

**PULMONARY SEQUESTRATION IN SCIMITAR SYNDROME
DEMONSTRATED BY
THREE-DIMENSIONAL ROTATIONAL ANGIOGRAPHY**

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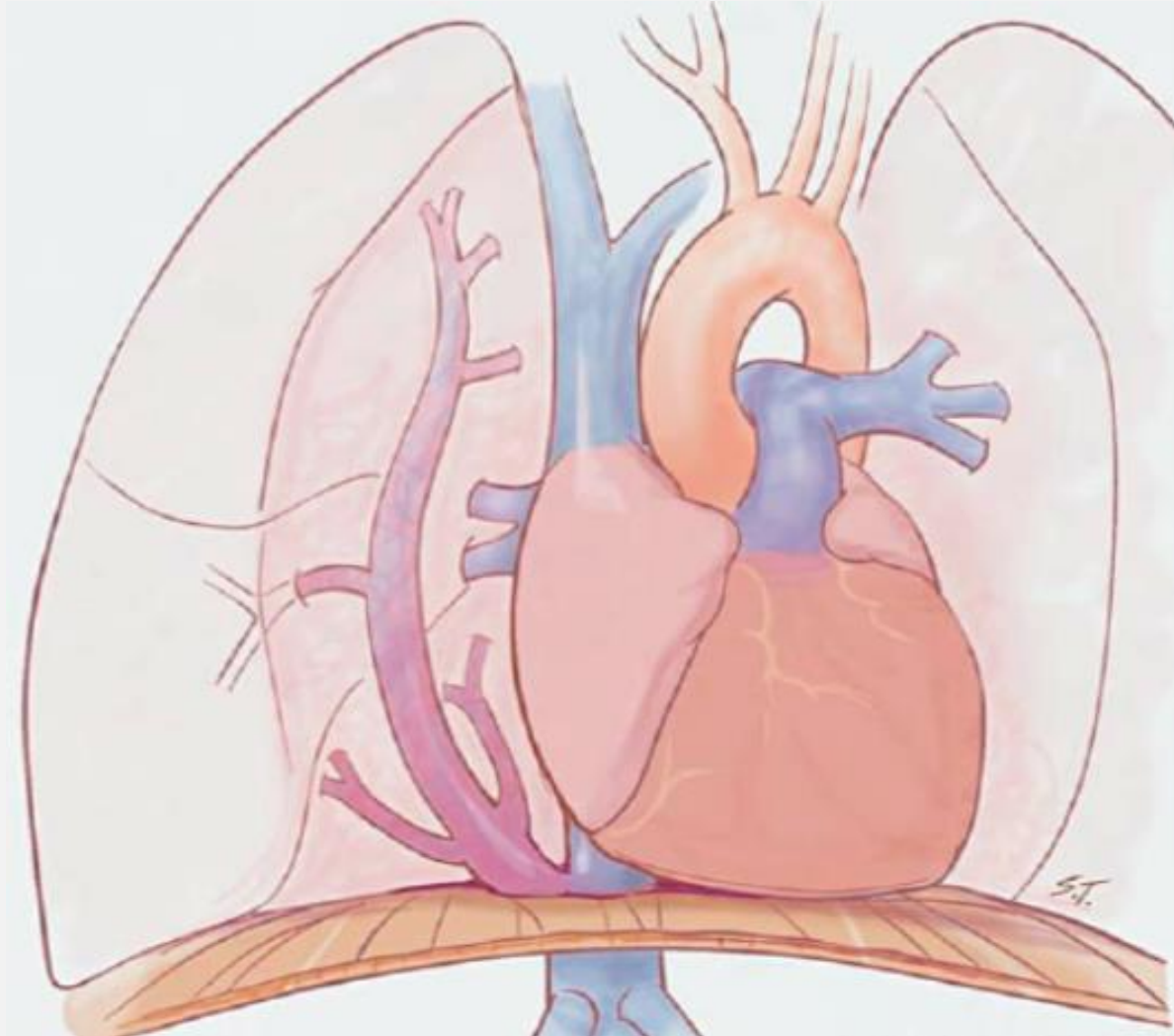
DISCLOSURE

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BACKGROUND

Scimitar Syndrome is a specific type of PAPVR and is a complex disorder consisting of

