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Pulmonary Sequestration

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What do these two patients have in common?

Patient 1: 50 y.o. non-smoking female with several months cough and hemoptysis;

CXR: posterior left lower lung consolidation

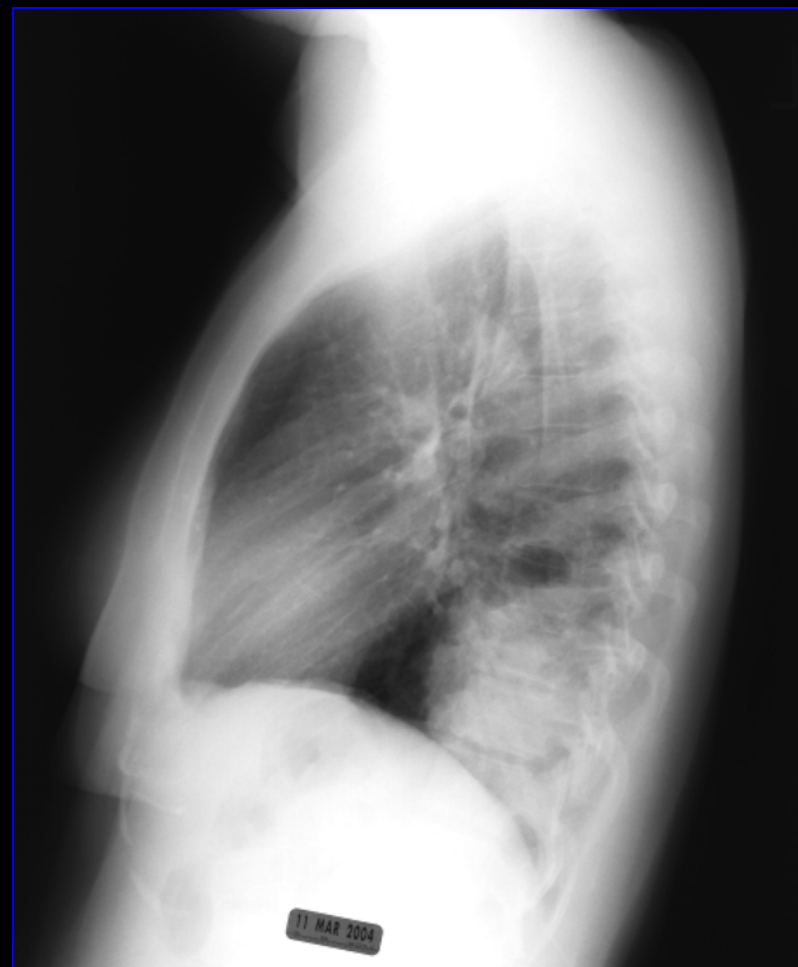
Patient 2: Asymptomatic neonate with incidental chest mass found on prenatal U/S;

CT: posterior left lower lung mass



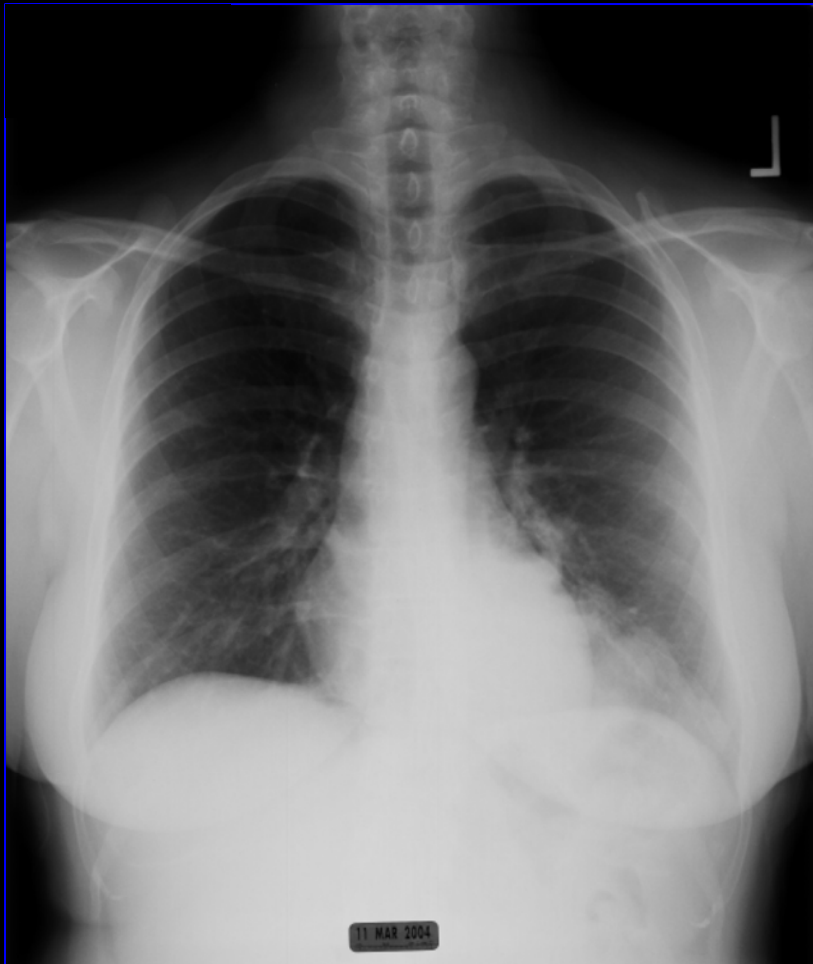
Patient 1

50 y.o. female with several months
of cough and hemoptysis



Patient 1

50 y.o. female with several months
of cough and hemoptysis



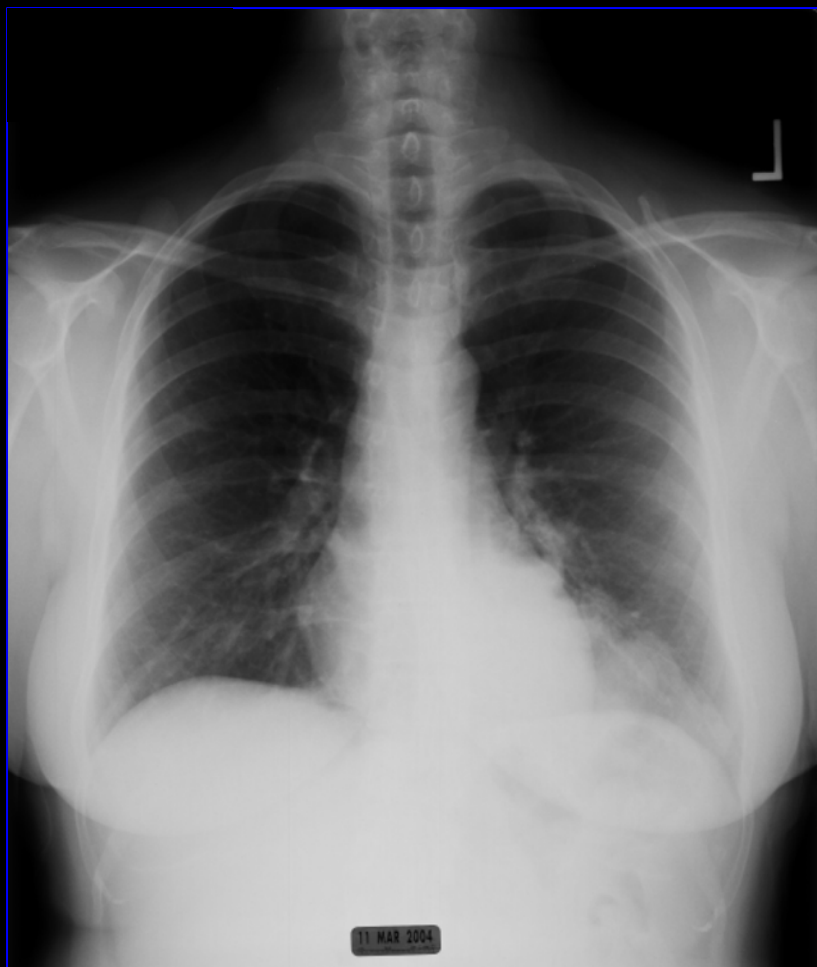
Findings:

- Ill-defined density in posterior LLL
- several rounded densities, some with air-fluid level



Patient 1

50 y.o. female with several months
of cough and hemoptysis



Wide differential for this infiltrate with
apparent cysts/cavitations includes:

- Bronchogenic carcinoma
- Metastatic Disease
- Abscess
- Infarction
- TB
- PCP
- Fungal infection
- Wegners
- Pulmonary sequestration

and more...



Patient 2

CT of Neonate with L chest mass on prenatal U/S



Courtesy of Dr. Fabio Komlos, Children's Hospital



Patient 2

CT of Neonate with L chest mass on prenatal U/S



Findings:

- Well-circumscribed heterogenous mass
- Posterobasal, above left hemi-diaphragm
- Arterial supply to mass from the aorta (*not visible on this slice*)



Patient 2

CT of Neonate with L chest mass on prenatal U/S



Differential of this congenital pulmonary mass includes:

- Diaphragmatic hernia
- Pulmonary sequestration
- CCAM (Congenital cystic adenomatoid malformation)
- Bronchogenic cyst
- Paraspinal lesion



What do the two patients have in common?

Patient 1 and 2 share a similar diagnosis--
admittedly uncommon, but not rare...

The clue: both patient's lesions are in the posterior left lower lung



What do the two patients have in common?

One of these diagnosis is characteristically
found in left lower lung...

Patient 1 DDx:

- Abscess
- Bronchogenic carcinoma
- Fungal infection
- PCP
- TB
- Metastatic
- Infarction
- Wegners
- Pulmonary sequestration

Patient 2 DDx:

- Diaphragmatic hernia
- Pulmonary Sequestration
- CCAM (Congenital cystic adenomatoid malformation)
- Bronchogenic cyst
- Paraspinal lesion



What do the two have in common?

