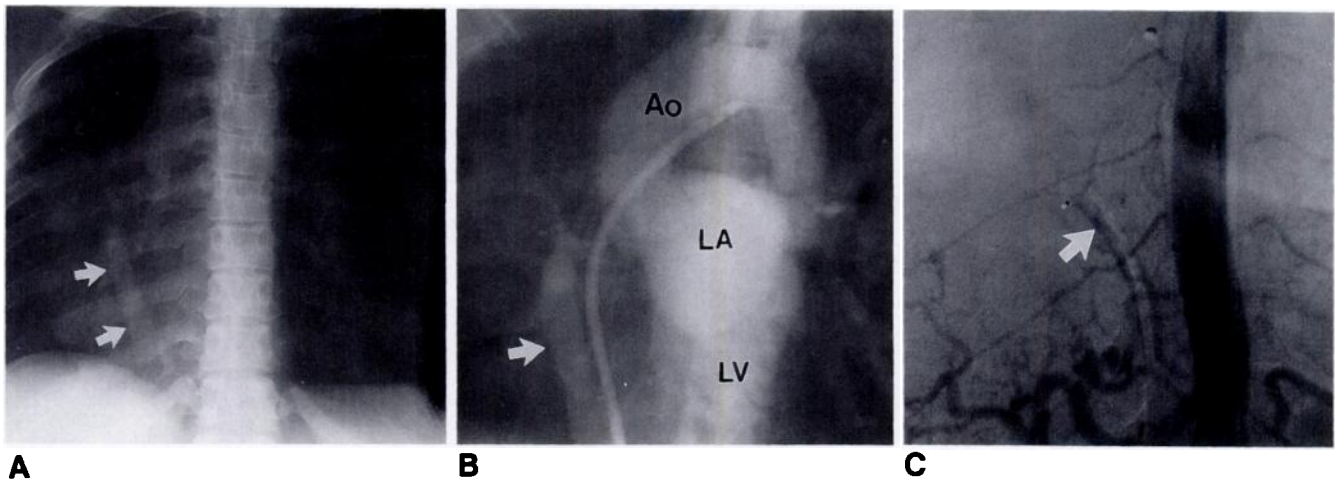


Fig. 4.—Classic scimitar syndrome in asymptomatic 24-year-old woman.

***A*, Routine preoperative chest radiograph shows hypoplasia of right lung and dextroposition of heart and anomalous pulmonary vein, or scimitar sign (*arrows*).**

***B*, Venous phase of pulmonary arteriogram shows anomalous vein (*arrow*) draining into inferior vena cava. Left pulmonary vein is shown emptying into left atrium (LA) with filling of ventricle (LV) and aorta (Ao). Arterial phase of pulmonary arteriogram (*not shown*) showed hypoplastic right pulmonary artery. Left pulmonary artery was normally developed.**

***C*, Thoracic aortogram with subtraction technique shows very small supplying artery (*arrow*) from upper abdominal aorta to right lower lobe.**

from the aorta. The basilar segments of the left lower lobe are more frequently involved with no radiographic abnormality of the pulmonary parenchyma. These patients may be totally asymptomatic. Eventually, however, left ventricular enlargement and congestive heart failure caused by the left-to-left shunt may develop. The combination of this type of systemic arterial supply to lung with normal bronchial connections but with coexistent infection has been referred to as pseudosequestration. It is presumed that chronically inflamed lung activates neovascularization of the relatively high-pressure systemic circulation to lung. The radiographic findings may be identical to those of classic pulmonary sequestration (Fig. 5) [28]. However, in some cases the distinction between true pulmonary sequestration and pseudosequestration is made only after disappearance of aberrant vessels and alleviation of symptoms after antibiotic therapy.

In primitive types of pulmonary sequestration there may be communication with the gastrointestinal tract. In such cases a tubular structure lined by columnar or squamous epithelium and containing intramural cartilage connects the esophagus or stomach to anomalous pulmonary tissue supplied by a systemic artery. These have been referred to as esophageal lung or gastric lung [16].

