AMSER Case of the Month September 2025

34 y.o. F G3P1 at 28w6d Presents With A Prenatal Fetal Lung Lesion

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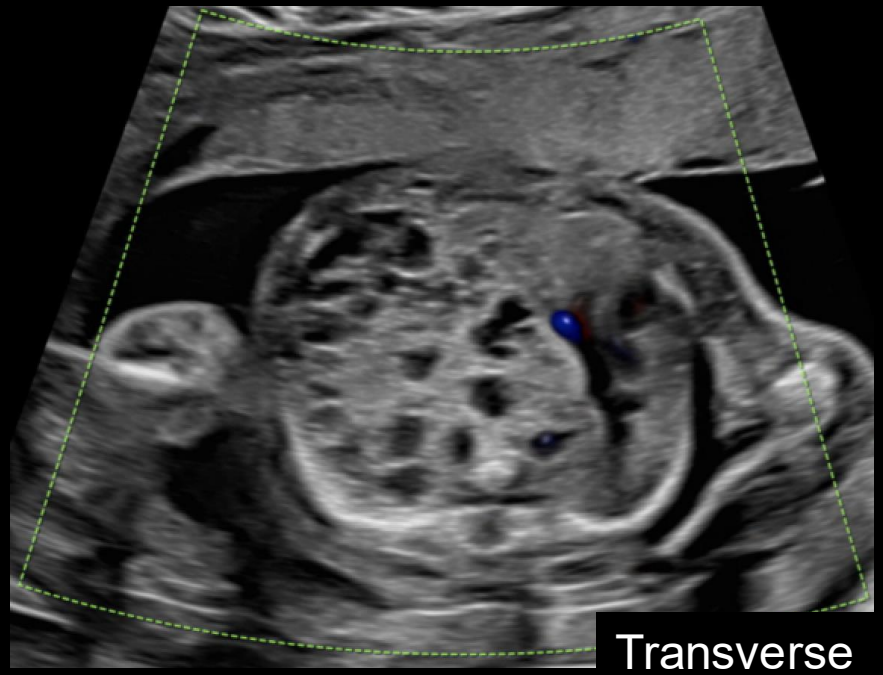
Patient Presentation

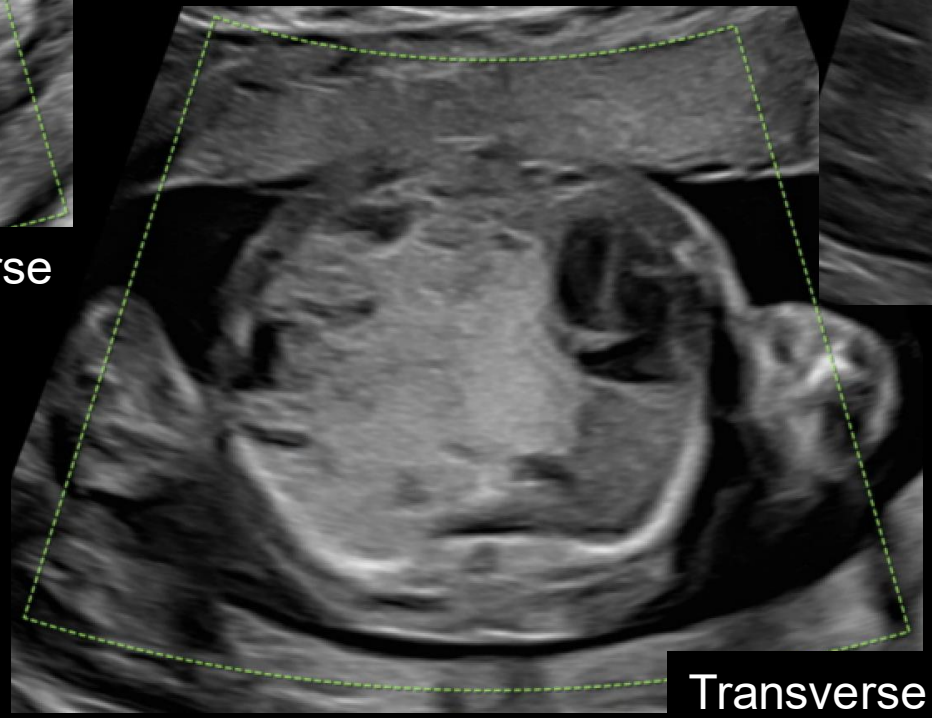
- HPI: 34 y.o. F G3P1 at 20w1d
  - Initially presents with limited 1st trimester ultrasound views of the heart then referred for early anatomy ultrasound

- Past Medical History: Gestational diabetes mellitus

Fetal Anatomy Ultrasound at the level of the heart (unlabeled)