When you encounter a persistent lesion in the left lower lung,
add this to your differential: **Pulmonary Sequestration**

Patient 1 DDx:

- Abscess
- Bronchogenic carcinoma
- Fungal infection
- PCP
- TB
- Metastatic
- Infarction
- Wegners
- **Pulmonary sequestration**

Patient 2 DDx:

- Diaphragmatic hernia
- **Pulmonary Sequestration**
- CCAM (Congenital cystic adenomatoid malformation)
- Bronchogenic cyst
- Paraspinal lesion



Pulmonary Sequestration

- A Congenital lung malformation:
 - A mass of abnormal, nonfunctioning, Pulmonary tissue
 - No communication with the tracheobronchial tree
 - Receives blood from an anomalous systemic artery (instead of pulmonary arterial system)
 - Usually occurs in left lower lung



Pulmonary Sequestration

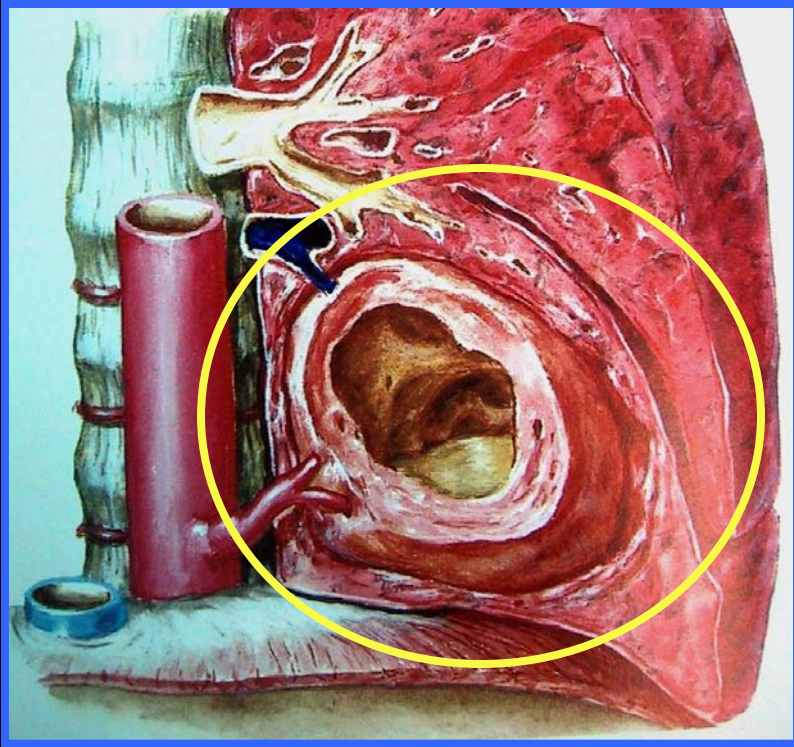
- Pathophysiology:
 1. Primitive foregut gives rise to accessory lung bud
 2. Pluripotent tissue migrates caudally with the developing normal lung
 3. Remains connected to aortic blood supply for the primitive foregut



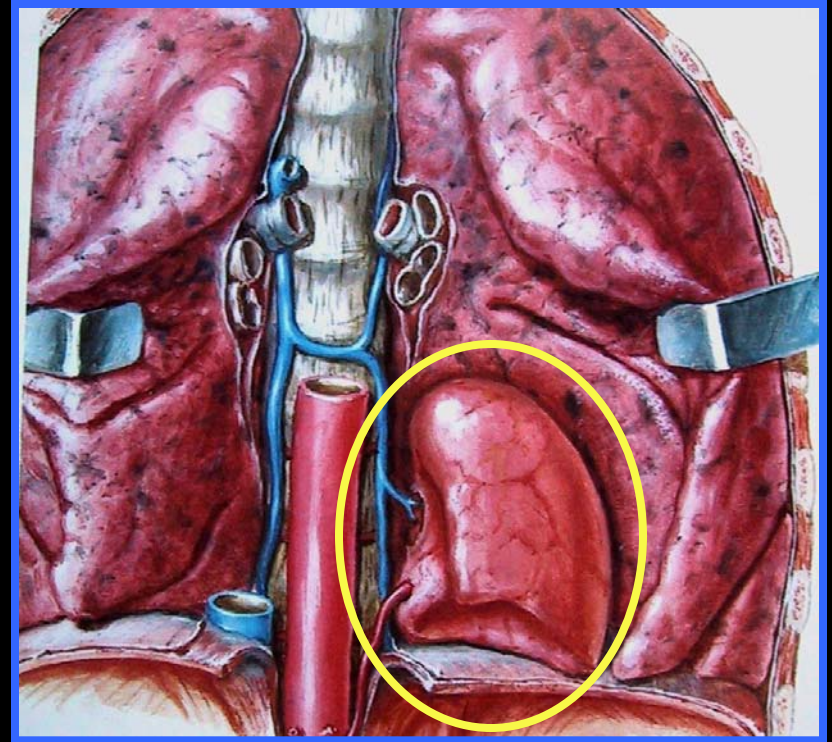
Two Types of Sequestration

- **Intralobar (75%)** = **WITHIN** visceral pleura of a pulmonary lobe
- **Extralobar (25%)** = “Accessory lung”—tissue in its own pleura

Intralobar vs. Extralobar

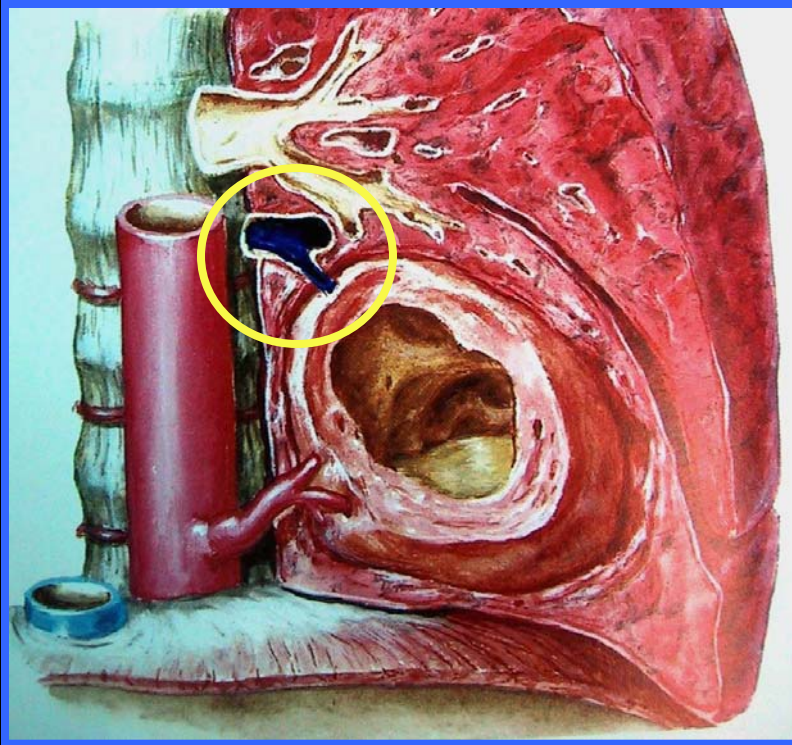


WITHIN visceral pleura of a pulmonary lobe

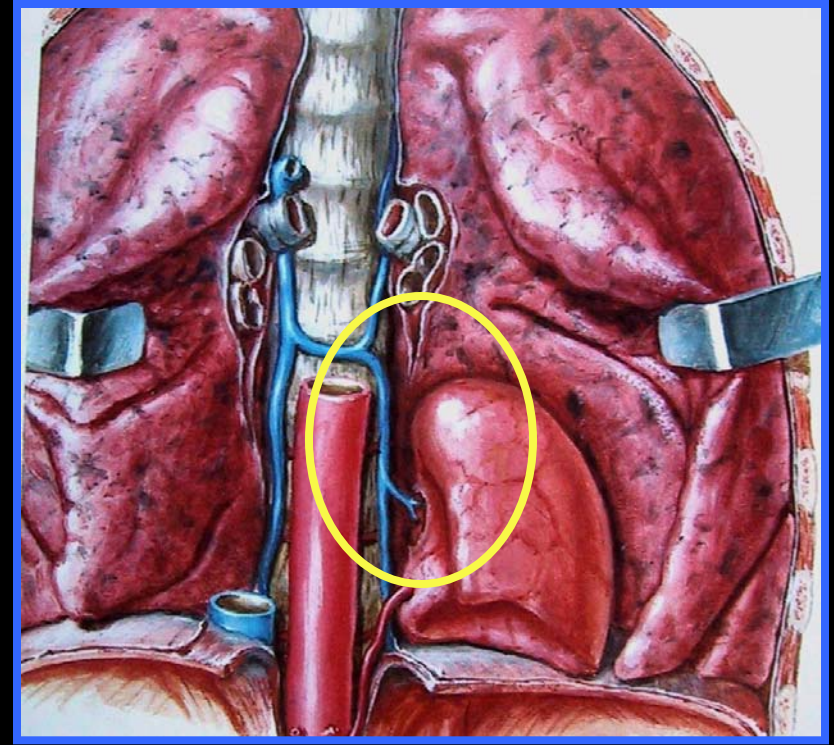


•“Accessory lung”: lung tissue in its own pleura

Intralobar vs. Extralobar

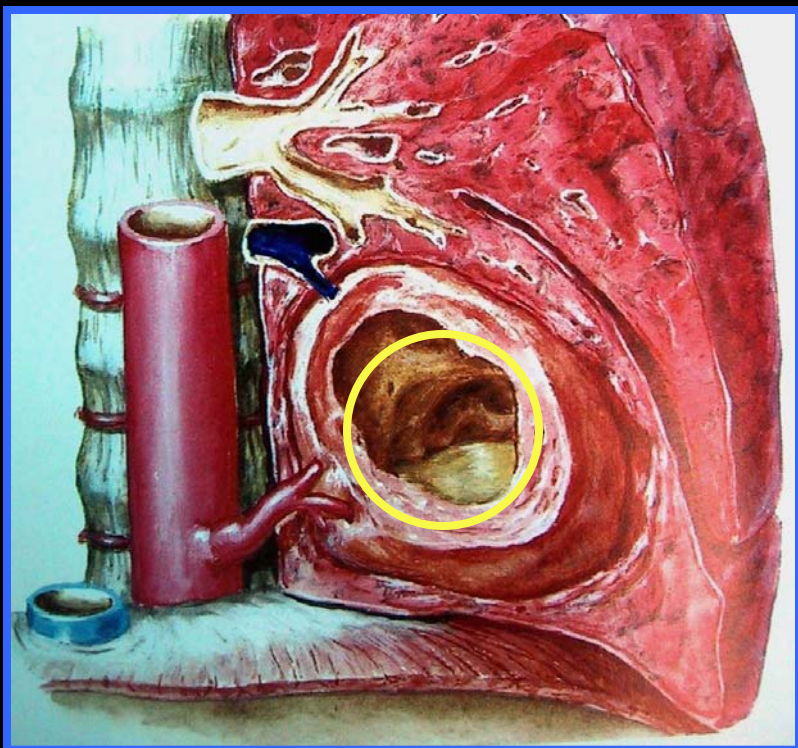


Venous Drainage into
pulmonary veins
(L→L recirculation)