Complications Associated with Sequestration and Its Variants

Infection is the most common complication of pulmonary sequestration. It is more commonly seen with ILS [17].

Because of the shunting that may occur, congestive heart failure is a rare but well-documented complication of sequestration and its variants [29]. In fact, it may occur in any patient with a systemic arterial connection to lung. Sequestration and its variants should be included in the differential diagnosis of any infant or child with congestive heart failure and suspected pneumonia or atelectasis. Additional reported complications of pulmonary sequestration and its variants include lobar emphysema [30], hemoptysis, hemothorax [31], and pleural effusion. Massive fatal pleural effusion has been reported with ELS [32].

Imaging Studies

Diagnosis should be directed toward identification of each of the six components of the sequestration spectrum: (1) sequestered or dysplastic lung mass; (2) aberrant arterial supply; (3) anomalous venous drainage; (4) communications with the bronchus or gastrointestinal tract; (5) gross lung anomalies, such as horseshoe lungs or hypoplasia; and (6) defects of the diaphragm. Any combination of these primary lesions can occur in an individual patient. The particular mixture of findings may not conform exactly to the strict definition of the more common sequestration forms; hence, precise categorization may not be possible.

Chest Radiographs

Plain radiographs of the chest often show abnormal lung or vascular shadows that suggest the diagnosis (Fig. 6A). The usual presentation is a single homogeneous opacity, as demonstrated in Figure 1, or, less often, a cystic mass in the base of one lung, as shown in Figure 7 [16]. Less specific findings include recurrent pneumonias and focal bronchiectatic changes. By far the majority of sequestrations are located in the lower lobes. In fact, any persistent abnormality in the posteromedial basal segment of a lower lobe in a child or young adult should suggest the diagnosis. ELS, in particular,

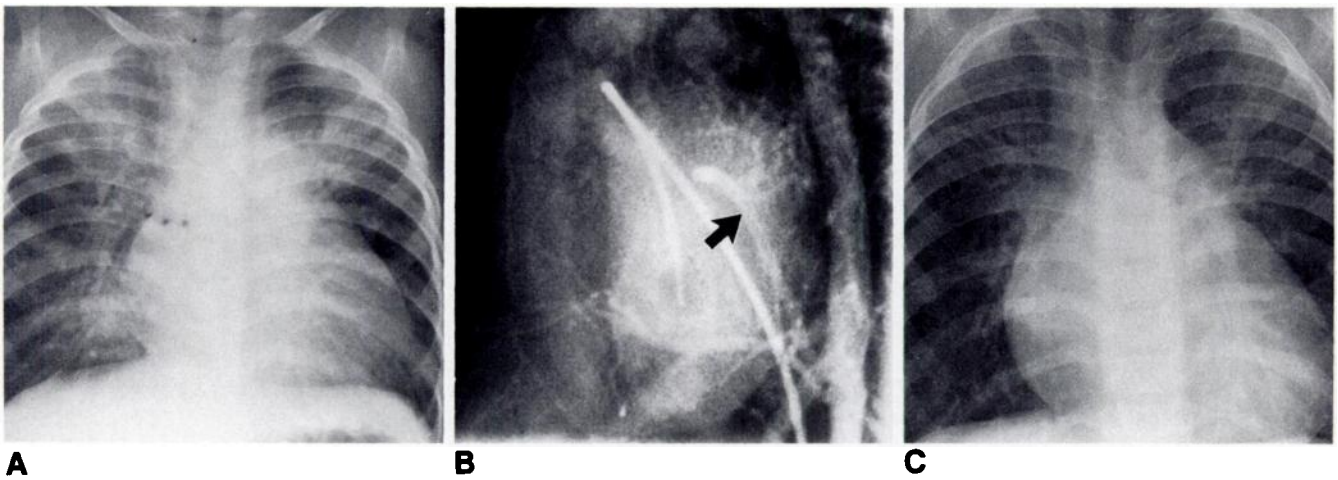


Fig. 5.—3½-year-old boy with atrioventricular canal defect and systemic arterial supply to infected lung (pseudosequestration).

