Fetal Lung Mass Controversies

Richard A. Barth, M.D.
Lucile Packard Children's
Hospital at Stanford University

Overview

- Prenatal Diagnosis of Chest Masses
  Historical and Current State
- Predicting Outcome of Chest Masses
- MRI of Chest Mass
  Is there value-added?
- Can Prenatal MRI reduce need for Postnatal Imaging?

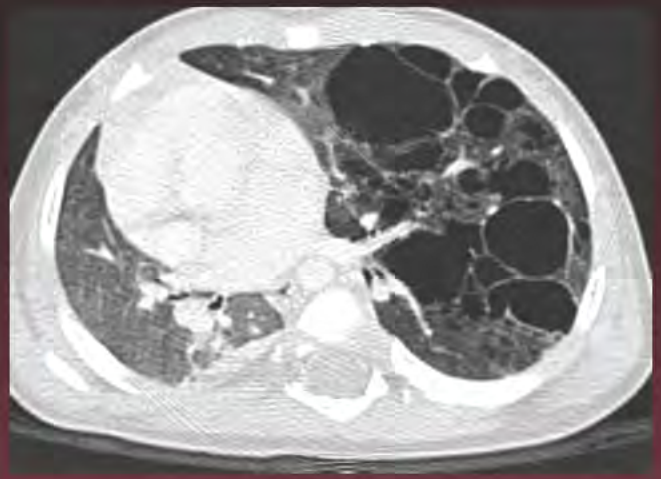
Distribution of Pathologically Proven Fetal Lung Lesions
CHOP Experience (108 Cases)*

CPAM	47%
Hybrid (Sequestration and CPAM)	25%
Overinflation/Bronchial Atresia	20%
Sequestration	8%

* Epelman M, et al. Seminars in Ultr, CT and MRI, 2010; 31: 141-157

Historical Background- Congenital Lung Masses

- Pre-Sonographic Era
  - Majority of lung masses presented with respiratory distress or pneumonia in infancy