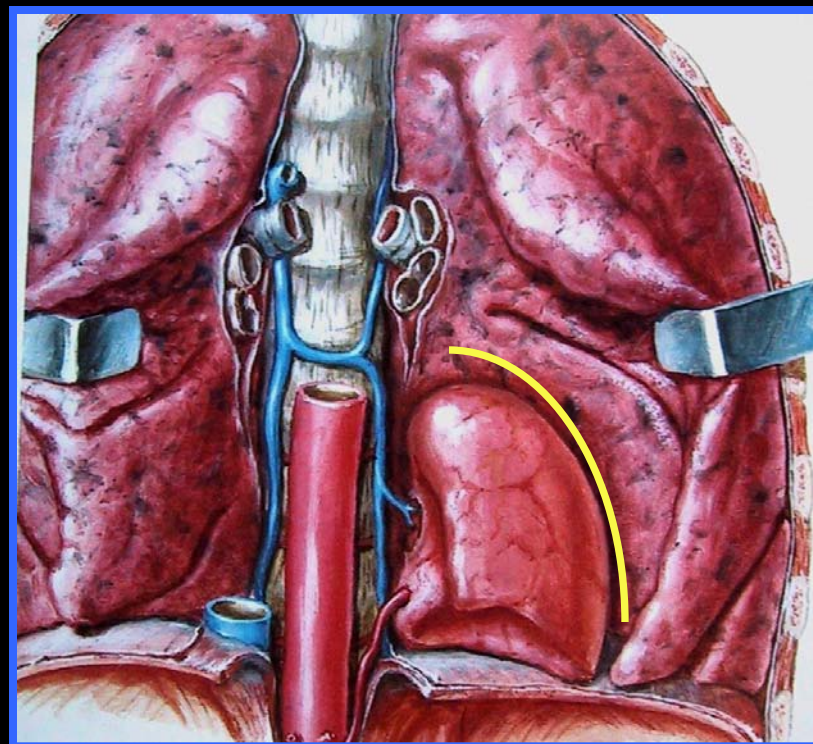


Venous Drainage into
systemic (hemiazygous)
(L→R shunt)

Intralobar vs. Extralobar



Cystic, frequently infected—
air gets in thru pores of Kohn
between adjacent alveoli



Less likely to get infected—
isolated from lung



Intralobar vs. Extralobar

Diagnosis usually made in
adolescence or adulthood...

Presenting with recurrent
pneumonia

Diagnosis usually made in
neonates or infants...

Often asymptomatic, but
discovered during evaluation
of other anomalies...

If not diagnosed
prenatally/neonatally, usual
presents by 6 months with
cyanosis or difficulty feeding



Intralobar vs. Extralobar

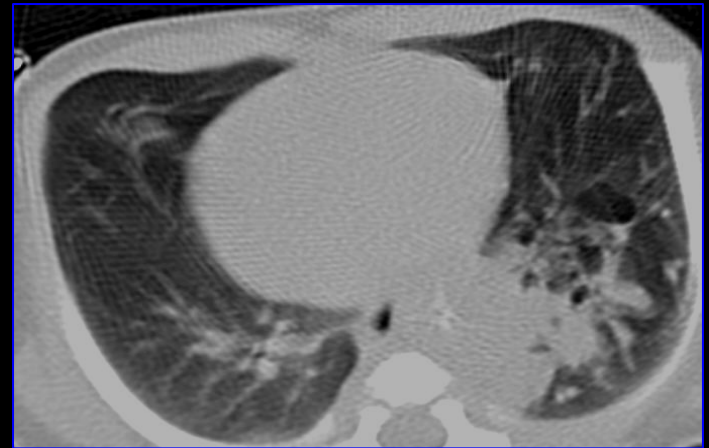
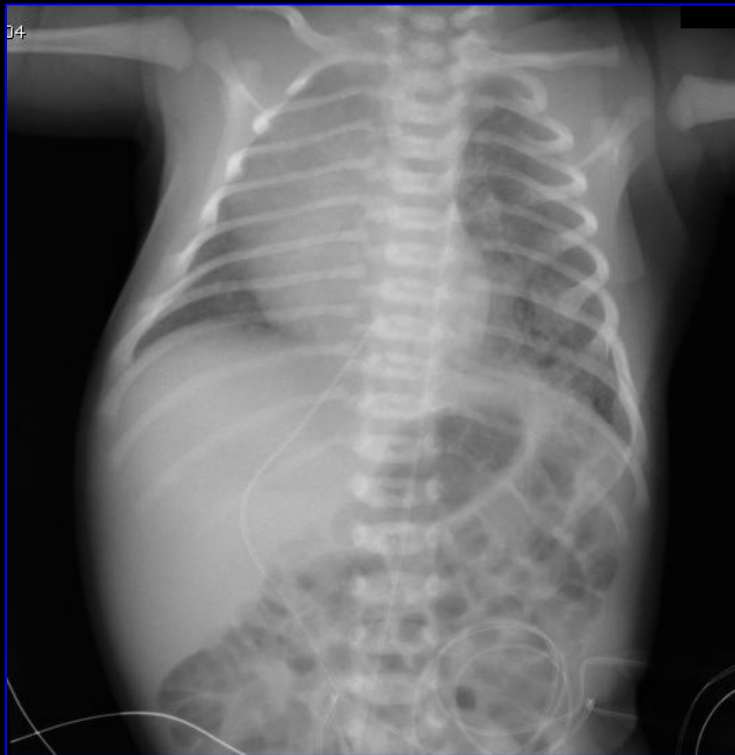
Not associated with other
anomalies

Often associated with:

- Diaphragmatic hernias
- Cardiac malformations
- Foregut anomalies



e.g. Neonate with Dextrocardia and Extralobar Sequestration





Which type of Sequestration?

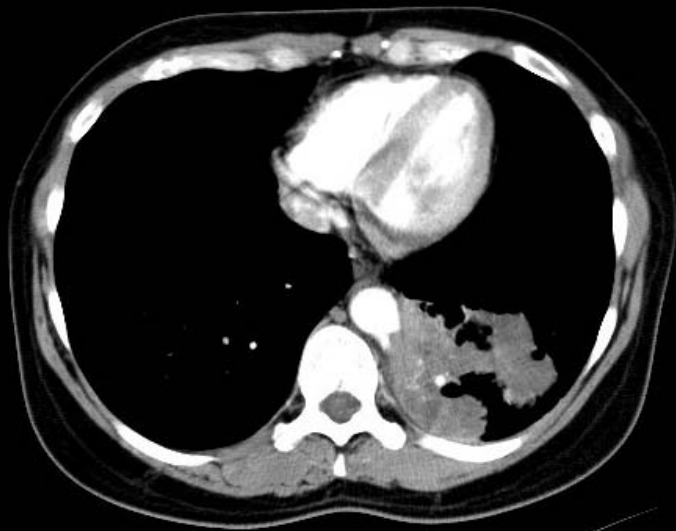
Intralobar

or

Extralobar

Patient 1:

50 y.o. female with several months cough and hemoptysis



PACS, BIDMC

Patient 2:

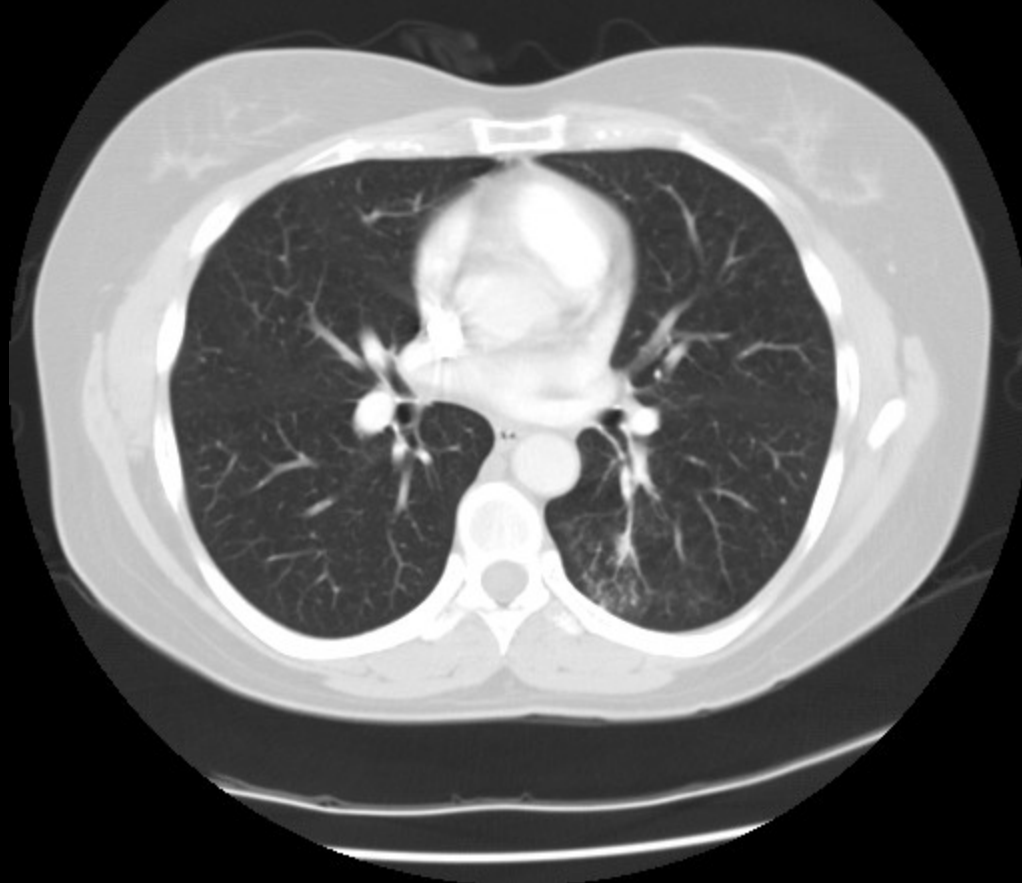
Asymptomatic neonate with mass found on prenatal U/S