Transverse



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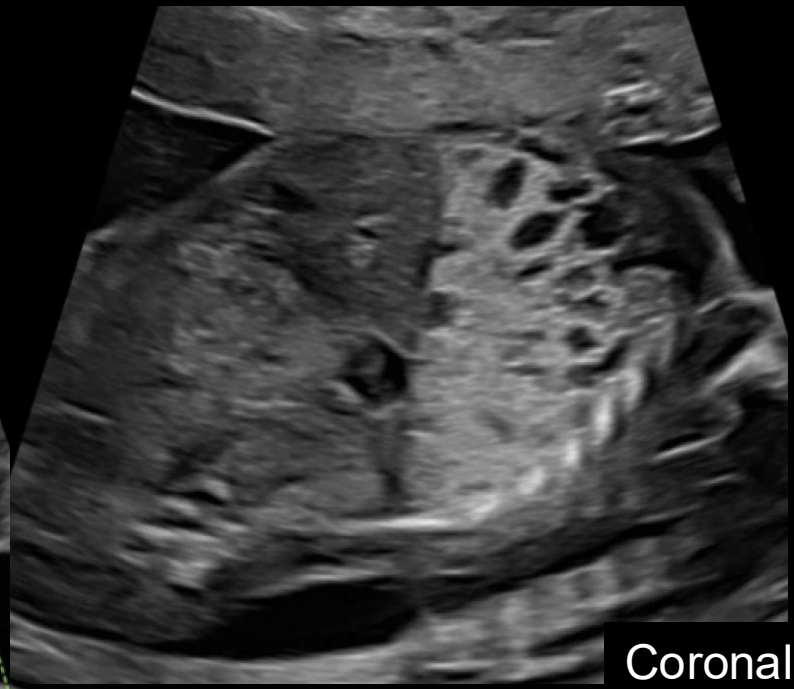

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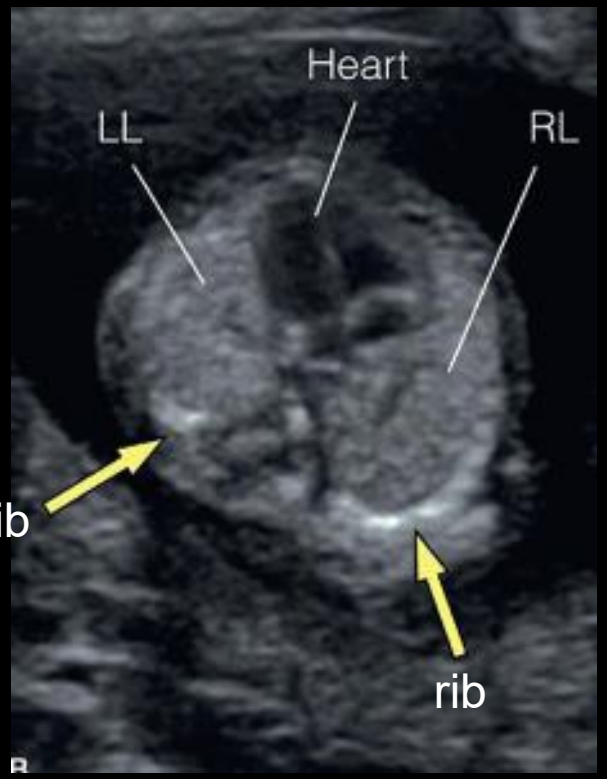
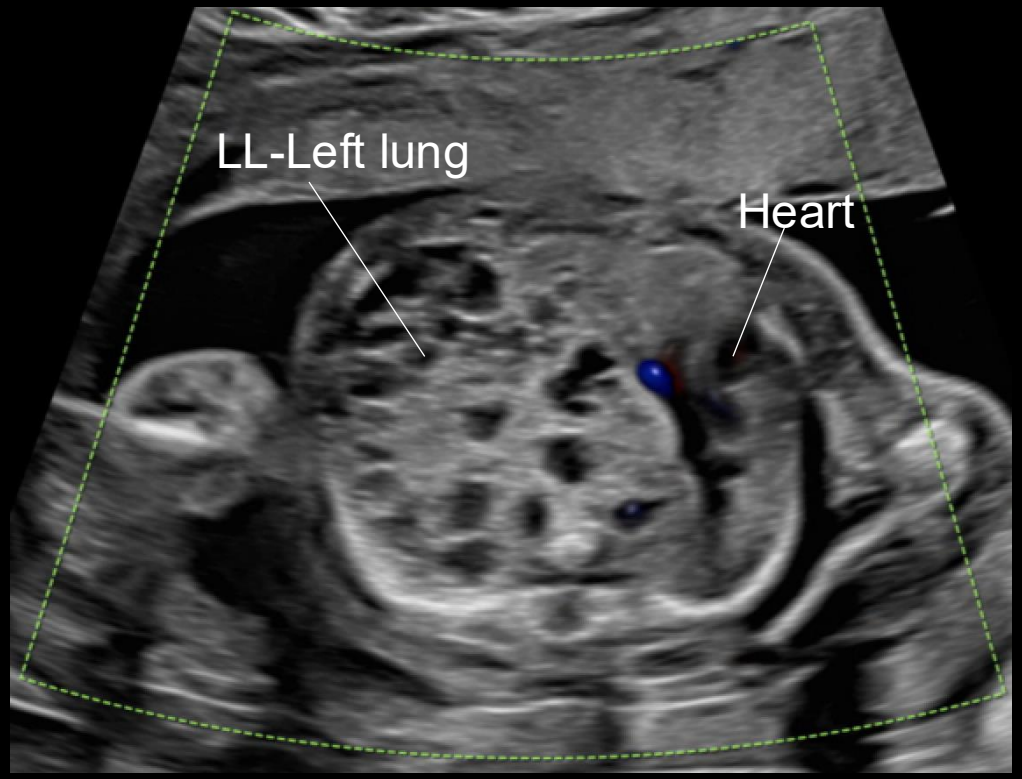
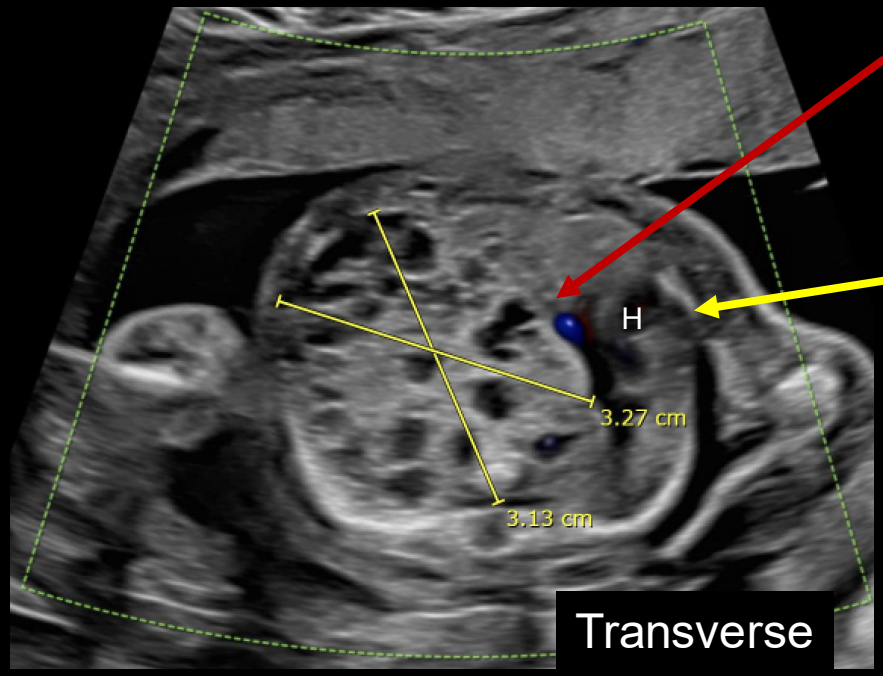
Fetal Anatomy Ultrasound at the level of the heart (Reference normal versus our case)