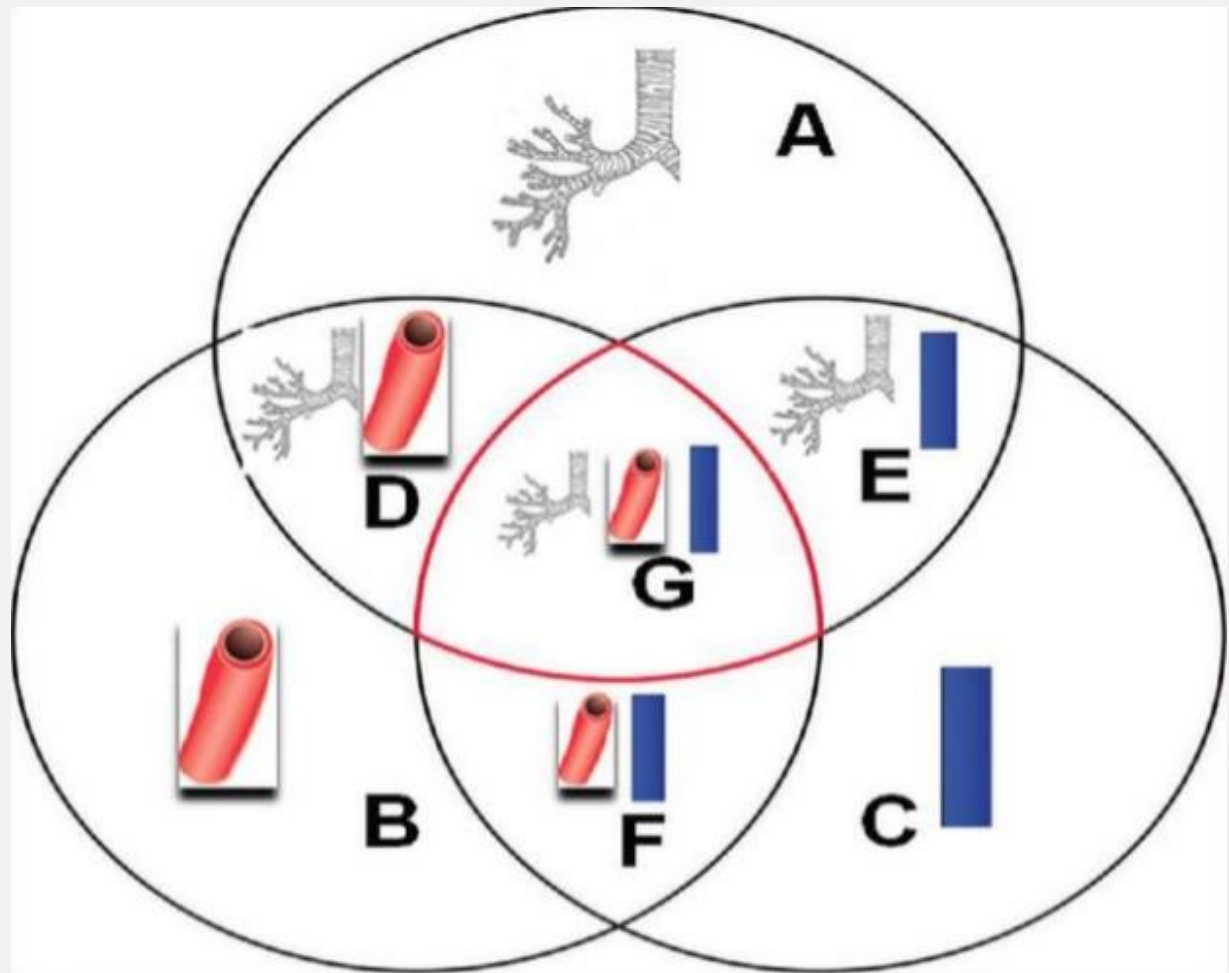
- ❖ Anomalous pulmonary venous drainage of the right lung to the IVC
- ❖ Systemic arterial supply of the right lower lobe from aorta
- ❖ Hypoplasia of the right lung
- ❖ Right pulmonary artery hypoplasia



SCIMITAR SYNDROME



PULMONARY MALINOSCUATION SPECTRUM



PULMONARY SEQUESTRATION

- ❖ Nonfunctioning primitive lung tissue
- ❖ Cystic or solid mass
- ❖ Has anomalous systemic blood supply.
- ❖ Does not communicate with the tracheobronchial tree

TYPES OF SEQUESTRATION

- ❖ Intralobar (75%)

WITHIN visceral pleura of a pulmonary lobe

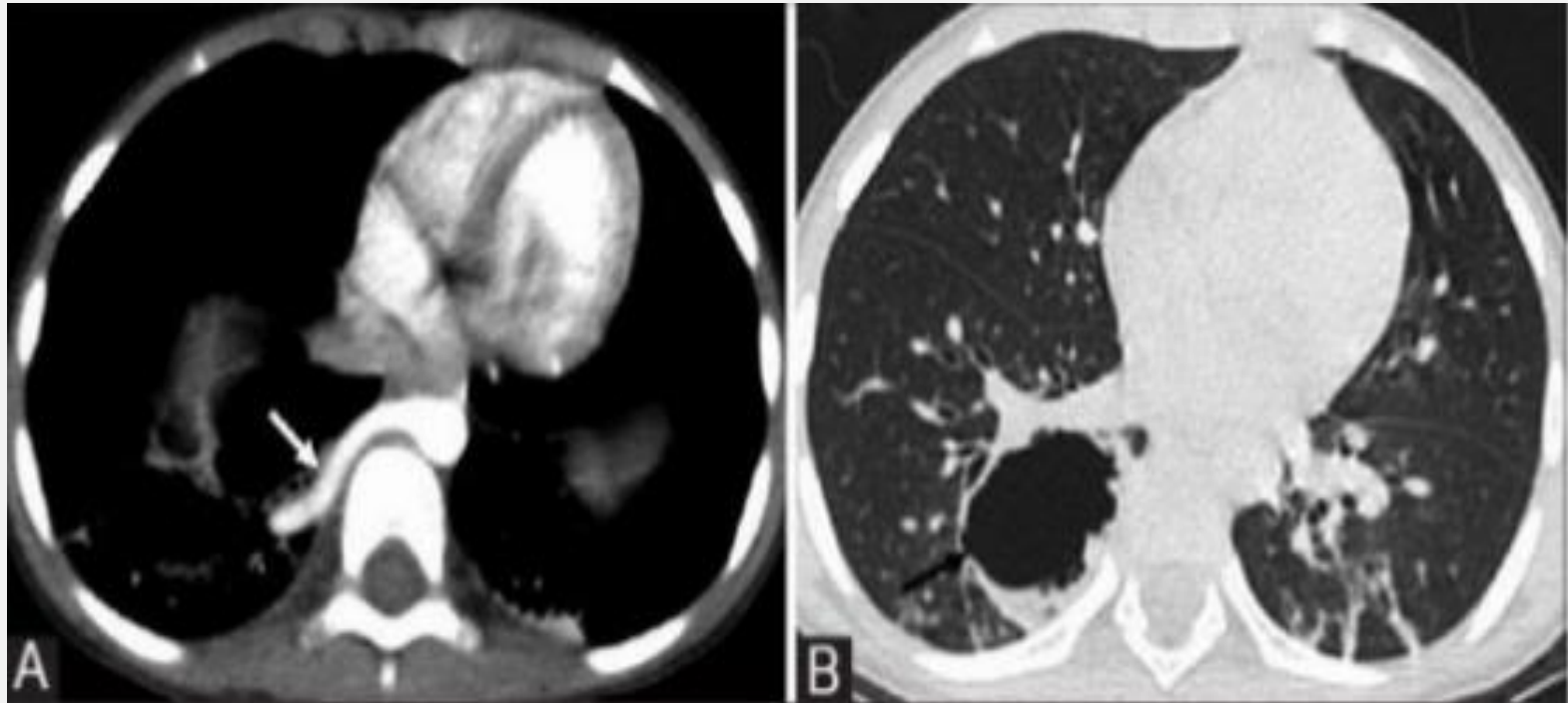
- ❖ Extralobar (25%)