Courtesy of Dr. Fabio Komlos, Children's Hospital

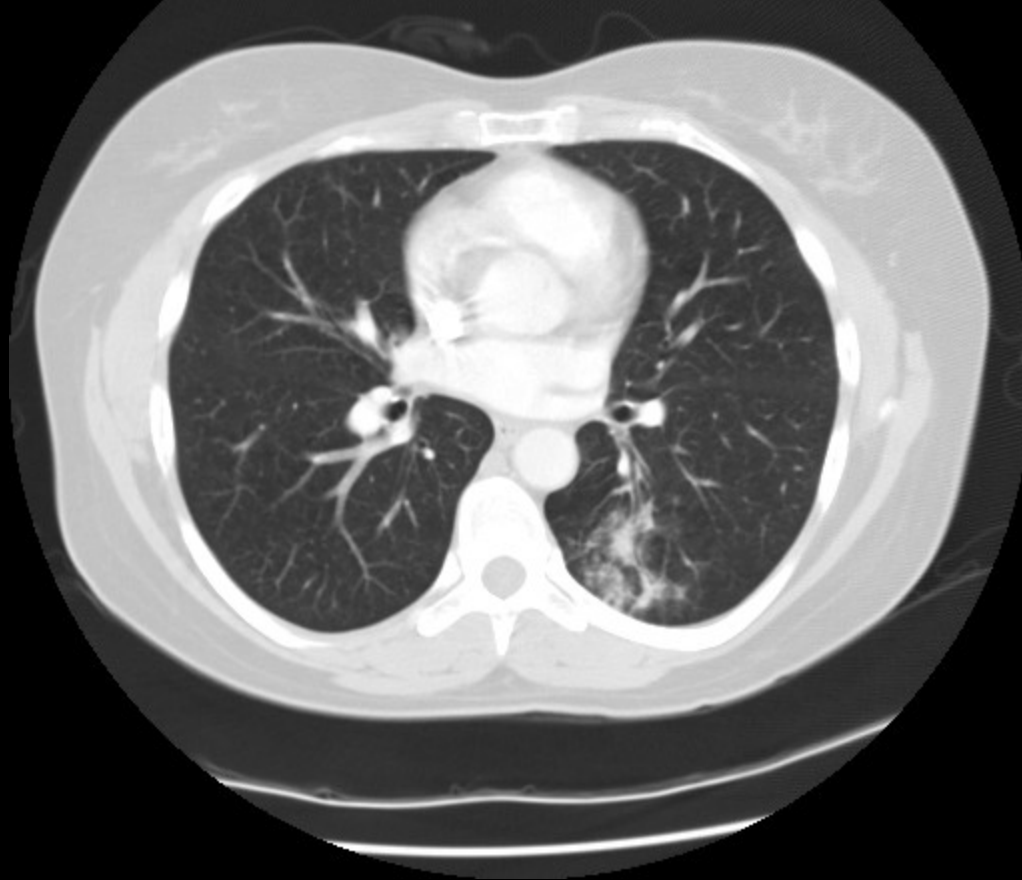


Patient 1 – Intralobar Sequestration



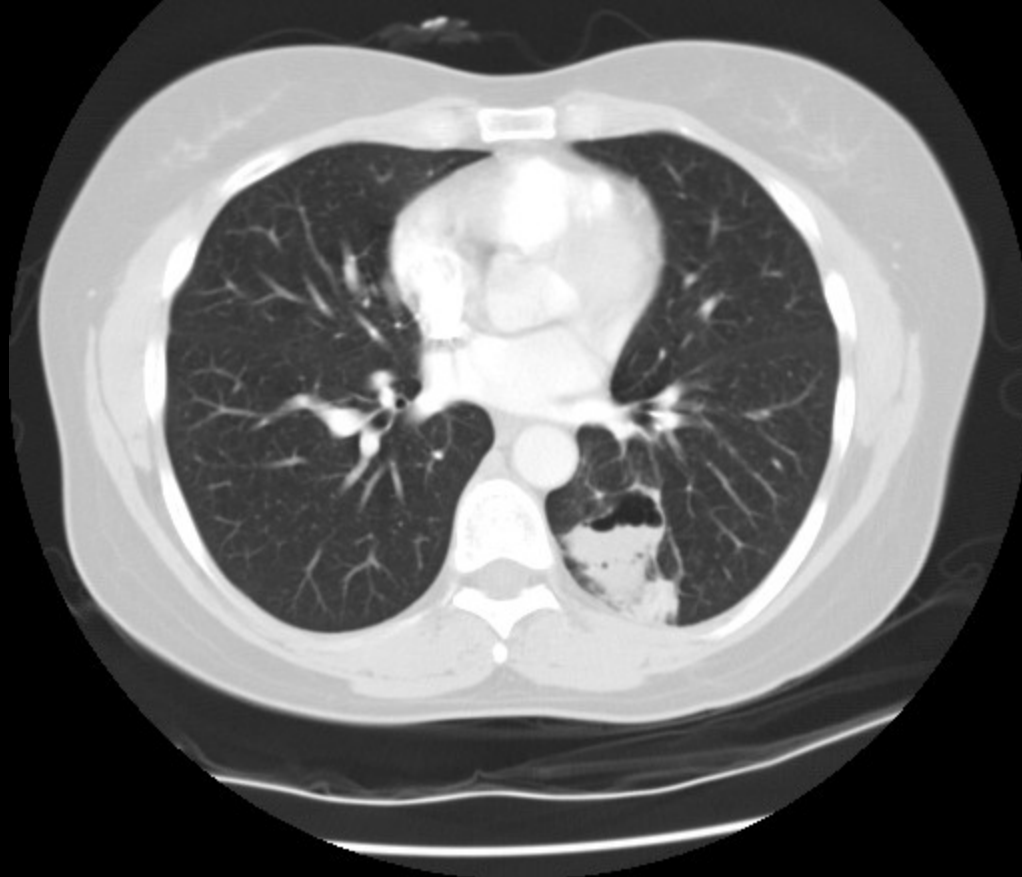


Patient 1 – Intralobar Sequestration





Patient 1 – Intralobar Sequestration





Patient 1 – Intralobar Sequestration



Patient 1 – Intralobar Sequestration





Patient 1 – Intralobar Sequestration

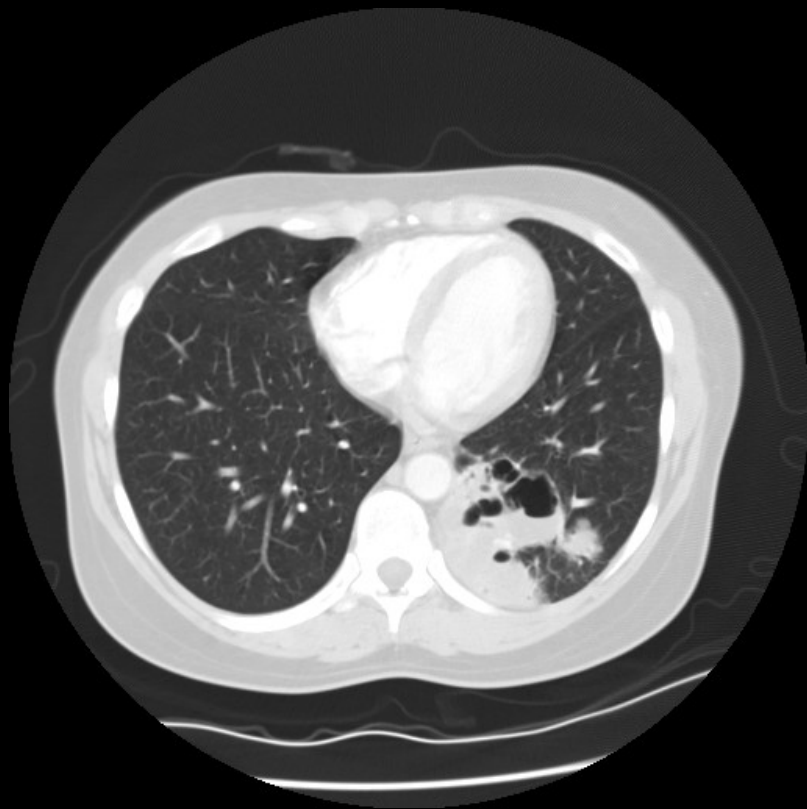




Patient 1 – Intralobar Sequestration



Patient 1 – Intralobar Sequestration



CT demonstrates typical findings of infected interlobar sequestration

- Dense, heterogeneous opacity within the posterobasal segment of the LLL
- Cystic appearance with multiple fluid-air levels

Patient 1 – Intralobar Sequestration



5 months later, after extended
fluoroquinolone therapy

-Resolution of fluid

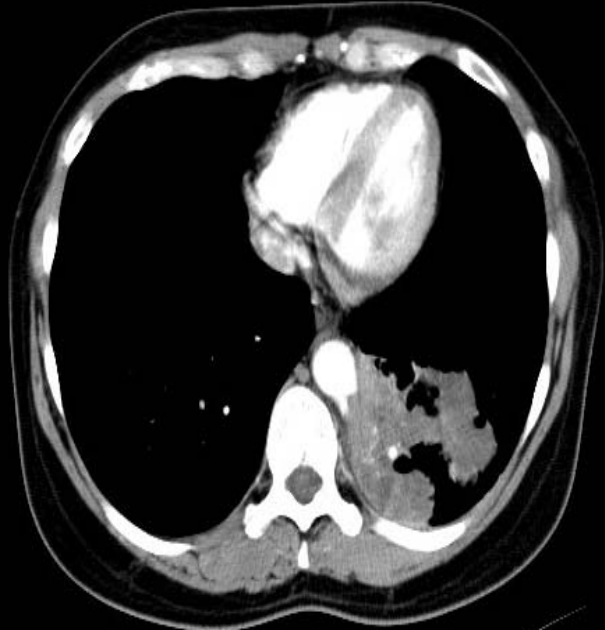

-persistent cystic spaces and
consolidation



Treatment

- Surgical resection for both intralobar and extralobar sequestrations, to avoid/treat
 - Chronic infection
 - Symptoms from compression of normal lung