Reference image courtesy “The fetal Chest/Radiology Key

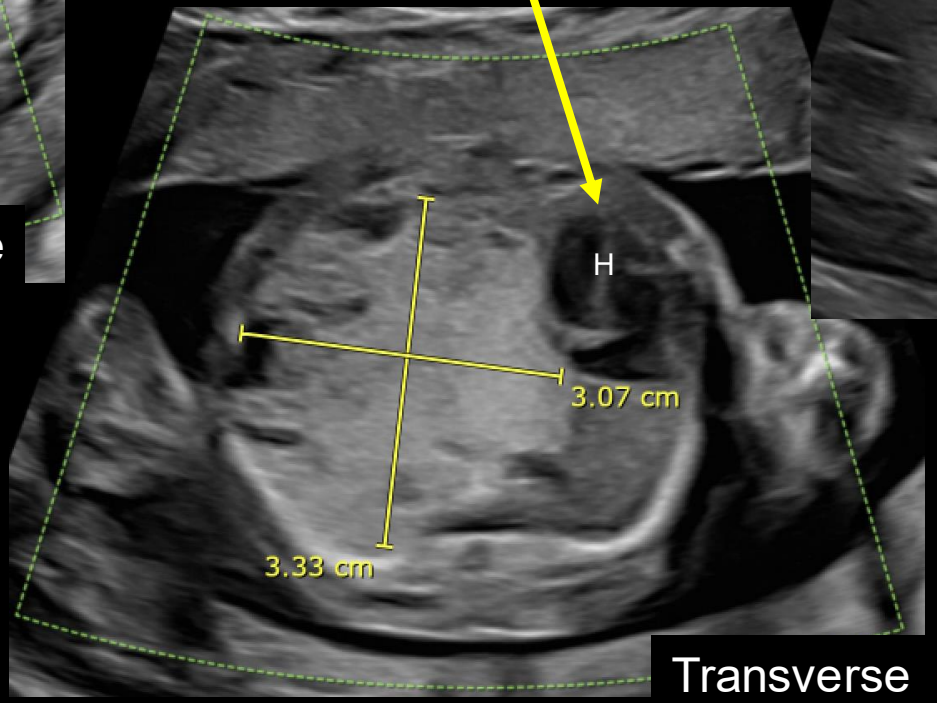


Fetal Anatomy Ultrasound (labeled)



Transverse

Enlargement and increased echogenicity of the left lung with multiple cystic spaces & significant dextroposition of the heart to the right.