***A*, Posteroanterior chest radiograph shows alveolar opacities in right lower lobe and upper lobes, compatible with bronchial pneumonia or asthmatic bronchitis.**

***B*, Late phase of left ventriculogram in four-chamber view shows large supplying artery (*arrow*) from abdominal aorta to right lower lobe with pulmonary venous drainage to right lower pulmonary veins (*not shown*).**

***C*, Follow-up chest radiograph 3 years later shows no alveolar disease. Cineangiocardiogram obtained at the same time (*not shown*) no longer showed supplying artery to right lower lobe. Total cardiac repair was performed without removal of pseudosequestration involving right lower lobe. The patient is currently asymptomatic.**

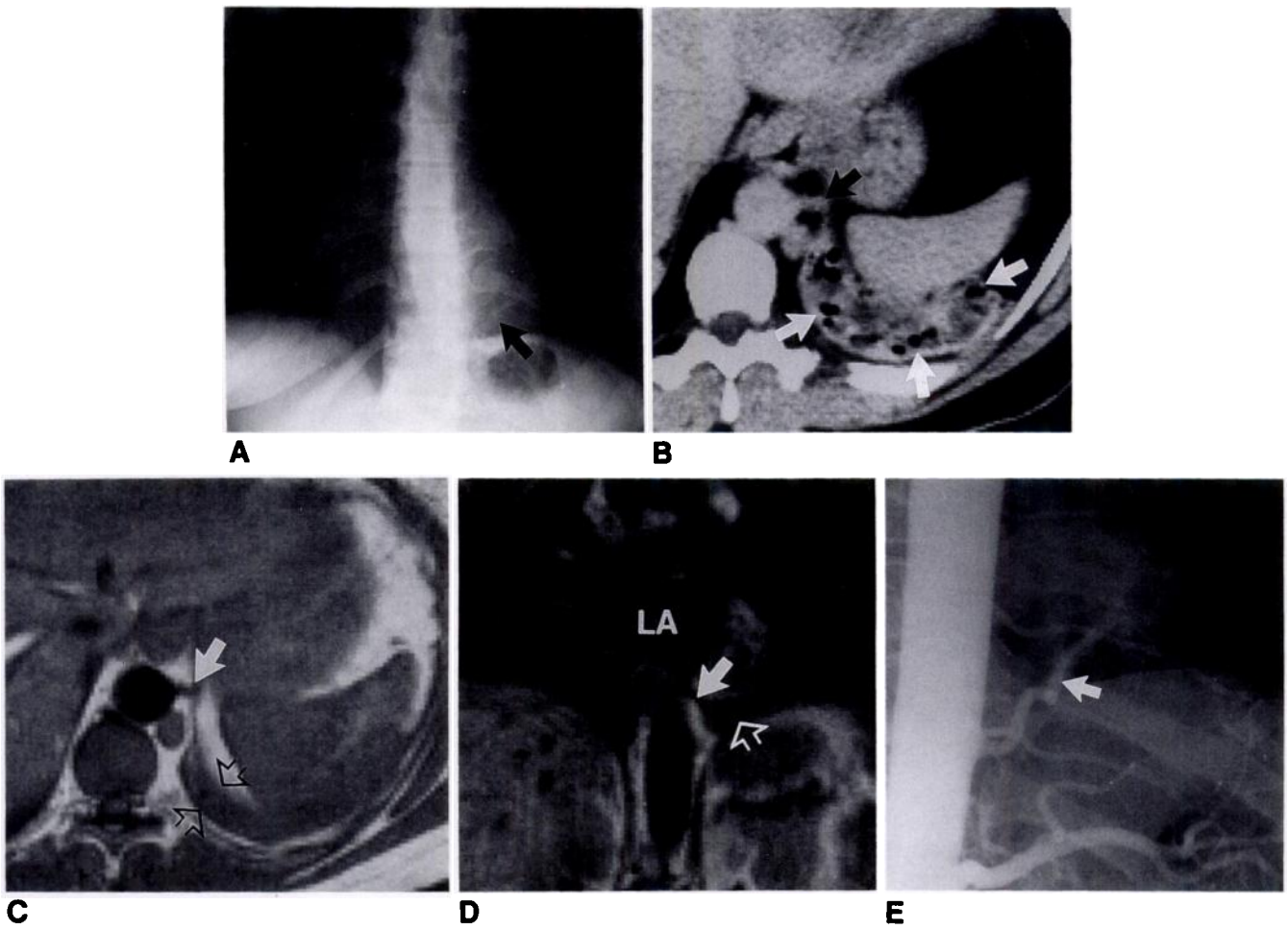


Fig. 6.—Intralobar sequestration in 30-year-old woman with pneumonia.

***A*, Posteroanterior chest radiograph at time of automobile accident, 10 years before pneumonia symptoms, shows vascular shadow in left lower lung (*arrow*). This was not recognized at the time.**

***B*, Several years later after persistent pneumonia. CT scan shows multicystic mass in left lung base (*white arrows*) and supplying artery (*black arrow*) from aorta.