Intralobar vs. Extralobar Anatomy

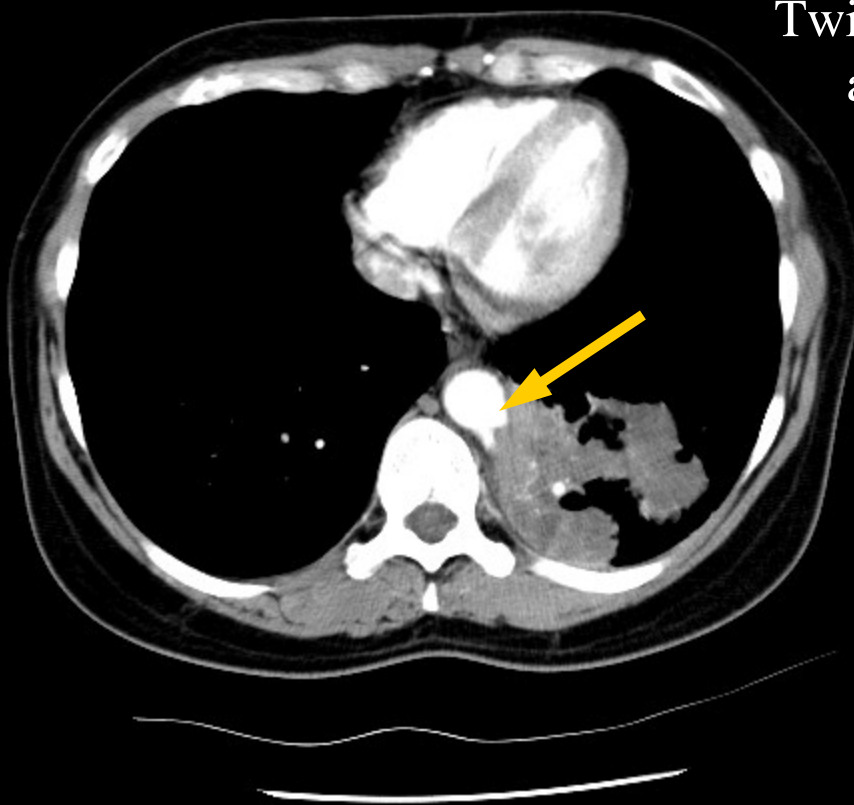
	Intralobar	Extralobar
Intra vs. Extra?	Within visceral pleura	Has own pleura— is an “accessory lobe”
		



Intralobar vs. Extralobar Anatomy

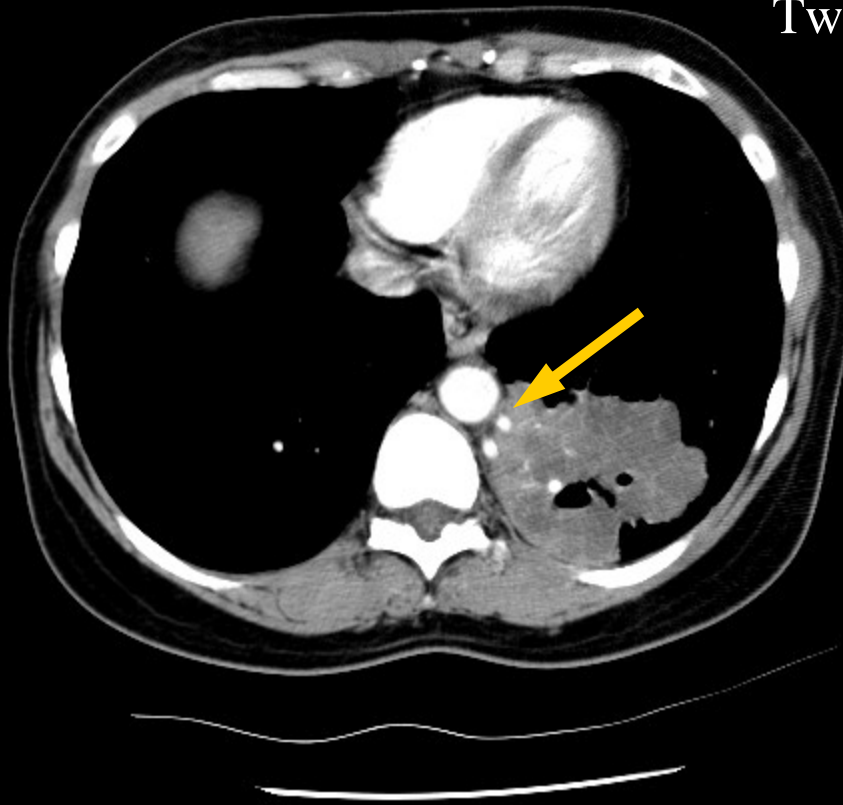
	Intralobar	Extralobar
Intra vs. Extra?	Within visceral pleura	Has own pleura— is an “accessory lobe”
Location	<u>in</u> Left Lower Lobe (67%)	<u>on</u> Left Hemidiaphragm (95%)
Arterial Supply	Aorta (Abdominal/thoracic)	Aorta (usually abdominal)

Patient 1 – CT with contrast (vessels)



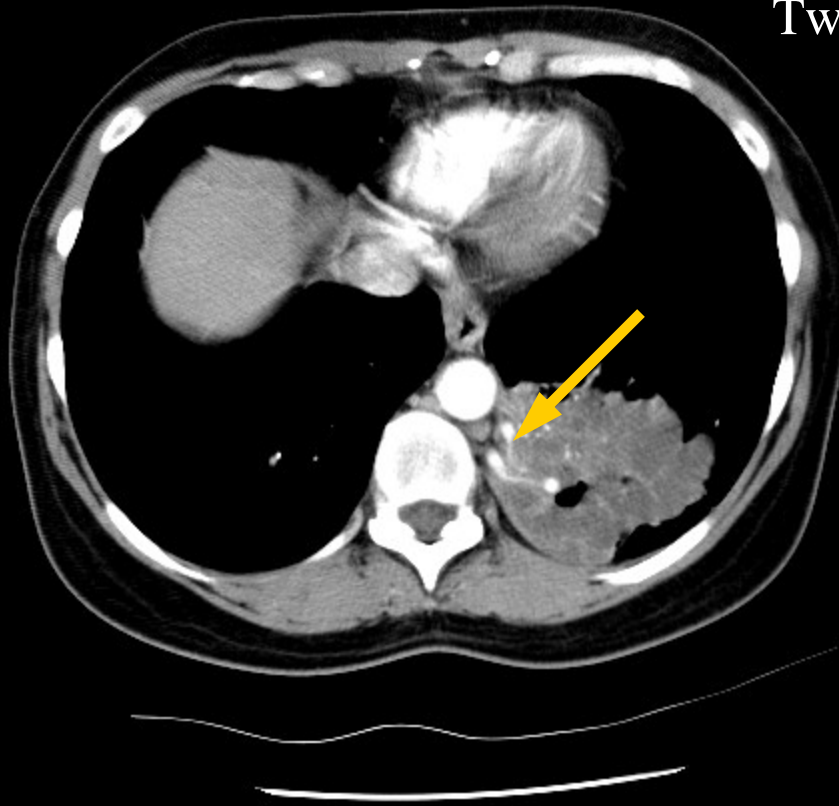
Twin anomalous arteries
aorta → sequestration

Patient 1 – CT with contrast (vessels)



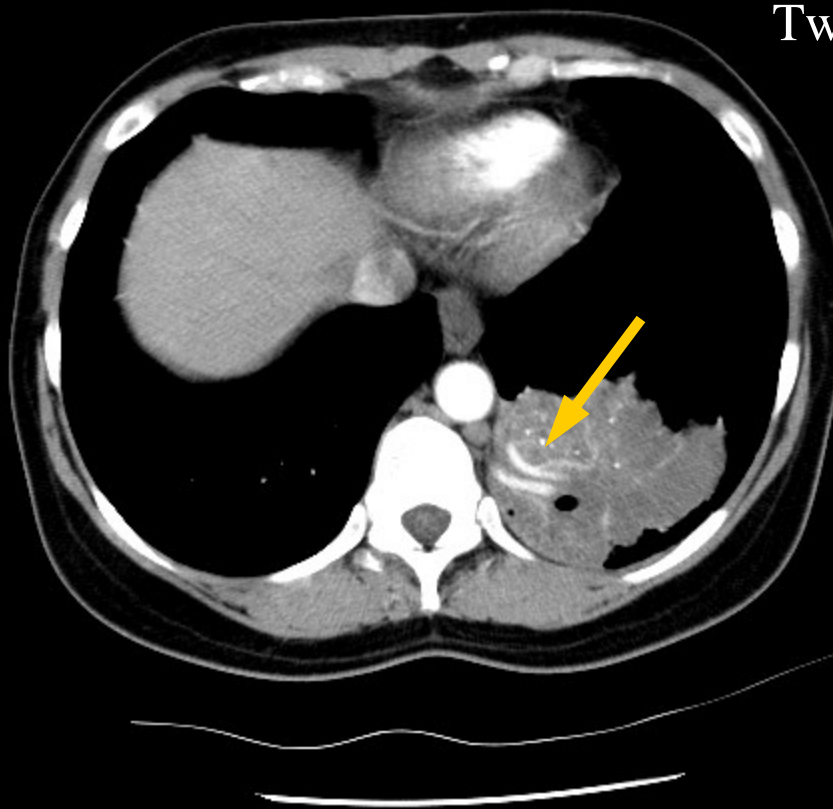
Twin anomalous arteries
aorta → sequestration

Patient 1 – CT with contrast (vessels)



Twin anomalous arteries
aorta → sequestration

Patient 1 – CT with contrast (vessels)