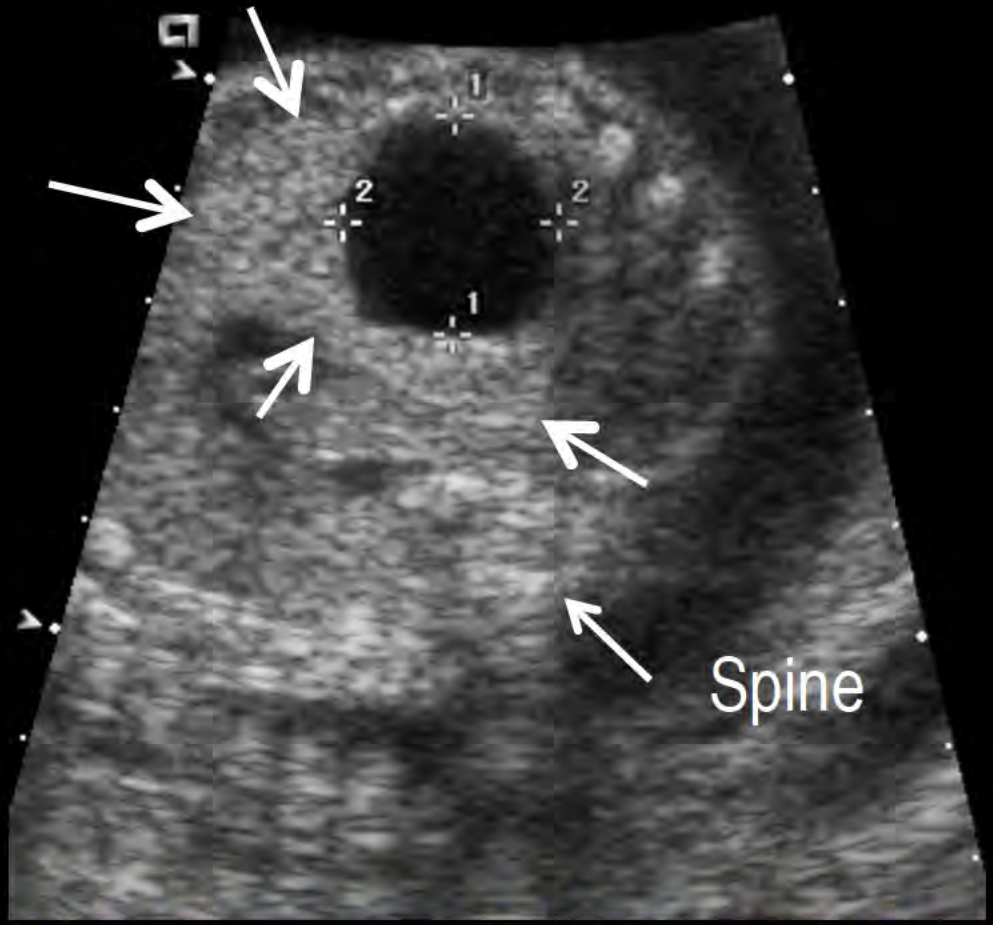


Infant with Respiratory Distress 2° to CPAM

Historical Background- Congenital Lung Masses

- Sonographic Era
  - Majority of masses detected as incidental finding on pre-natal Ultrasound or MRI
  - Prenatal Imaging plays a key role in predicting outcome, family counseling, and management decisions

Incidental Congenital Cystic Lung Mass- 22wks GA

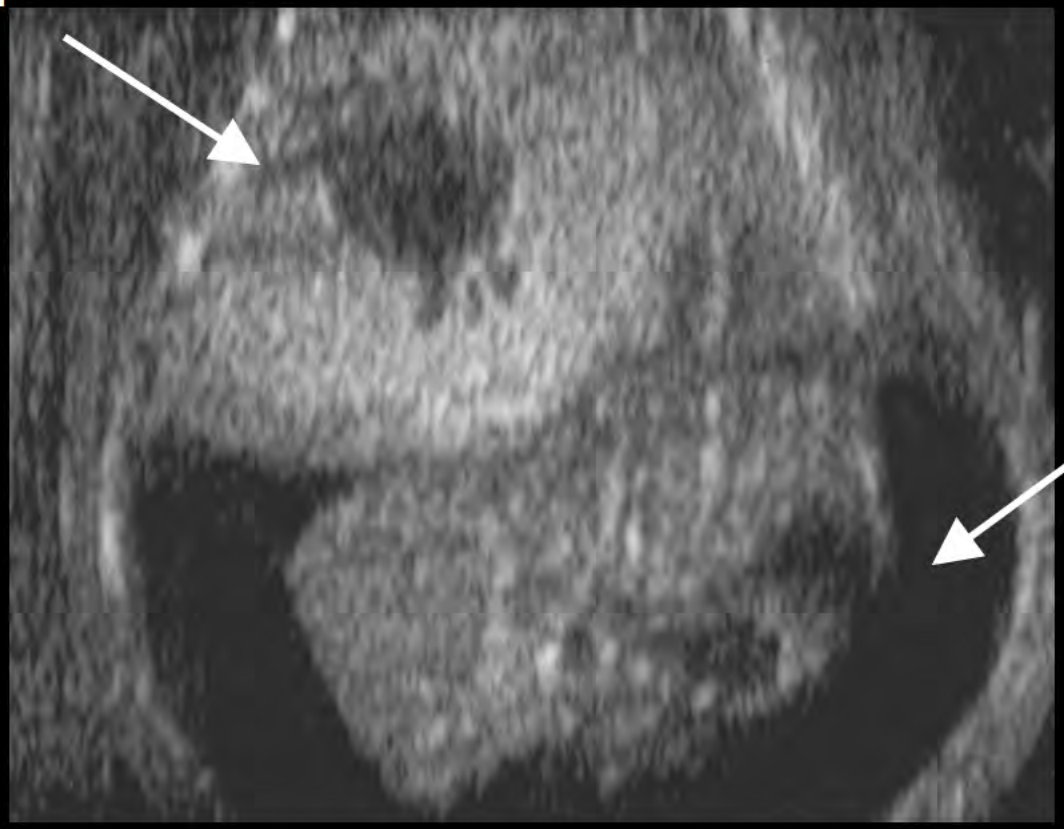


Congenital Pulmonary Airway Malformation is Most Common Cause of a Cystic Fetal Lung Mass