**

***C*, Axial MR image shows supplying artery (*solid arrow*) as well as abnormal lung in posterior basilar segments (*open arrows*).**

***D*, Coronal MR image shows abnormal left lower lobe lesion (*open arrow*) with draining vein (*solid arrow*) to left atrium (LA).**

***E*, Aortogram shows supplying artery (*arrow*) seen on chest radiograph, CT scan, and MR image. Intralobar sequestration was confirmed by surgical pathology.**

is commonly identified incidentally on routine chest radiographs in asymptomatic patients [17].

Bronchography

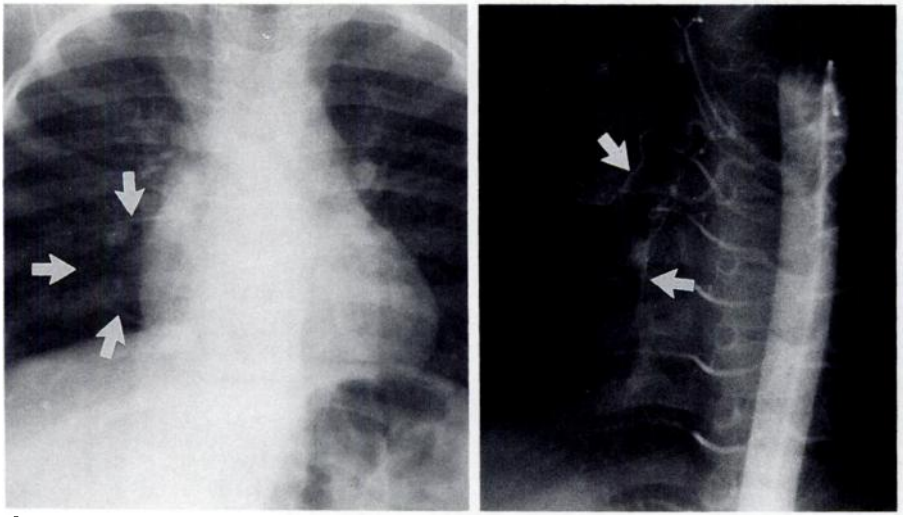
Bronchograms, rarely used today, are unnecessary because the combination of other findings is sufficient for making the diagnosis. However, if contrast material is used, it should be introduced selectively into the bronchus. Classically there is no bronchial supply to the aberrant sequestered lung, with normal bronchi draped around but not communicating with the involved segment of lung. Rarely, contrast material may leak into the sequestration [34].