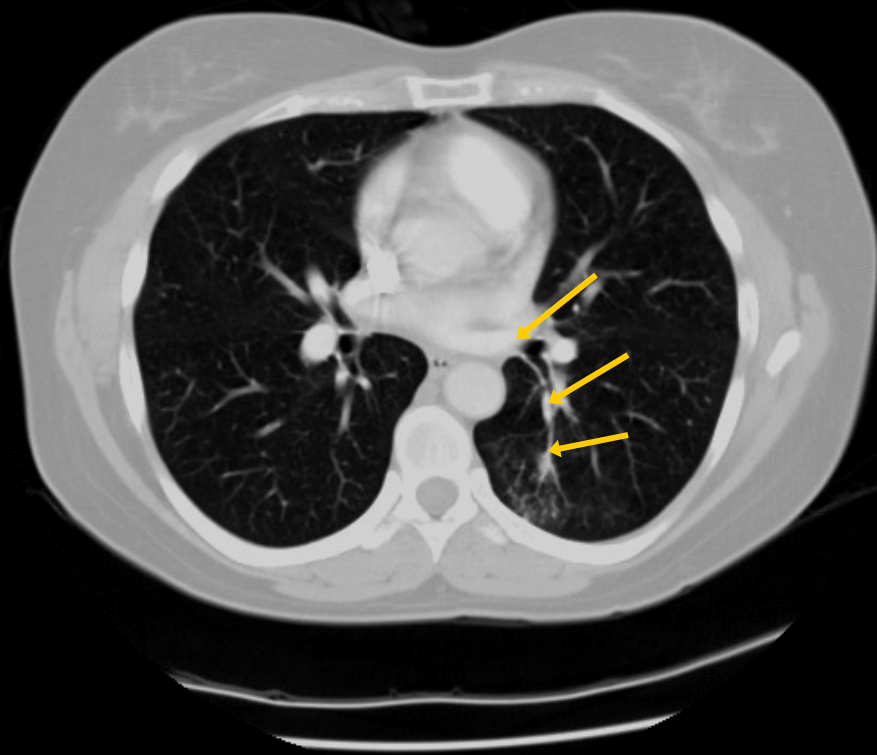
Twin anomalous arteries
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Intralobar vs. Extralobar Anatomy

	Intralobar	Extralobar
Intra vs. Extra?	Within visceral pleura	Has own pleura— is an “accessory lobe”
Location	<u>in</u> Left Lower Lobe (67%)	<u>on</u> Left Hemidiaphragm (95%)
Arterial Supply	Aorta (Abdominal/thoracic)	Aorta (usually abdominal)
Venous Return	Pulmonary Veins	Systemic Veins

Patient 1 – CT with contrast (vessels)



- Venous drainage into pulmonary veins
- Characteristic of **intralobar sequestration**



Imaging Pulmonary Sequestrations

- Identifying anomalous vessels = key to definitive diagnosis
- Useful for planning surgical resection

Variety of arterial patterns:

- »Descending Thoracic aorta (usually)
- »Infradiaphragmatic aorta (20%)
- »Dual arteries (10%)
- »Coronary arteries (rare reports)

Imaging Pulmonary Sequestrations

Various techniques have been used...

Definitive Diagnosis traditionally by

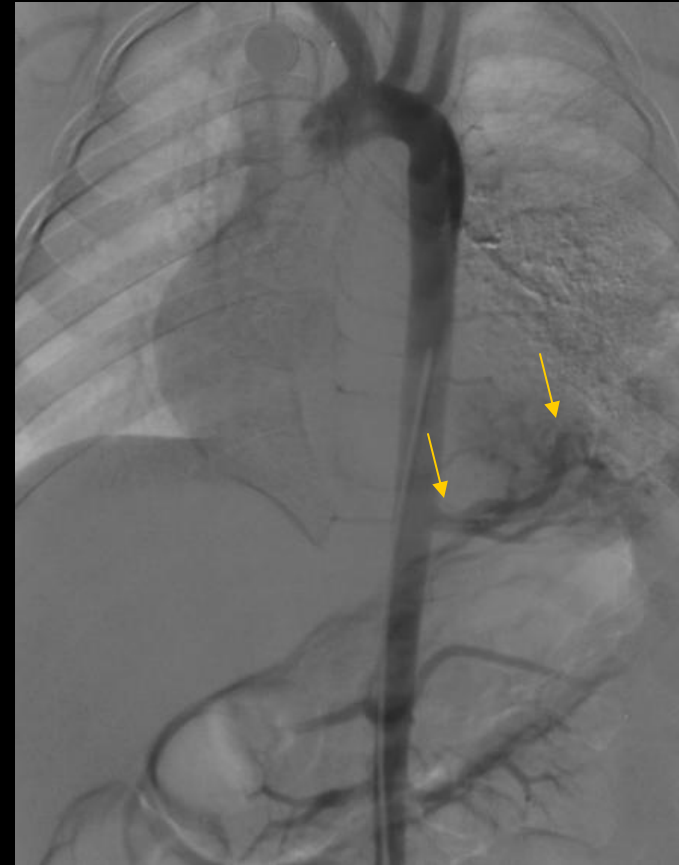
- **Invasive Arteriography**



Imaging Pulmonary Sequestrations

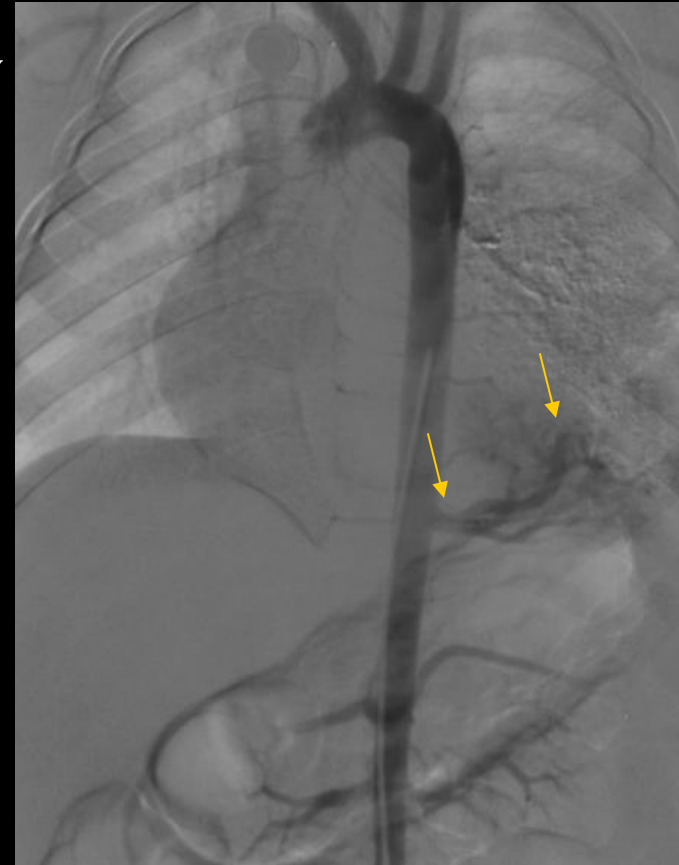
Aortagram:

- Anomalous vessel from infra-diaphragmatic aorta to density at left lung base
- Venous phase (not shown): drainage to the hemiazygous.
- Typical for an **extralobar sequestration**



Imaging Pulmonary Sequestrations

Conventional angiography is largely being replaced by non-invasive techniques: **CT, U/S, and MRI**





CXR for Sequestrations

- Remains an important screening tool
- But doesn't show the vessels

Radiographic Appearance of Intralobar Sequestrations

- **CXR** typically shows:
 - Lower lobe paraspinal opacity
 - Often cavitory or cystic

DDx

Pneumonia

Lung abscess

CCAM



Courtesy of Dr. Fabio Komlos, Children's Hospital

Intrapulmonary sequestration with large air-fluid level

Radiographic Appearance of Extralobar Sequestrations

- **CXR** typically shows:
 - Solid retrocardiac opacity
 - rounded or triangular
 - supra or sub-diaphragmatic

DDx (in neonate)

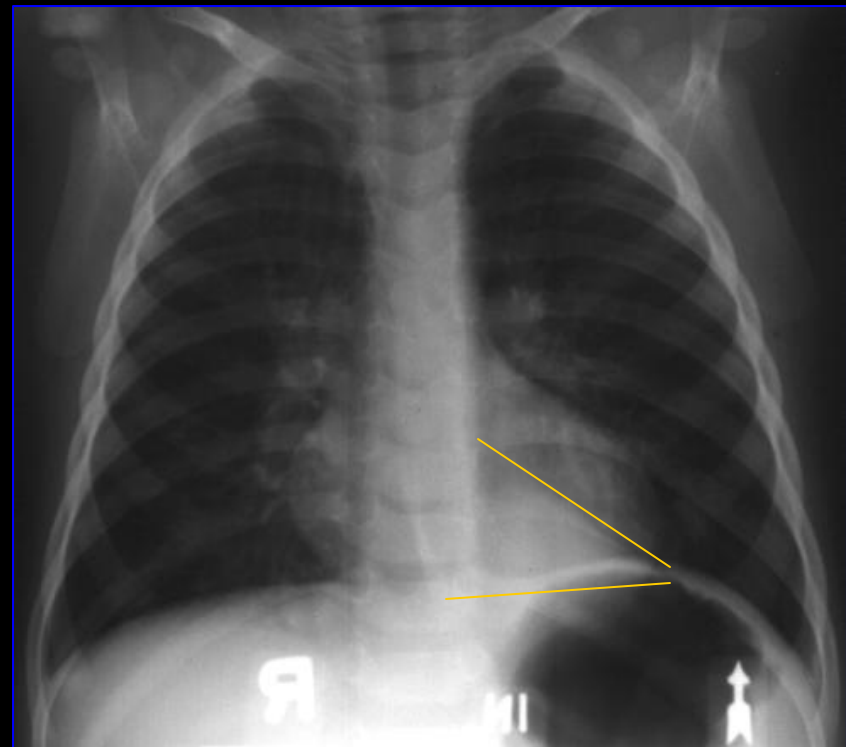
Diaphragmatic hernia

Loculated pleural effusion

Esophageal duplication cyst

Neuroblastoma

Adrenal hemorrhage



Triangular extralobar sequestration in a 13 month-old
p/w 1 month of wheezing, coughing and rhinorrhea.

CT Appearance of Sequestrations

- **CT/CTA**

- provides the best display of parenchyma

- Intralobar: mixed cystic/solid lesion or homogenous soft tissue.
 - Extralobar: well circumscribed homogenous lesion.