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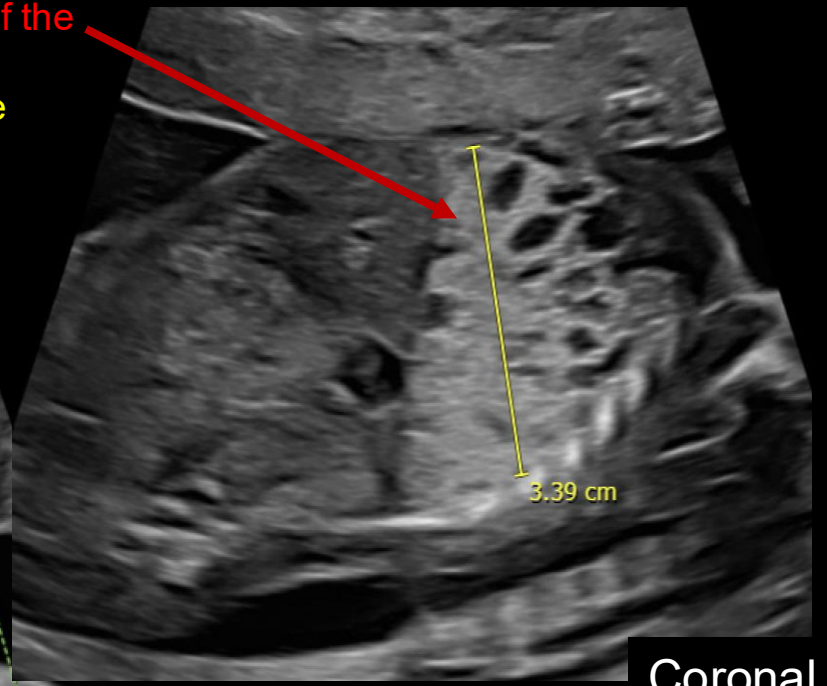

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What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 4: **Second and third trimester screening for abnormal finding on ultrasound: major anomalies. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
US pregnant uterus transabdominal detailed scan	Usually Appropriate	O
MRI fetal without IV contrast	Usually Appropriate	
US echocardiography fetal	Usually Appropriate	
US pregnant uterus transabdominal follow-up	Usually Appropriate	O
MRI fetal without and with IV contrast	Usually Not Appropriate	O