“Accessory lung”— tissue in its own pleura

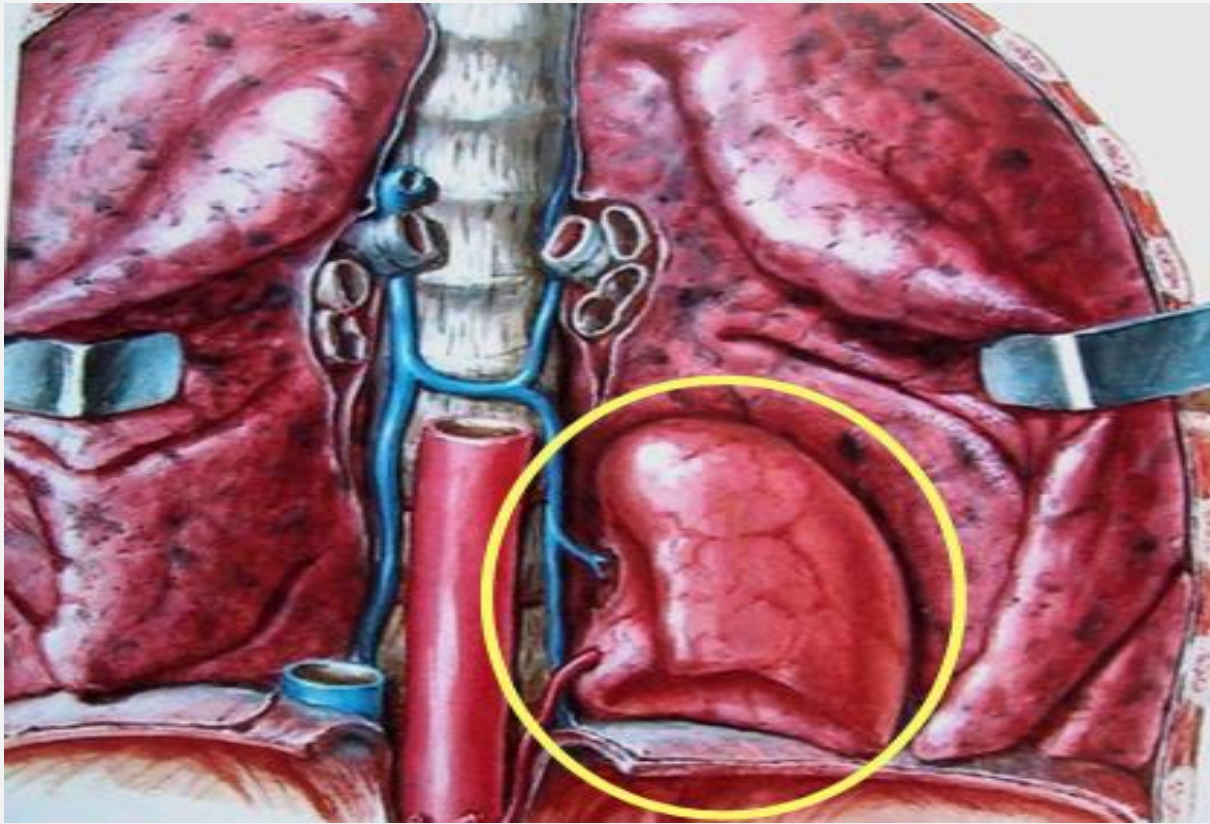
**INTRALOBAR
SEQUESTRATION**

CT Axial image

A- Mediastinal Window B- Lung window



EXTRALOBAR SEQUESTRATION



DIAGNOSIS

- ❖ 3DRA (3 Dimensional rotational angiography) has been used to define complex cardiovascular and airway anatomy.
- ❖ Can 3DRA help define areas of pulmonary sequestration

METHODS

- ❖ 16 year old female patient.
- ❖ Presented with two episodes of hemoptysis
- ❖ Diagnosed with PDA and Scimitar syndrome in Chicago in infancy