Dx of Isoechoic Solid Masses

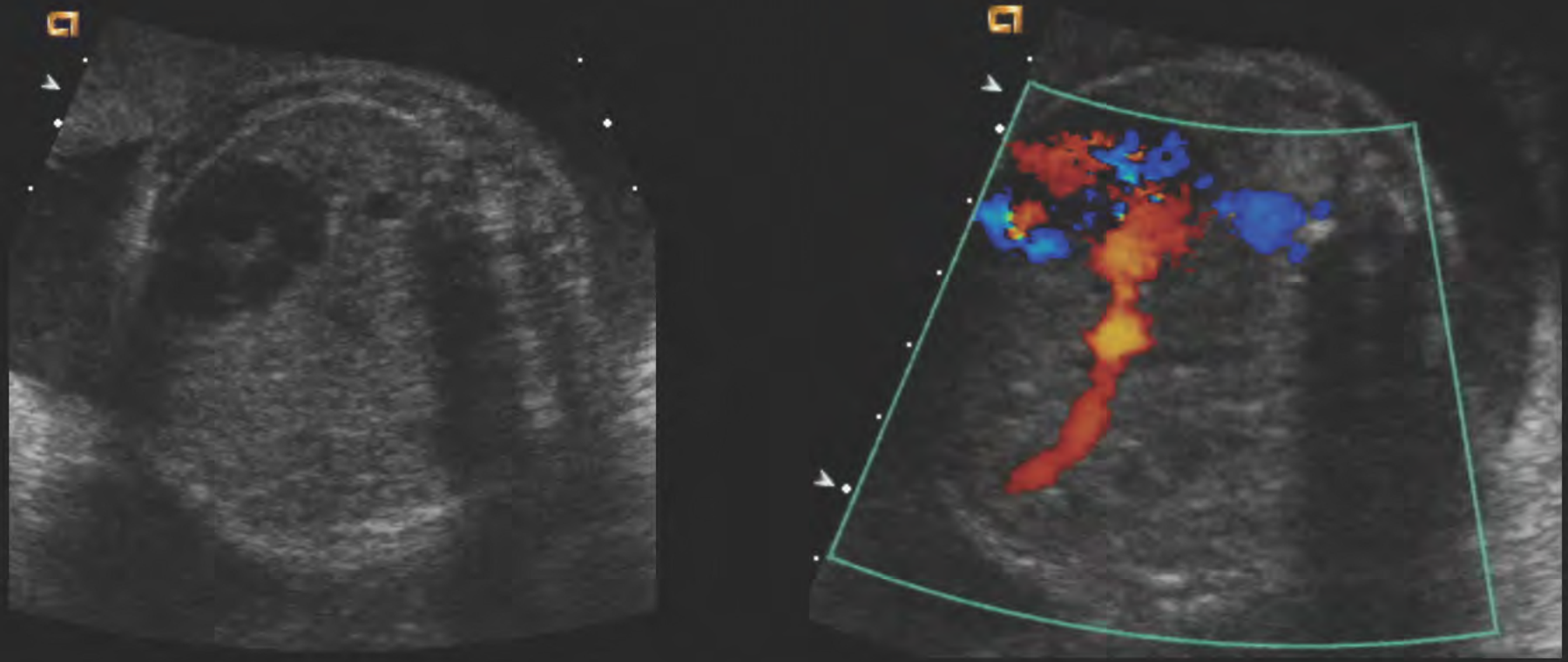
- Mass Effect
  - Mediastinal Shift
  - Altered Cardiac Position/Axis
  - Flat/Inverted Diaphragm