Arteriography

Traditionally, the diagnosis of pulmonary sequestration has been made definitively with arteriography. The classic findings are the anomalous systemic arterial supply and anomalous venous drainage shown by aortography (Figs. 1B and 1C). In neonates, arteriography may be performed with the use of an umbilical artery catheter placed by the neonatologist.

The pulmonary arteriogram may be useful in confirming the absence of pulmonary arterial supply to an area of lung [35]. Patients with pseudosequestration show similar findings with aortography and selective injections. However, in pseudosequestration the arterial supply is more commonly multiple and the arteries are not clearly anomalous [28].

Digital subtraction angiography has been an excellent imaging technique for evaluating patients with pulmonary se-



A B

Fig. 7.—2-year-old boy with bronchogenic cyst arising in an intralobar sequestration.
A, Posteroanterior chest radiograph shows thin-walled air-filled cyst (*arrows*) in right mid lung compatible with simple cyst or bronchogenic cyst.
B, Thoracic aortogram shows two unusual vessels (*arrows*) in wall of cyst extending from upper descending thoracic aorta and intercostal arteries. Surgical pathology confirmed an unusual type of bronchogenic cyst arising in intralobar sequestration.
(Reprinted with permission from Fitch et al. [33].)

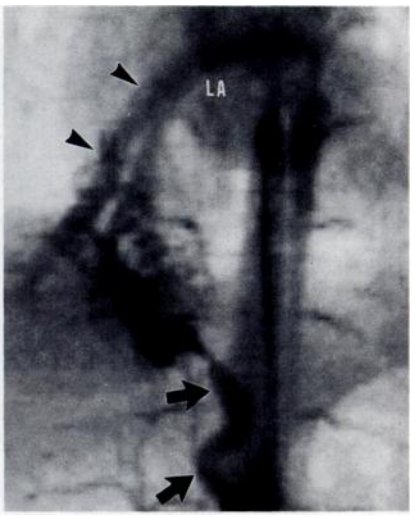


Fig. 8.—Intralobar sequestration in 2½-year-old girl with wheezing. Posteroanterior chest radiograph (*not shown*) showed hyperinflation and alveolar opacity in lateral basilar segment of right lower lobe. Intraarterial digital subtraction angiogram shows large supplying artery from abdominal aorta, just above celiac axis (*arrows*). There is a hypervascular mass involving right lower lobe with venous drainage (*arrowheads*) to left atrium (LA). Intralobar sequestration was confirmed by surgery. (Reprinted with permission from Tonkin [20].)

questration. The supplying artery is well imaged, and the venous anatomy is shown even better with digital subtraction angiographic techniques (Figs. 3B and 8) [20, 36].

Barium Studies

Barium studies usually are required to show communication with the esophagus or stomach. Figure 9 is an example of bilateral esophageal lung in which right and left lower lobe sequestrations communicate with the esophagus. The barium esophagogram is also useful in detecting associated anomalies such as vascular rings [37].

CT

CT can show both abnormal lung parenchyma and the aberrant vessels supplying a sequestration (Figs. 2C and 6B) [38]. Calcification, an uncommon but well-documented finding in sequestrations, is more readily appreciated with CT [39].

MR Imaging

MR imaging is ideally suited for the diagnosis of pulmonary sequestration and its variants because of excellent demonstration of vessels and pulmonary parenchymal abnormalities in multiple imaging planes. It may alleviate the need for angiography. In fact, MR may identify vessels not recognized by arteriography [40]. Several cases of MR diagnosis of pulmonary sequestration have been reported [41–43]. We are reporting two additional patients in whom ILS was shown by MR (Figs. 6 and 9).