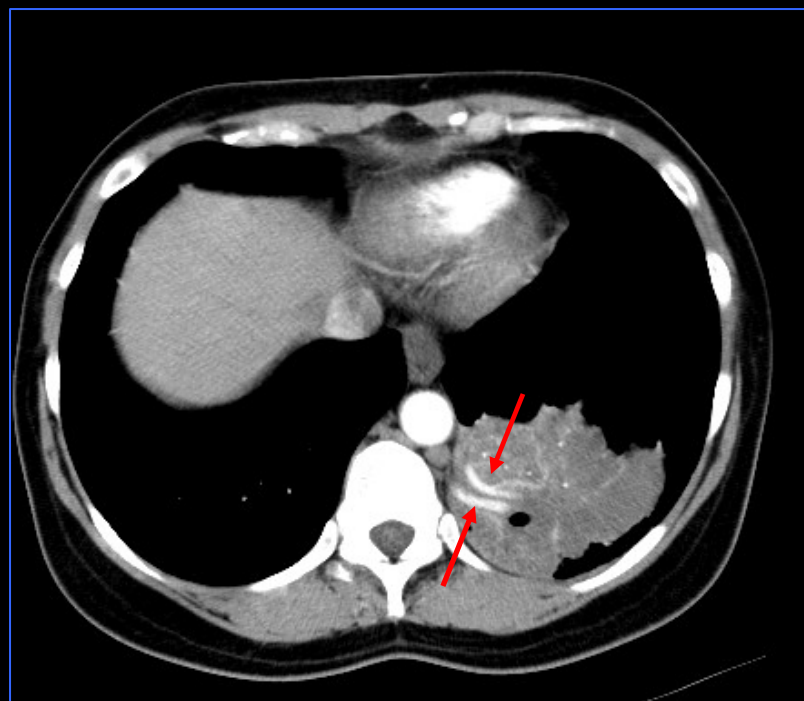


PACS, BIDMC

Patient 1- Intralobar Sequestration

CT Appearance of Sequestrations

- CT/CTA
 - typically can show the anomalous systemic arterial supply



PACS, BIDMC

Patient 1 – Arterial supply



CT Appearance of Sequestrations

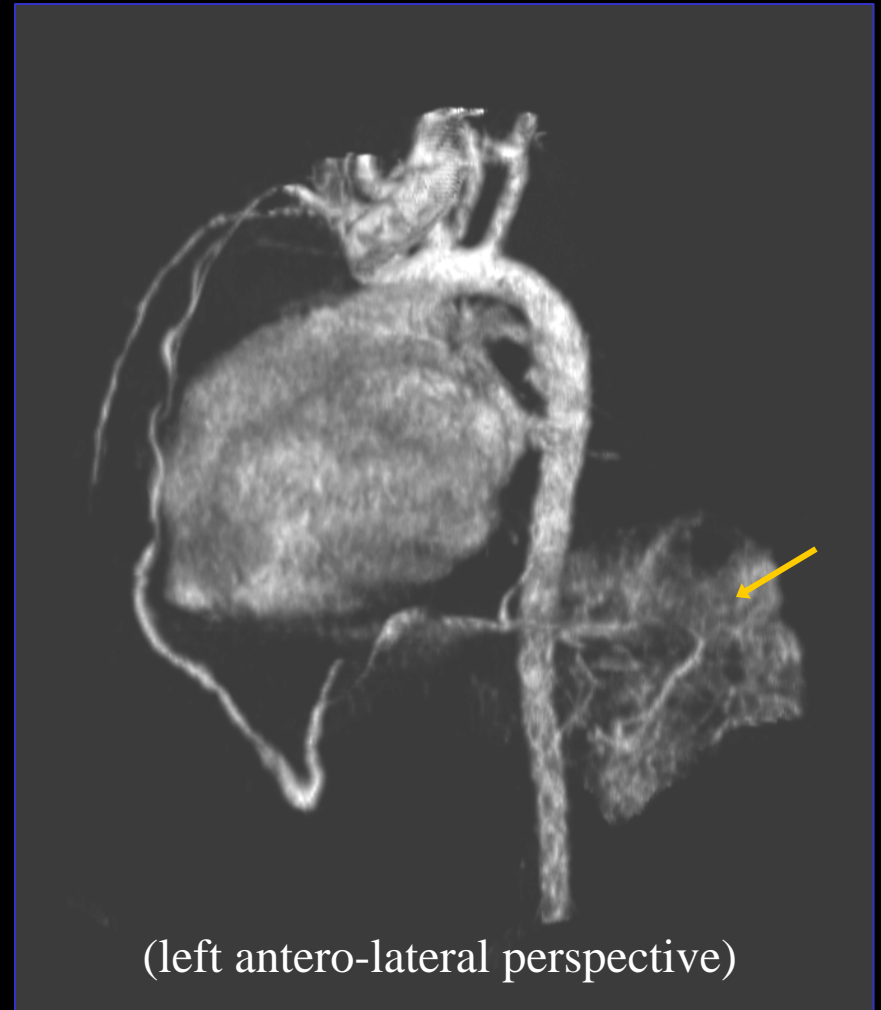
- Newer MultiDetector CT combined with 3D reconstruction can consistently identify both the arterial and venous anatomy



3D CT Reconstruction

Patient 2

Neonate with
extralobar sequestration



(left antero-lateral perspective)



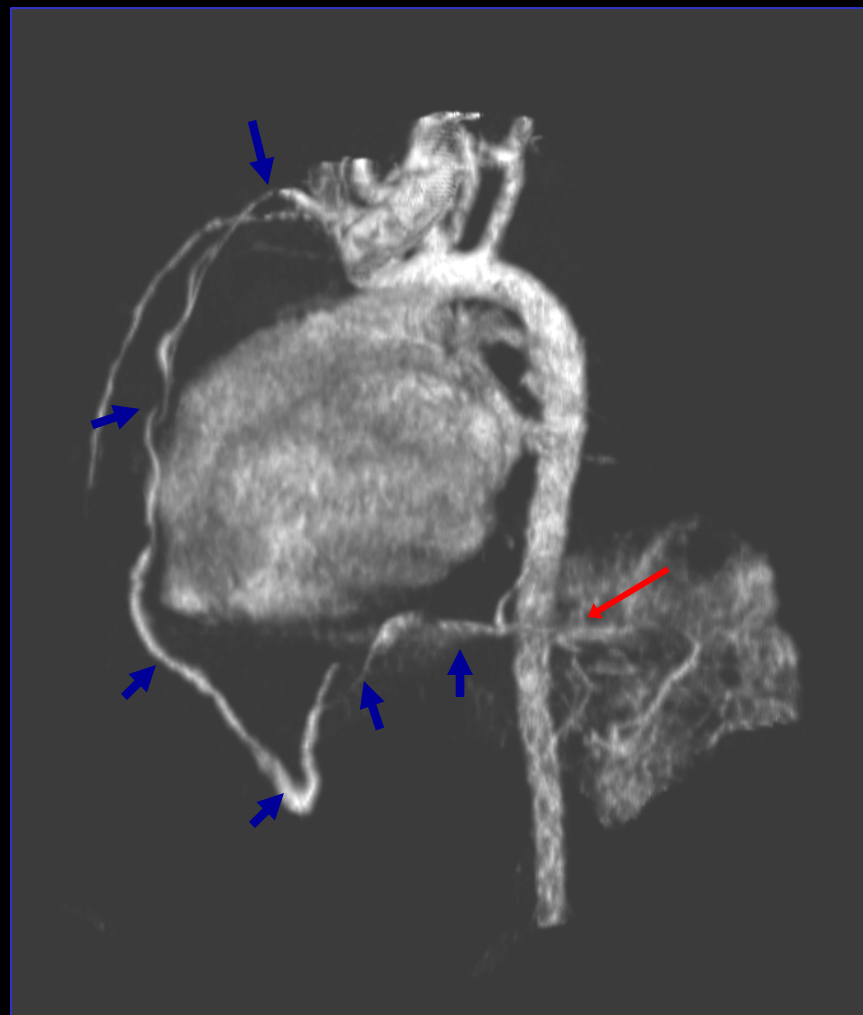
3D CT Reconstruction

Patient 2 – Neonate with extralobar sequestration

Not easily seen on axial scan

a 3-D reconstruction reveals:

- Typical **arterial supply** from the aorta
- Unusual systemic venous drainage via the **internal mammary vein**