This imaging modality was ordered by the OB physician

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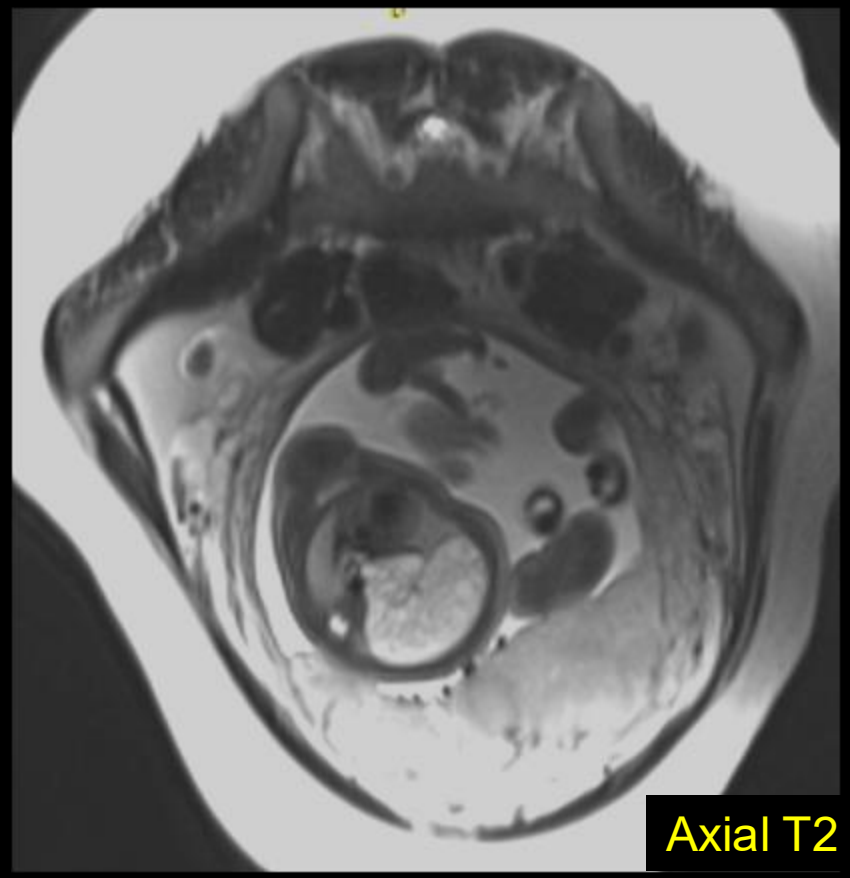
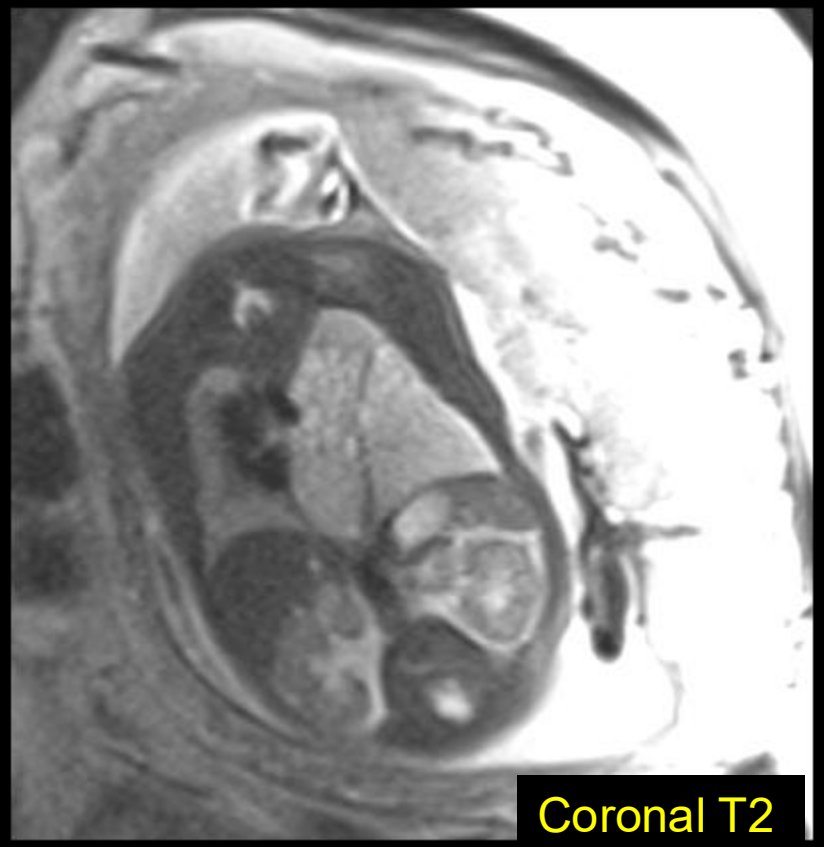
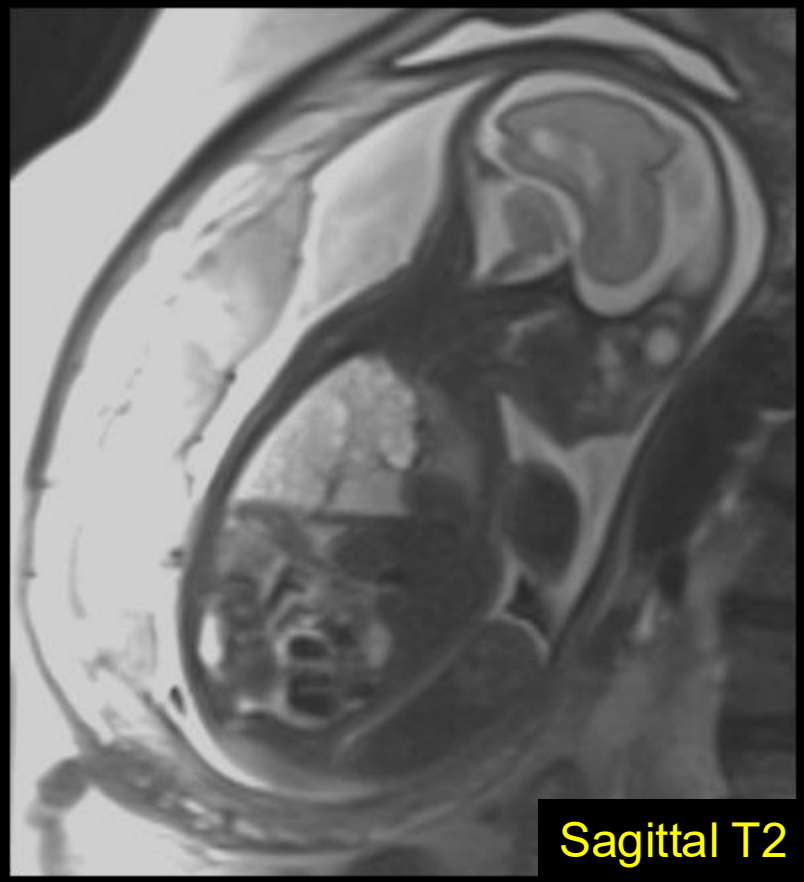
ACR Appropriateness Criteria (continued)

- Ultrasound was inconclusive/limited: Fetal ultrasounds had sub-optimally visualized structures complicated by maternal obesity (BMI >30), reducing image resolution
- Assessment of lesion type & mass effect: : Differentiating between macro- and microcystic CPAM is critical for prognosis & management. MRI would clearly depict potential dextroposition of the heart, aiding in evaluating the degree of thoracic compromise affecting surgical planning.
- Objective lung volume assessment: Fetal MRI enables quantitative lung volume measurement & tracking of Lung-to-Head Ratio (LHR), which would guide delivery planning
- Detection of associated anomalies: Congenital Pulmonary Airway Malformation (CPAM) Type 2 is often associated with other congenital anomalies -MRI allows for comprehensive evaluation of non-pulmonary fetal anatomy.
- Surveillance of fetal hydrops risk: While the CPAM volume ratio (CVR) was 1.02 (below threshold), MRI can rule out hydrops—which if present, would dictate urgent intervention (steroids or fetal surgery).
- Meets ACR criteria for further evaluation of suspected fetal anomaly: ACR guidelines: MRI is appropriate when ultrasound detects or suspects anomalies that require further definition or are not fully visualized.