CPAM with Hydrops



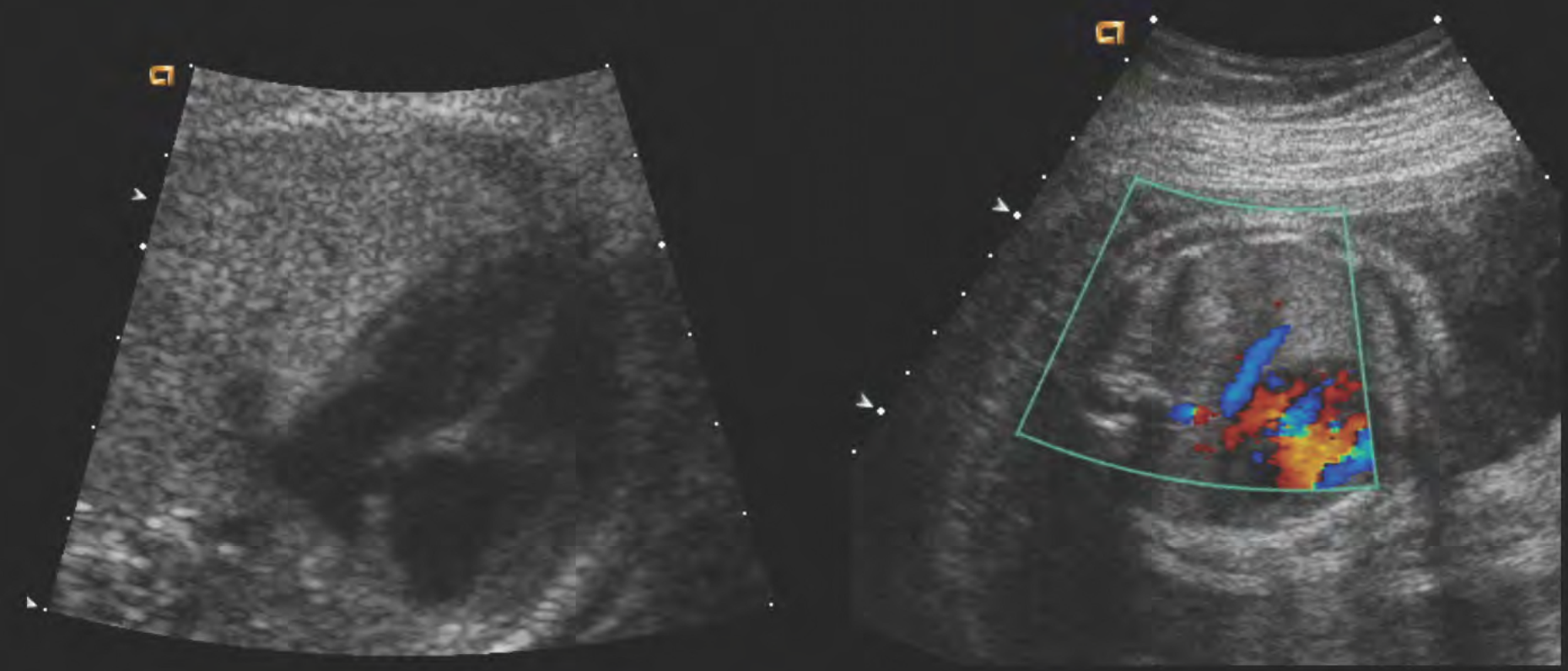
Mediastinal Shift 2° to Rt Lung Mass

Doppler US is Important for Characterizing Solid Masses



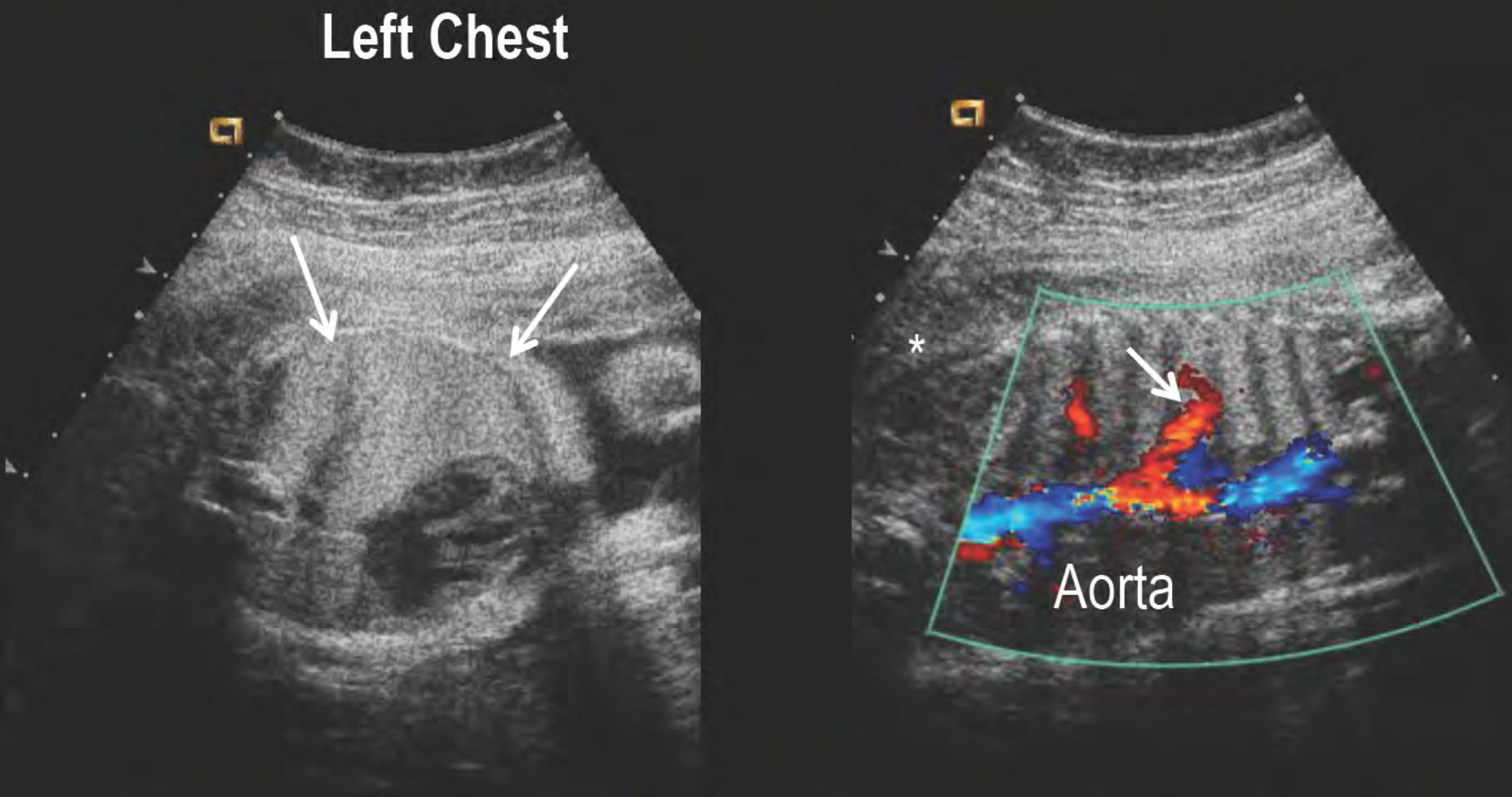
Right CDH with Liver Up

Doppler US is Important for Characterizing Solid Masses