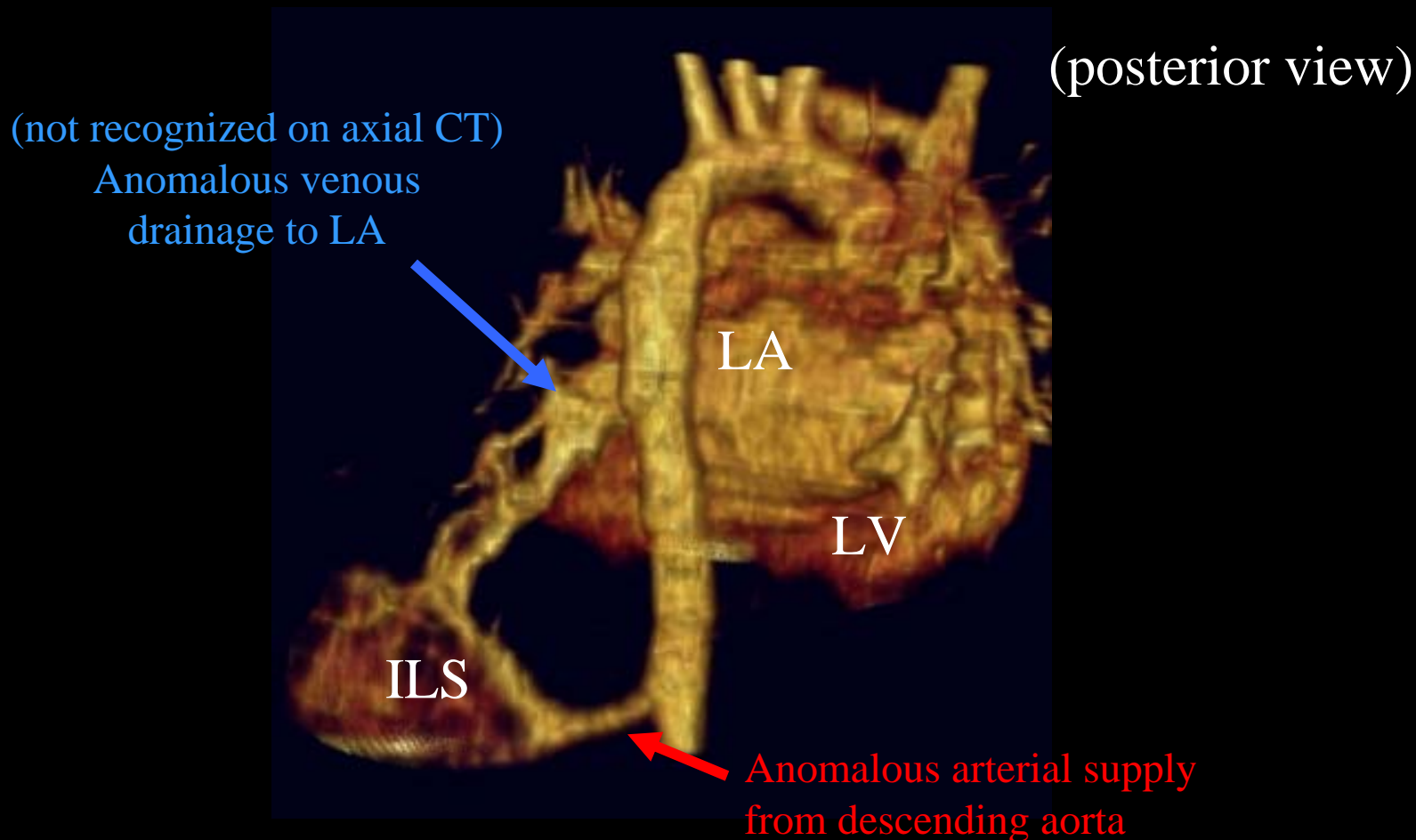


Courtesy of Dr. Fabio Komlos, Children's Hospital

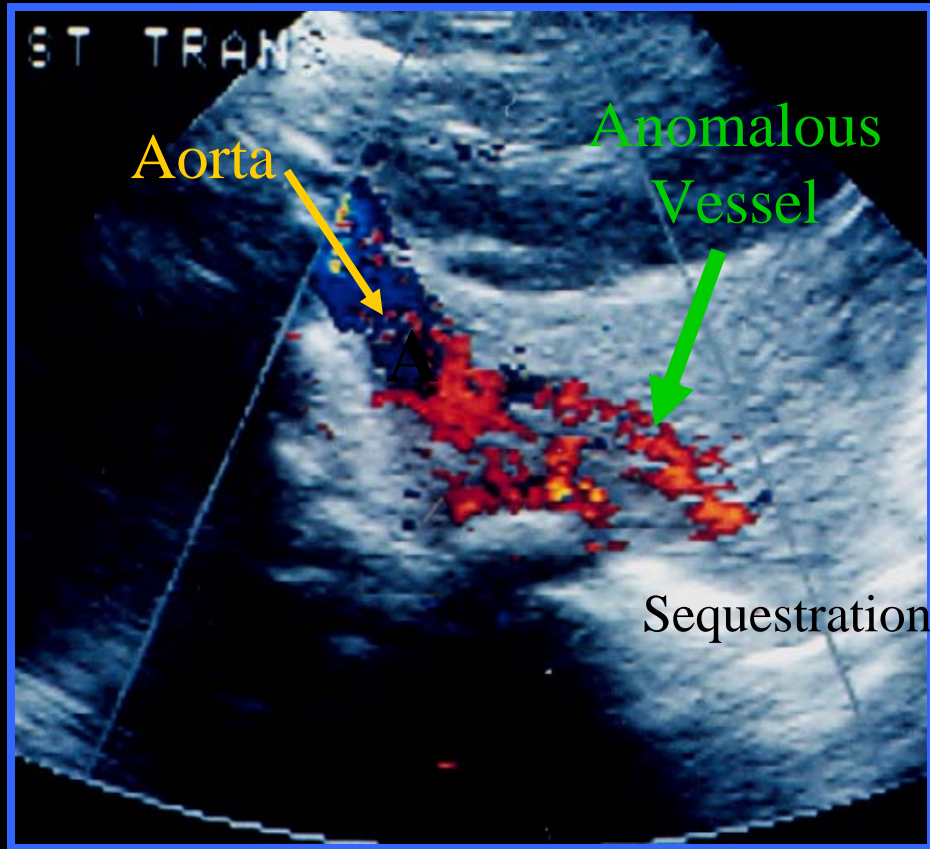


3D CT Reconstruction

A 6 month old girl with recurrent LLL PNA – Intralobar Sequestration



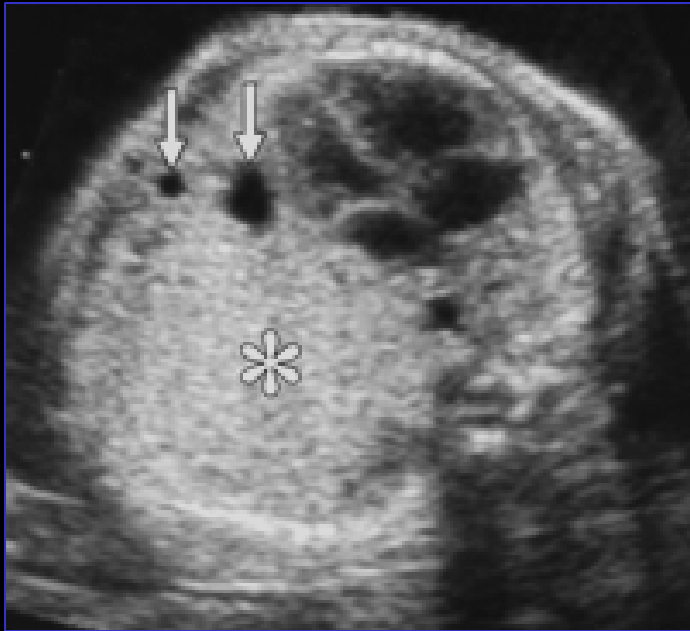
Sonogram of Sequestration



- May show anomalous vessel to the sequestration

Courtesy of Dr. Fabio Komlos, Children's Hospital

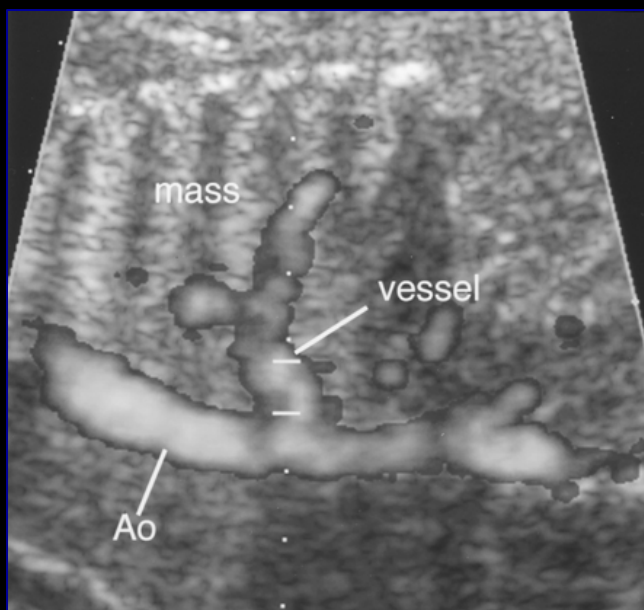
Sonogram of Sequestration



- Can also image prenatally

Axial sonogram of chest obtained at 22 weeks gestation in fetus with extralobar sequestration (*asterisk*). Cysts (*arrows*) are also visible.).

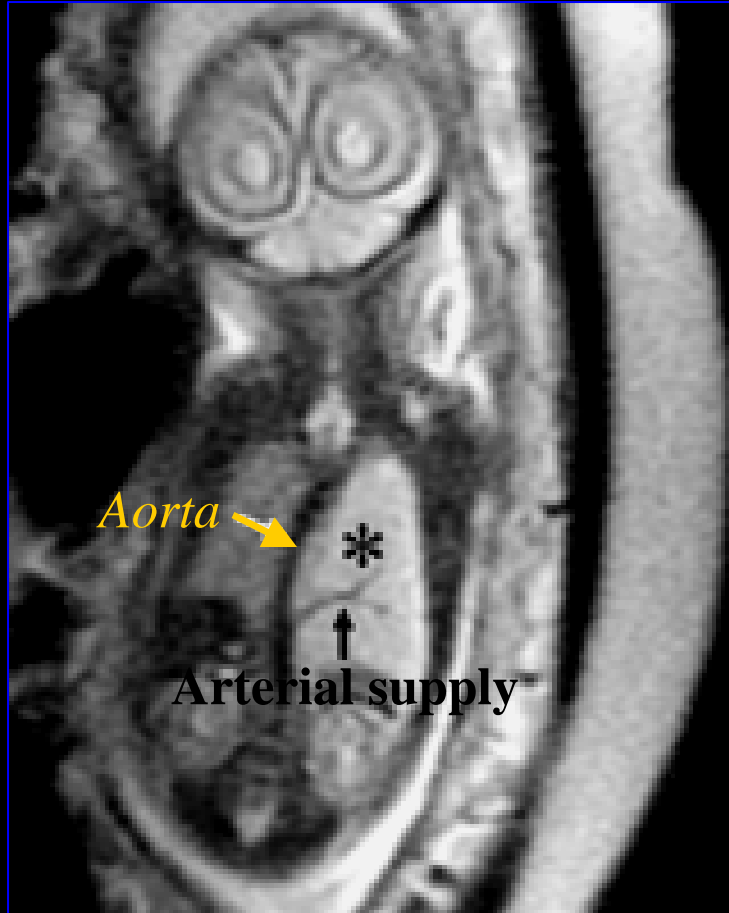
Sonogram of Sequestration



Sagittal sonogram of chest obtained at 32 weeks' gestation in fetus with extralobar sequestration (*mass*).

- Prenatal use of Doppler
- Distinguish sequestration from other congenital thoracic lesions

MRI of Sequestration



- MRI can also show precise anatomy of sequestrations (CT still better)
- MRI and U/S safe prenatally
- Accuracy still unknown

Coronal T2-weighted MRI obtained at 23 weeks' gestation, showing extralobar sequestration (*asterix*).



Recap

Intralobar

1. Adults/adolescents
2. Pulmonary veins
3. No anomalies

Extralobar

1. Neonates/infants
2. Systemic veins
3. Often other anomalies



Acknowledgements

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- Pamela Lepkowski
- Larry Barbaras



References

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