Sonography

Pulmonary sequestration has been demonstrated by sonography both in utero [44] and in infants [45]. Lesions adjacent to the diaphragm or liver are best imaged with sonography. The diagnosis should be suspected in any fetus with a lung mass, particularly if hydramnios is present. The mass usually is homogeneously echogenic but may have a cystic or complex character. This appearance is not specific for pulmonary sequestration but should arouse suspicion for a number of possibilities in the pulmonary sequestration spectrum. The aberrant arterial and venous drainage may be shown with sonography [46]. Duplex Doppler may be useful in showing the aberrant arterial supply [47]. Color Doppler flow imaging shows promise as a noninvasive tool for identifying arterial supply and venous drainage in both infants and fetuses. In addition, sonographically directed fine-needle biopsy may be useful in making a definitive diagnosis of ELS. The retrieval of respiratory epithelium in an extrapulmonary site confirms the diagnosis [48].

Treatment

Surgery usually has been necessary for treatment of pulmonary sequestration, as infection almost universally occurs

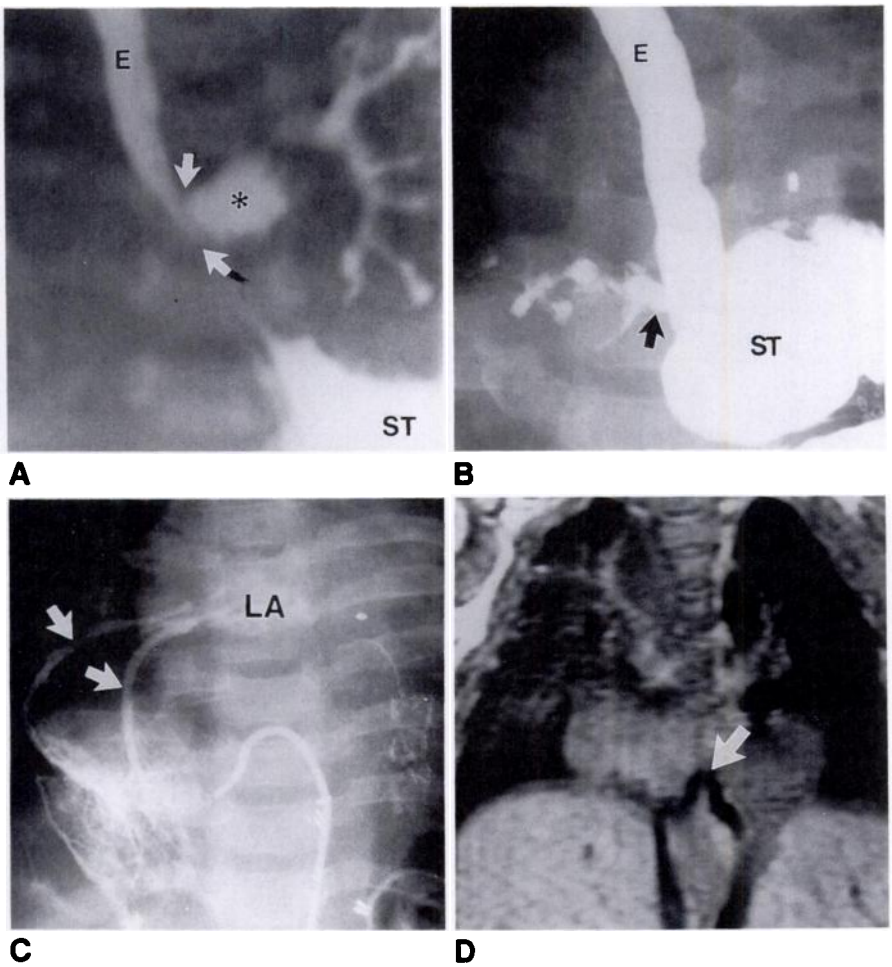


Fig. 9.—Neonate with congestive heart failure with bilateral lower lobe sequestrations communicating with esophagus (esophageal lung).