Had device closure of PDA
No further intervention

ECHOCARDIOGRAM

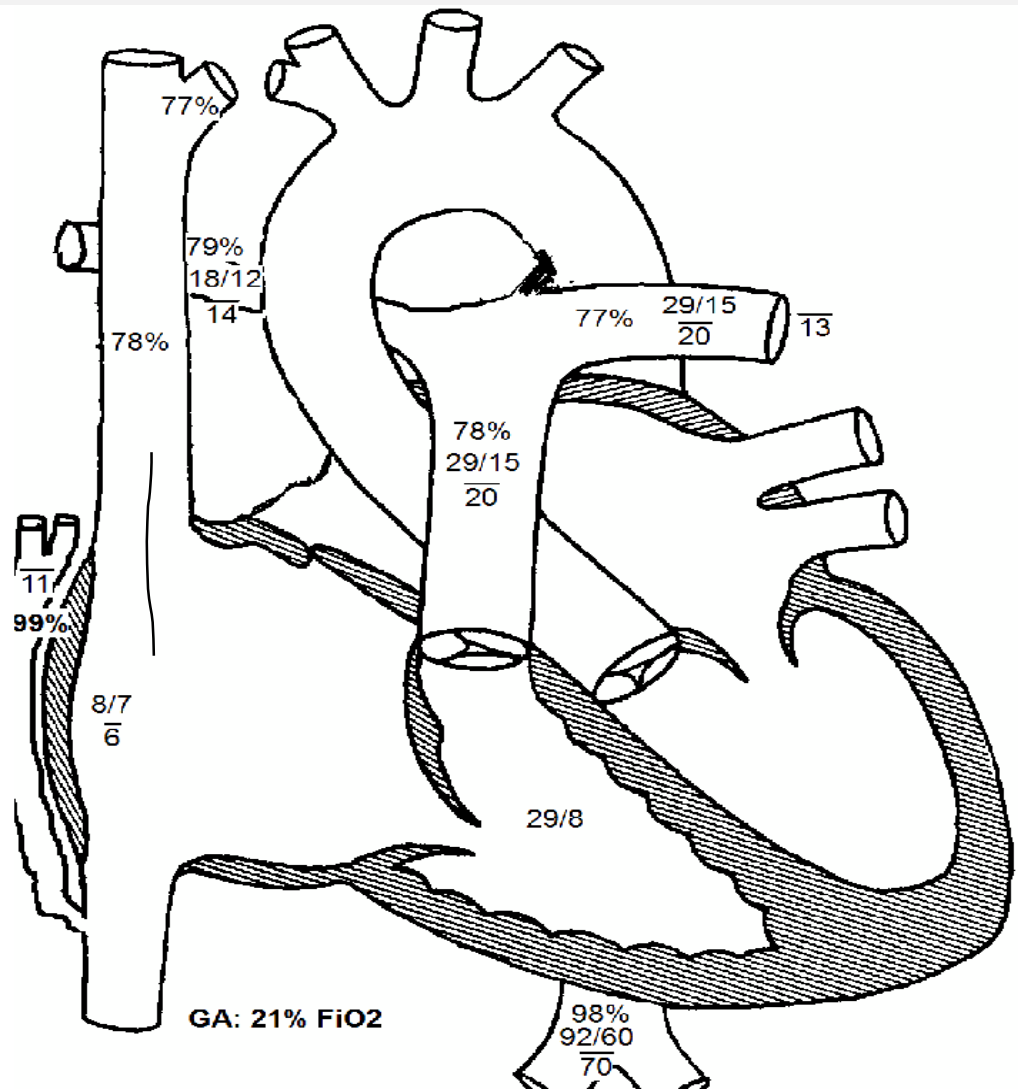
- ❖ Anomalous pulmonary venous drainage on right
- ❖ Dextroposition of heart
- ❖ Normal Biventricular size and subjective systolic function

CT SCAN

- ❖ Scimitar syndrome (hypogenetic right lung with variant bronchial anatomy, hypoplastic right pulmonary artery, and partial anomalous pulmonary venous drainage of right lower lobe to inferior vena cava).
- ❖ Area of airspace disease in the right lower lobe (pneumonia versus Focal pulmonary hemorrhage).
- ❖ No systemic arterial supply is demonstrated to suggest pulmonary sequestration.