MRI Fetal w/o Contrast (unlabeled)







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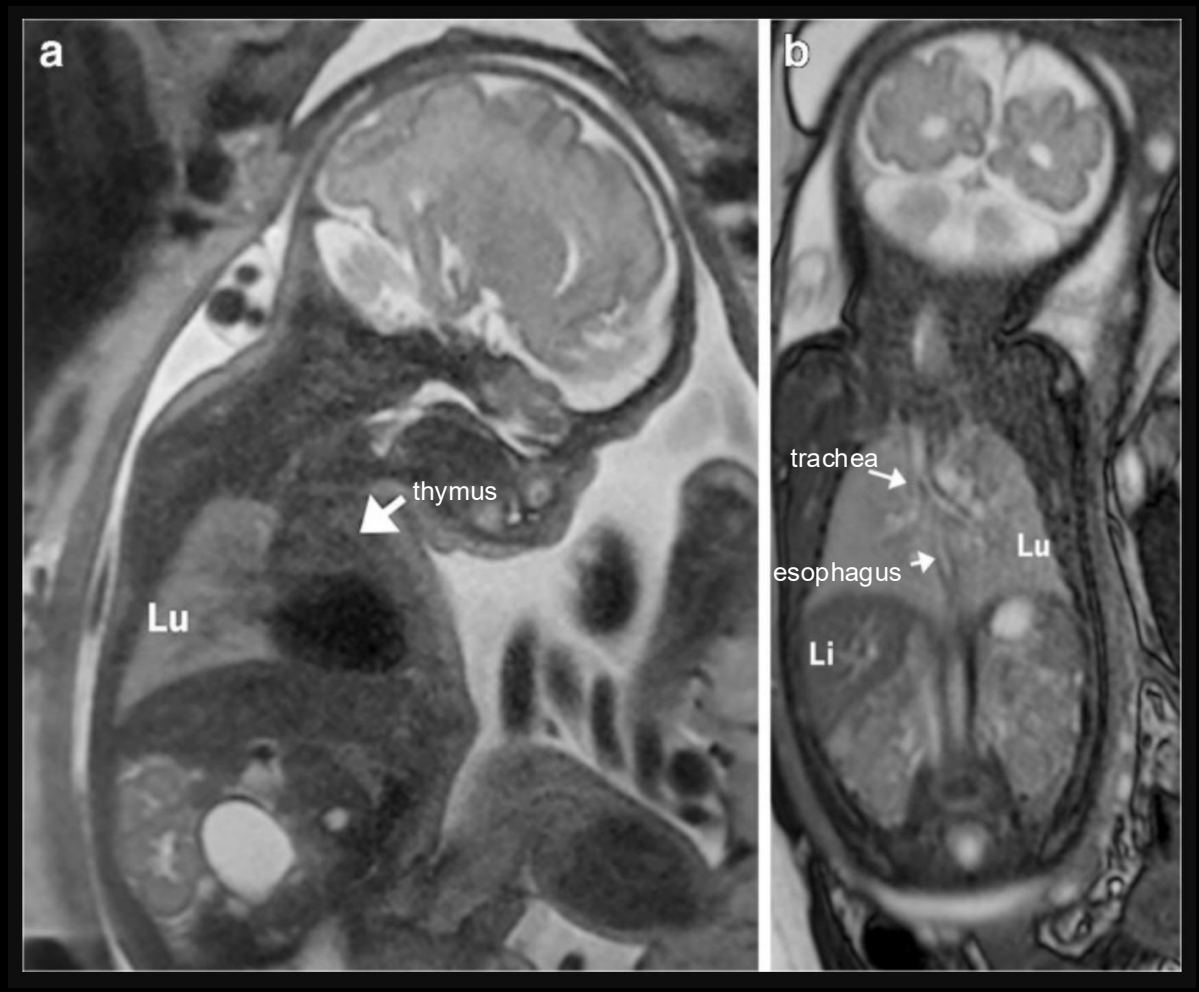
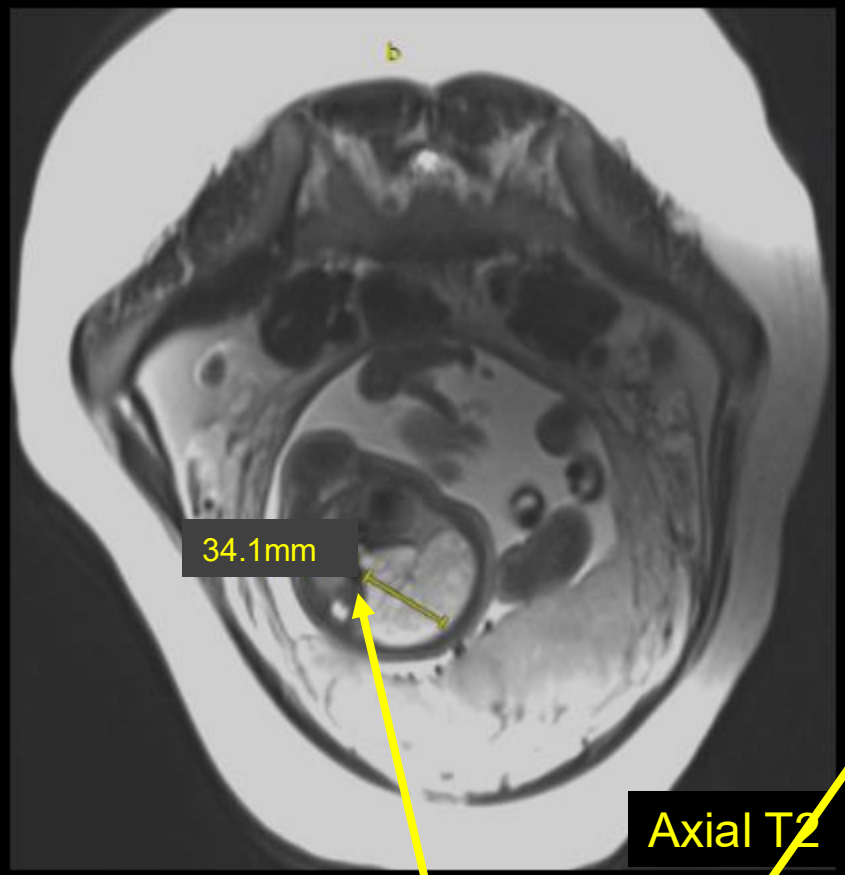
MRI Fetal w/o Contrast (Reference normal)

