Sequestration of the Lung

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- Congenital malformation of primitive foregut consisting of
  - Non-functioning segment of lung
  - Doesn’t communicate with bronchial tree
  - Supplied by systemic artery
- LLL:RLL 2:1
- Intralobar (80%): extralobar (20%)
Intralobar Sequestration
General

- Lies within same visceral pleura as lobe in which it occurs
- Non-functioning — intralobar sequestrations are closed systems not communicating with tracheobronchial tree unless infected
Intralobar Sequestration
Blood Supply

- Arterial supply from aorta
  - Most commonly descending thoracic aorta
- Venous drainage almost always to pulmonary venous system (left to right shunt)
Intralobar Sequestration

General

- Left lower lobe, posterior segment in about 2/3 of cases
  - In remainder, right lower lobe, posterior segment
- Not usually associated with other anomalies
  - Can be in 14%
- Frequently recognized in adulthood due to pneumonia
Intralobar Sequestration

X-ray

- When not infected, appear as solid masses in left lower lobe usually touching diaphragm
- If infected and communicating with bronchial tree, may be cystic, air-containing with fluid levels
Intralobar Sequestration

X-ray

- May be obscured by surrounding pneumonia in normal lung
- Classically, bronchogram shows bronchi draped around mass
Extralobar Sequestration

General

- Develops as accessory lung contained within its own pleura
- Rarely becomes infected
- Almost always presents as homogeneous soft tissue mass
- Related to left hemidiaphragm in 90% of cases
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.
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.
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 anglogram 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.
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;
(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.
Fig. 4-3-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. Cineangiocardiacogram 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 a symptomatic
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 posteromedi al basal segment of a lower lobe in a child or young adult should suggest the diagnosis.
ELS is commonly identified incidentally on routine chest radiographs in asymptomatic patients [}
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.
Intralobar sequestration in a 2 and 1/2 year-old girl with wheezing.

Posteroanterior chest radiograph (not shown) showed hyperinflation and alveolar opacity in the lateral basilar segment of the right lower lobe. Intralobar sequestration was confirmed by surgery.

Intraarterial digital subtraction angiogram shows large supplying artery from the abdominal aorta, just above the celiac axis (arrows). There is a hypervascular mass involving the right lower lobe with venous drainage (arrowheads) to the left atrium (LA). Intralobar sequestration was confirmed by surgery.
Pulmonary sequestration has been demonstrated by sonography both in utero and in infants. 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.

Duplex Doppler may be useful in showing the aberrant arterial supply.

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.
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 opacities 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.
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).
Surgery usually has been necessary for treatment of pulmonary sequestration, as infection almost universally occurs if the abnormal lung is not removed.

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.
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.