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CARDIAC CATHETERIZATION

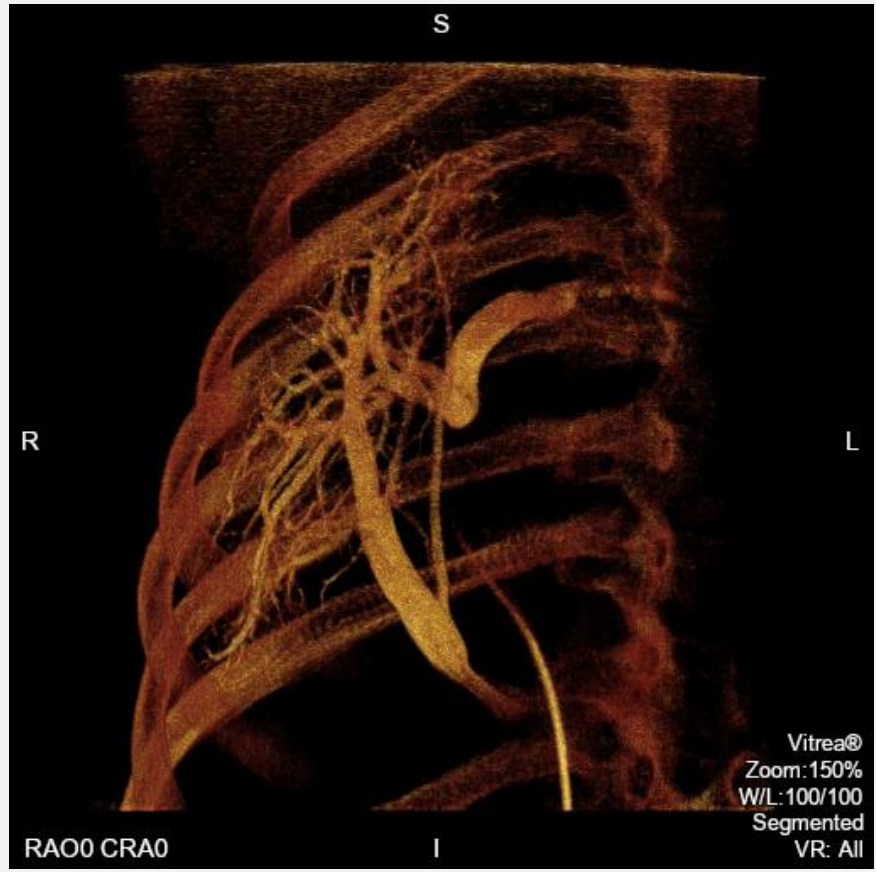


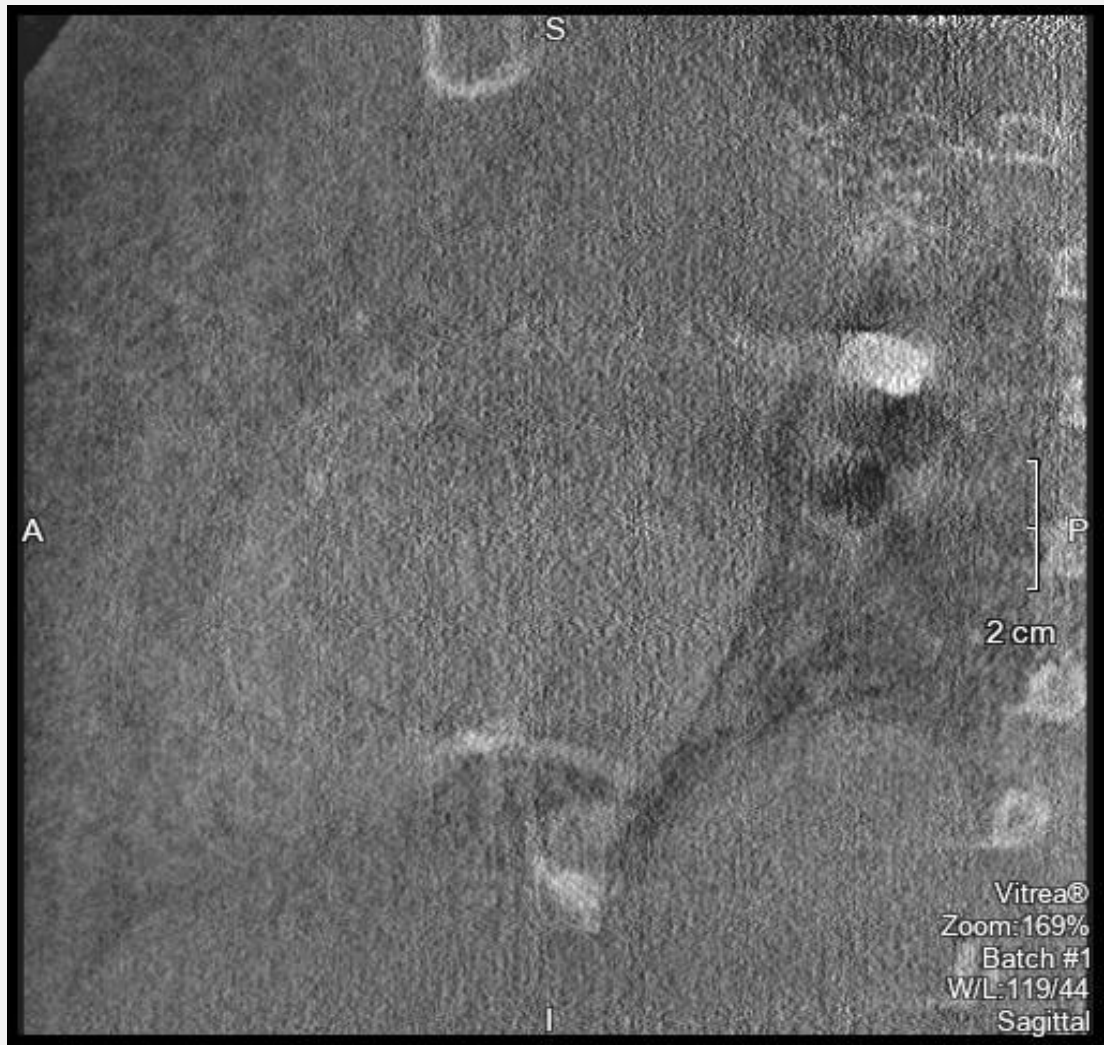
- ❖ No step up in Sats from SVC to RPA
- ❖ Normal PA Pressure
- ❖ Qp:Qs : 1.05.:1
- ❖ Pressure gradient of 5 mm Hg between Abn PV and RA