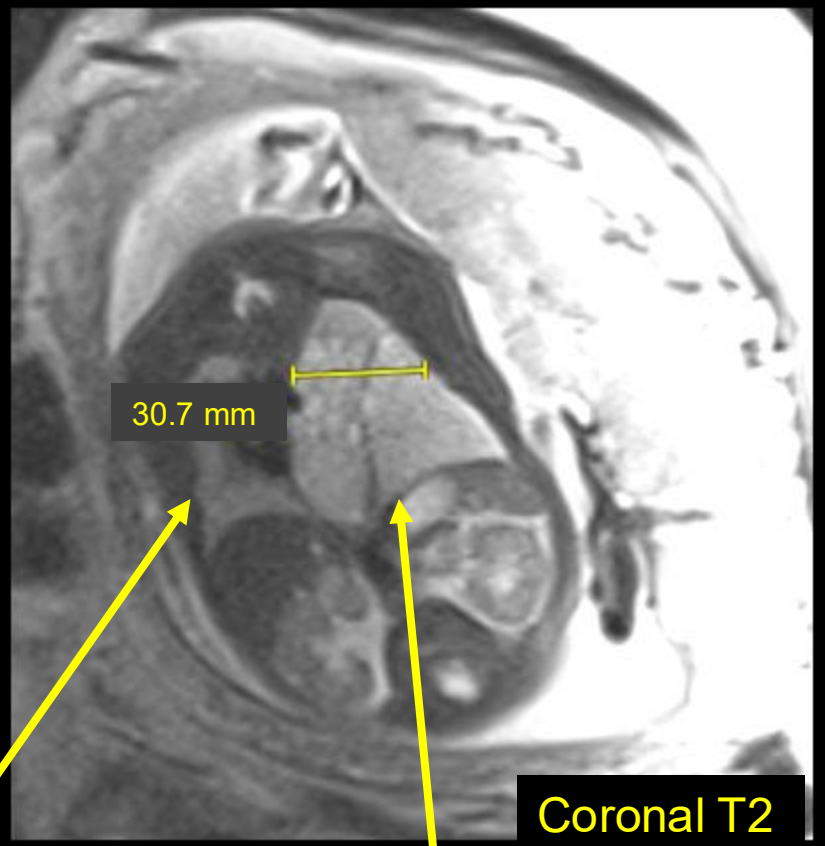


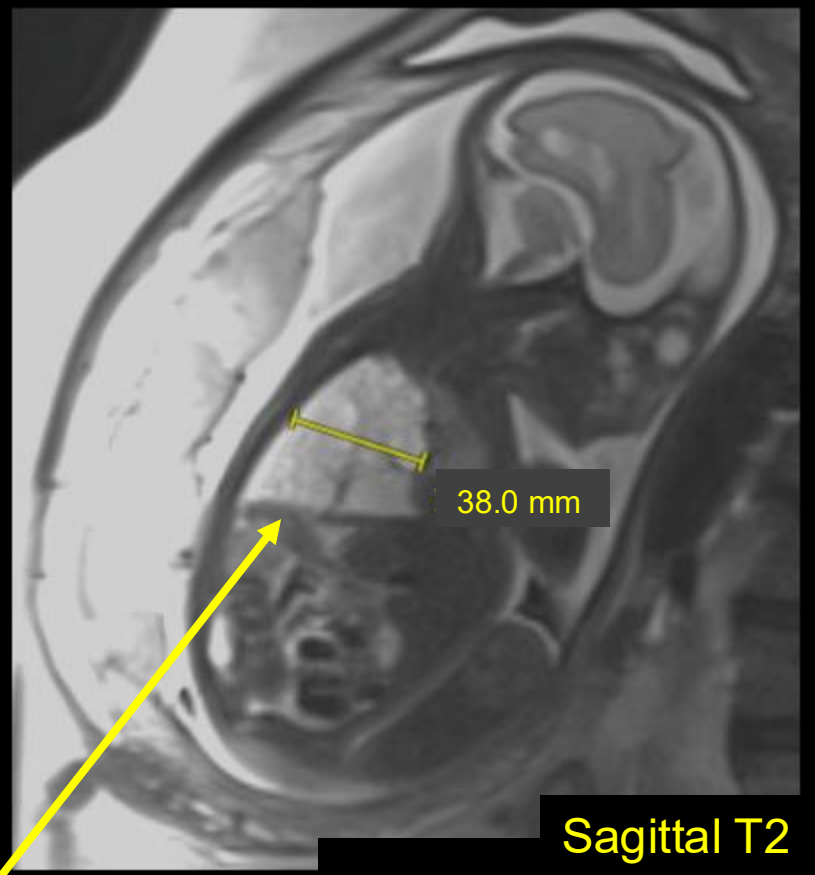
Adams, N.C., Victoria, T., Oliver, E.R. *et al.* Fetal ultrasound and magnetic resonance imaging: a primer on how to interpret prenatal lung lesions. *Pediatr Radiol* **50**, 1839–1854 (2020). https://doi.org/10.1007/s00247-020-04806-x

Lu = lung, Li = liver

MRI Fetal w/o Contrast (labeled)







Rightward mediastinal shift with asymmetrically small right lung.