***A*, Esophagogram shows communication (*arrows*) between esophagus (E) and bronchi in left lower lobe via barium-filled cavity (*asterisk*). A sequestration was resected. ST = stomach.**

***B*, Several months later, patient was in congestive heart failure. Posteroanterior chest radiograph (*not shown*) showed evidence of right thoracotomy for repair of tracheoesophageal fistula and clips in left lower lobe from left thoracotomy for removal of left lower lobe sequestration. There was a right-sided opacity along right heart border and signs of congestive heart failure. Second esophagogram shows communication (*arrow*) between esophagus and right lower lobe bronchi. There is also barium within stomach.**

***C*, Aortogram with selective injection into supplying artery from aorta opacifies right lower lobe vascular mass. Venous drainage is to pulmonary veins (*arrows*) and into left atrium (LA).**

***D*, Coronal MR image shows supplying artery (*arrow*) from abdominal aorta to sequestered right lower lobe. This is an unusual case of bilateral pulmonary sequestrations communicating with esophagus or bilateral esophageal lung.**

(Courtesy of D. Mulvihill and A. Robinson, New Orleans, LA.)

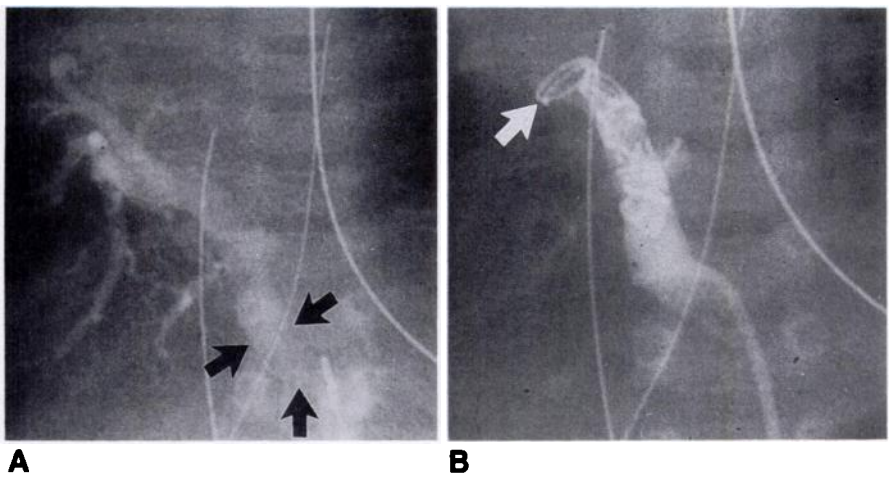


Fig. 10.—Neonate with congestive heart failure and systemic vessel to right lower lobe. Vessel was successfully embolized and heart failure resolved. Surgery has not been performed.

***A*, Cineaortogram via umbilical artery catheter shows large supplying artery from upper abdominal aorta (*arrows*).**

***B*, Supplying artery was successfully embolized with multiple coils (*arrow*).**

if the abnormal lung is not removed [17]. It is imperative that the arterial supply and venous drainage of the sequestered segment be identified preoperatively to prevent massive intraoperative hemorrhage due to transection of unidentified vessels.

Preoperative medical treatment may result in regression of some systemic pulmonary anastomoses, particularly if acquired through infection. Embolization of the anomalous vessels with angiographic techniques may be helpful in reducing operative blood loss. Also, embolization may be the definitive treatment for systemic supply to lung to prevent shunting and its subsequent complications (Fig. 10).

Conclusions

The pulmonary sequestration spectrum represents a heterogeneous and complex group of abnormalities involving

anomalous connections of pulmonary parenchyma, pulmonary and systemic vasculature, and, rarely, the gastrointestinal tract. In the individual patient, emphasis should be given to identifying the particular components of the spectrum rather than to categorizing the abnormalities. This approach is essential for proper diagnosis and appropriate therapy of the numerous abnormalities that may be present.

In the workup of sequestration we recommend plain chest radiographs followed by barium esophagography and MR imaging. Arteriography is recommended if MR is inconclusive. CT, sonography, and bronchography have a lesser role in diagnosis but may provide additional useful information.

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