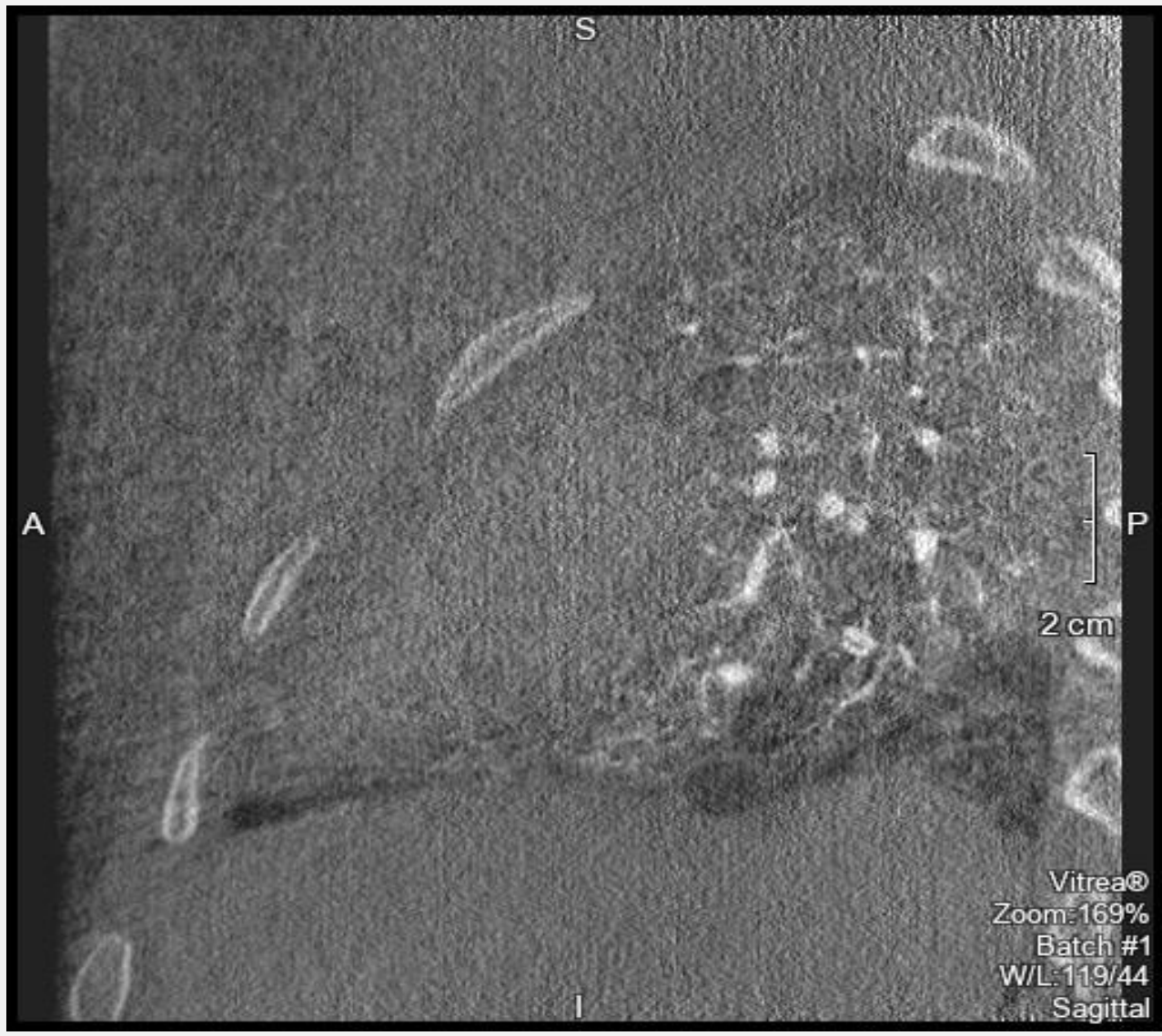


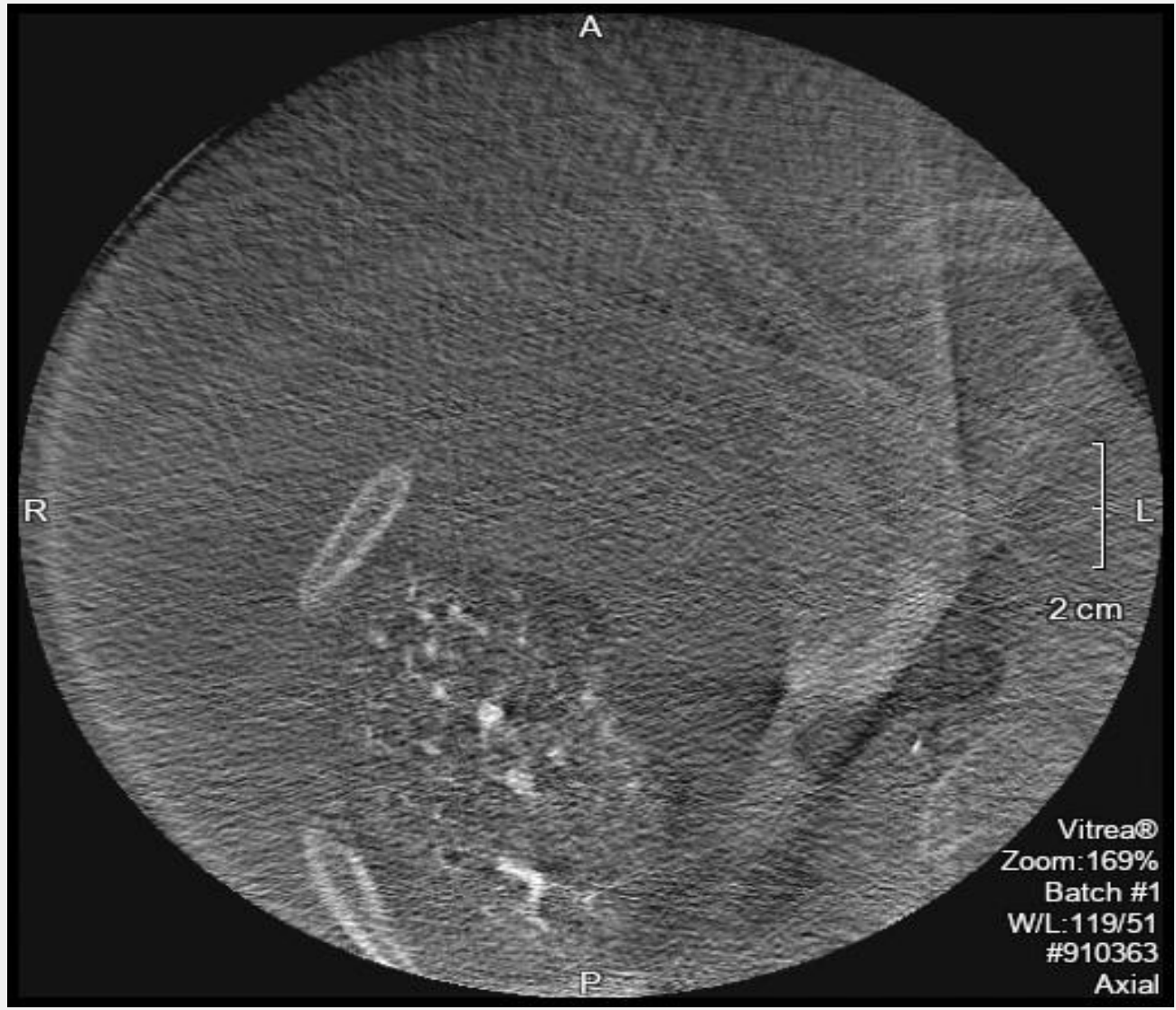
PA Angiogram

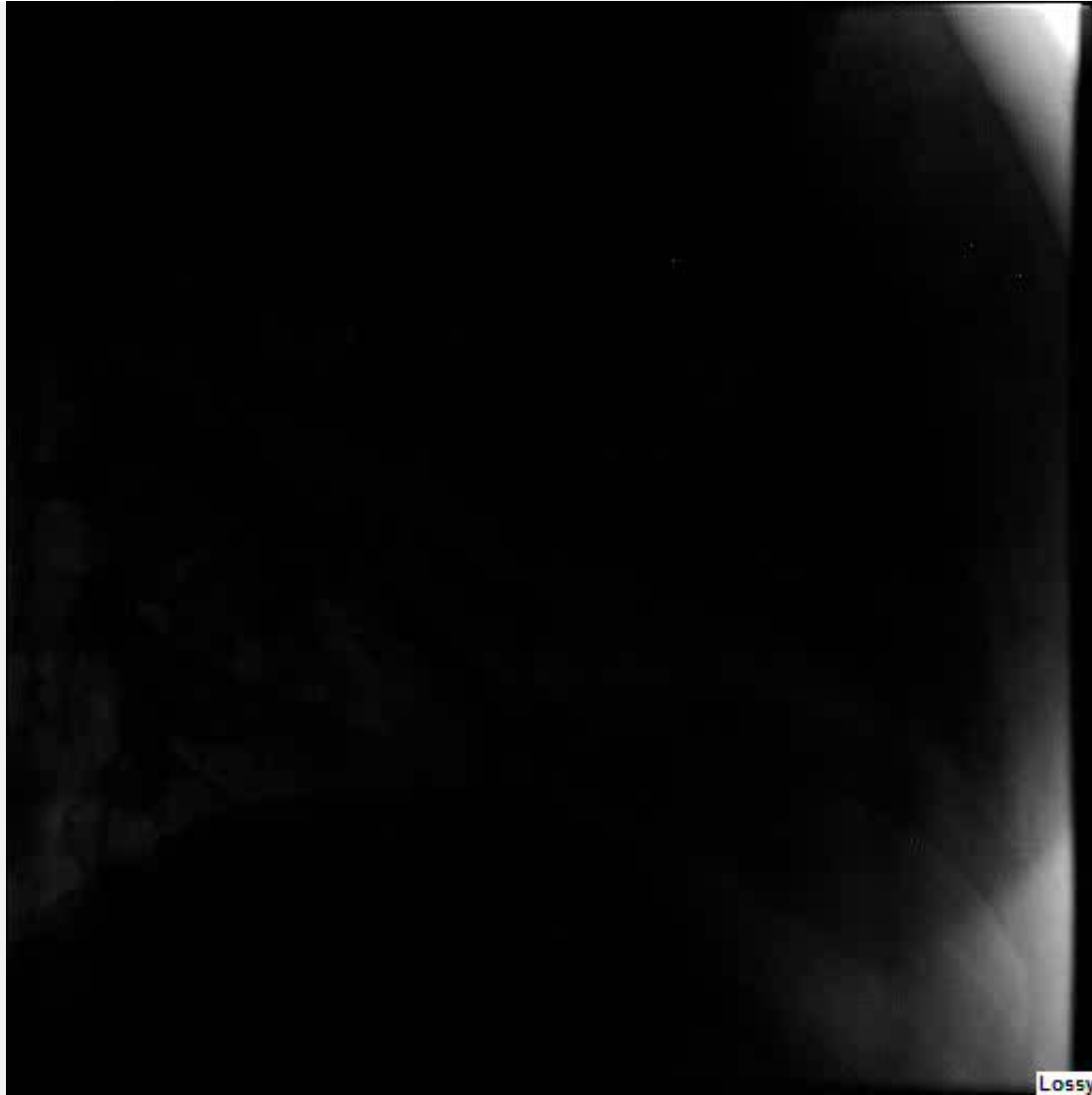




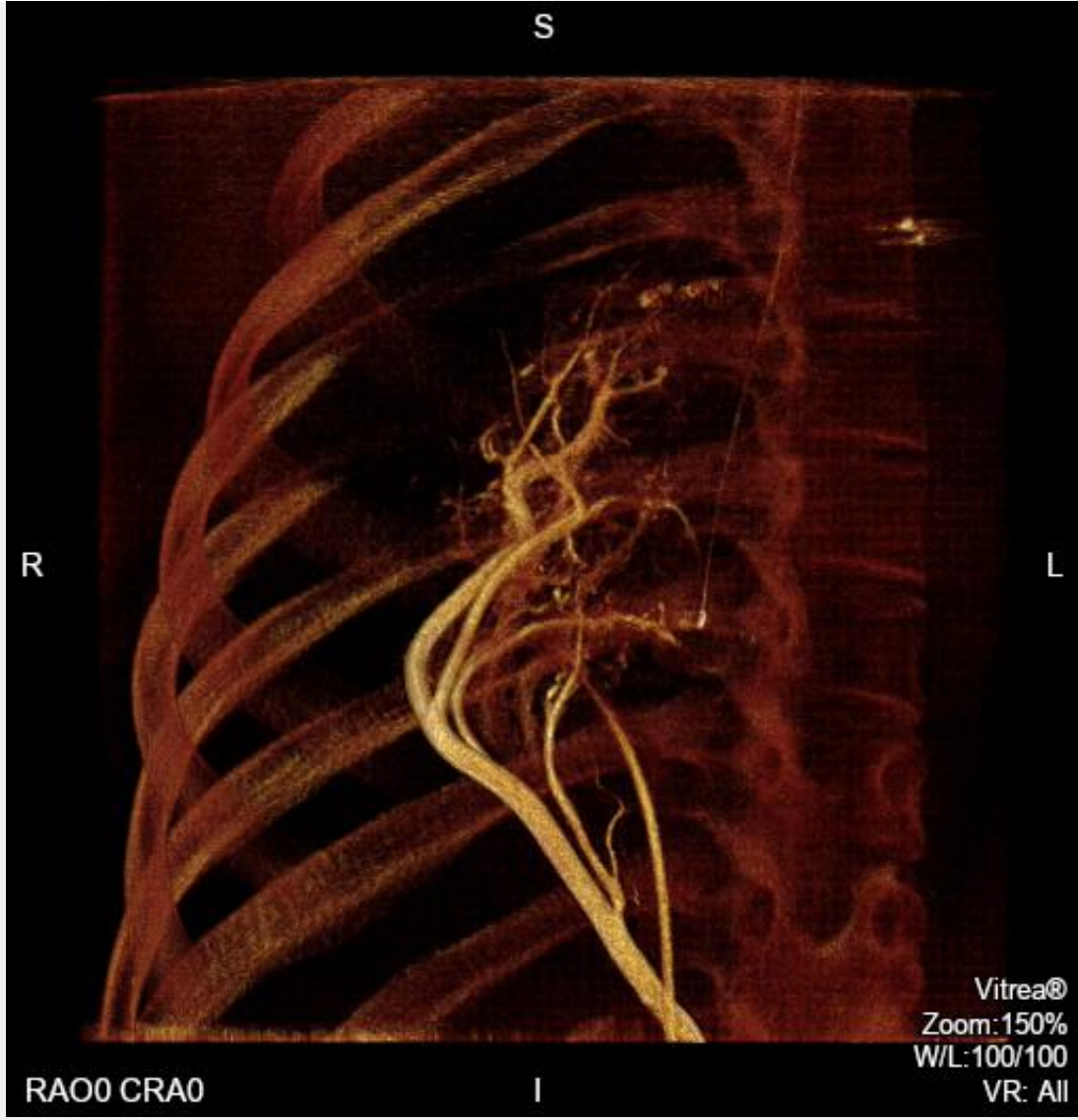








Lossy





RESULTS

- 3DRA from RPA injection demonstrating hypoplasia of the RPA, Scimitar Vein (SV) draining the entire right lung and stenosis of the distal SV.

MPR from same angiogram demonstrating bronchioles normally related to pulmonary vessels.

- 3DRA from APC supplying the sequestered posterior medial lung segment MPR from same angiogram demonstrating sequestered lung with no contributing airway

CONCLUSIONS

- 3DRA was used to accurately diagnose Scimitar Syndrome and define components of pulmonary sequestration.
- 3DRA may obviate the need for additional diagnostic testing such as cardiac CT in patients referred for cardiac catheterization.