Enlarged and T2 hyperintense left lower lobe with multifocal cystic areas

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Final Diagnosis:

Congenital Pulmonary Airway Malformation (CPAM) Type 2

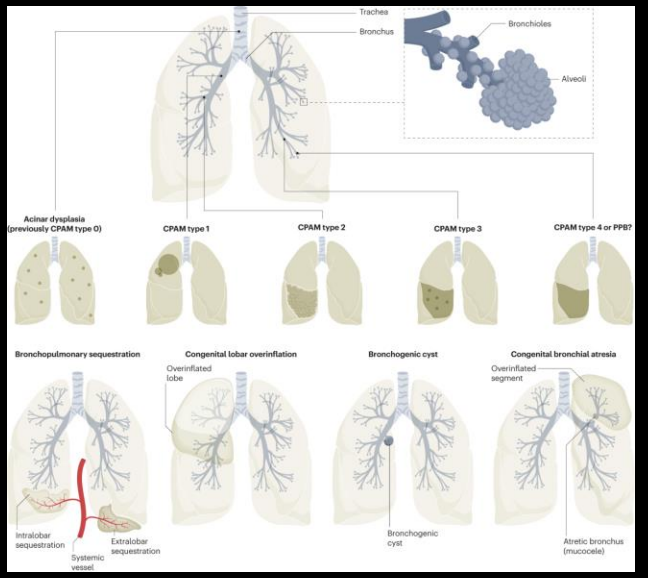
Congenital Pulmonary Airway Malformation (CPAM) – Type 2

- Rare, benign developmental cystic lung lesion
  - Occurs in ~1 per 25,000–35,000 live births
  - Classification: Types 0–4 (Stocker classification) based on cyst appearance & region in lung
  - Type 2 lesions (multiple small cysts of bronchiolar regions) often associated with renal, cardiac, or GI anomalies
- Presents with respiratory distress in neonates
  - Commonly detected prenatally on routine ultrasound
  - Many are asymptomatic at birth
  - Can present later with recurrent infections & resemble tumor or hyperinflation
- Work-up: Prenatal US → Postnatal CT/MRI → Surgical Resection
  - If CVR > 1.6, risk of hydrops → consider fetal intervention or steroids

Imaging Characteristics & Differential

- Imaging characteristics vary with type & age
  - Type 2: Multiple uniform small cysts, <2 cm
  - CT: Cystic, sharply demarcated, non-enhancing unless infected
  - MRI (fetal): High T2 signal, hyperintense, fluid-filled cysts
  - May be misdiagnosed as infection or mass on radiographs
- Cysts may enlarge with time; superinfection may distort appearance
  - Rarely show air-fluid levels or hemorrhagic debris
  - No systemic feeding vessel or solid component in Type 2 lesions
- Differential for suspected CPAM:
  - CPAM Types 0-4

CPAM	Characteristics
Type 0	Tracheal/bronchial Cysts < 0.5 cm
Type 1	Bronchial/bronchiolar Large cysts 2-10 cm
Type 2	Bronchiolar Solid tissue cysts <2 cm
Type 3	Alveolar ducts Microcysts <0.5 cm
Type 4	Acinar Large cysts >10 cm



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Treatment & Genetic Considerations

- Main treatment is resection of the involved lung segment/lobe
  - Elective surgery usually before 1 year of age
  - Resection even in asymptomatic cases to prevent infection or malignancy
- Preoperative planning:
  - CT or MRI to define anatomy, cysts, and exclude feeding vessels
  - Echocardiogram to evaluate for associated cardiac anomalies
  - Respiratory stabilization if symptomatic
- Genetic testing not standard, but:
  - Consider DICER1 mutation screening in solid or atypical lesions (pleuropulmonary blastoma risk)
  - No known inherited syndromes with classic CPAM
- Post-op management and surveillance:
  - Chest imaging to confirm complete resection
  - Monitor for infections, compensatory hyperinflation
  - Long-term prognosis is excellent; lung function usually normal

Patient Outcome

- As of 30w3d:
  - Assessment
    - The CPAM volume ratio (CVR) is 1.7, accompanied by notable mediastinal shift but w/o evidence of cardiac compression or fetal hydrops/polyhydramnios.
    - Given presence of macrocystic lesions & considering maternal GDM, steroid administration is deferred
      - Will reconsider if lesion enlarges, hydrops develops, or cardiac function becomes compromised
  - Plan
    - 2x-weekly biophysical profile (BPP) evaluations
    - Maintain optimal glycemic control
    - Closely monitor maternal BP & observe for signs/symptoms of preeclampsia.
    - Consultations with NICU & pediatric surgery
    - Planned delivery at 37–38 weeks at Special Delivery Unit (SDU)

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