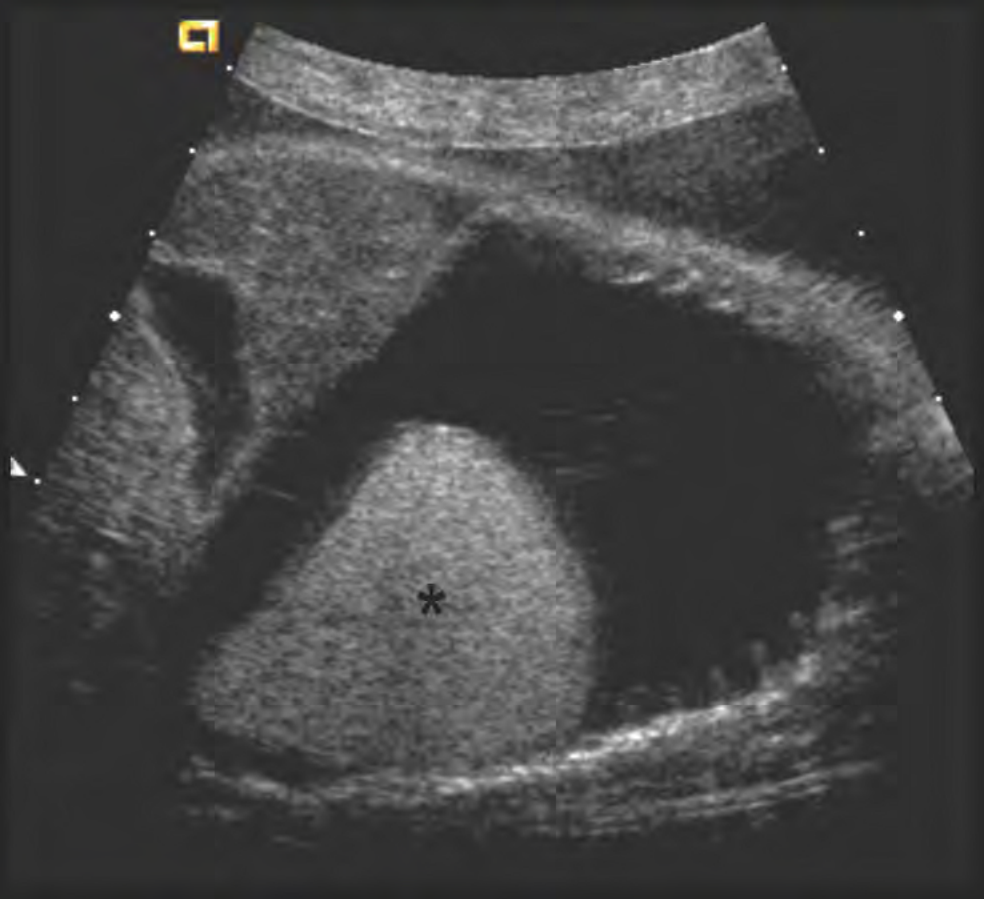
CPAM arterial supply via pulmonary artery

Doppler US is Important for Characterizing Solid Masses



Systemic arterial feeder confirms Sequestration

Torsion of Extralobar Sequestration



Large pleural effusion favors sequestration

Newborn-Torsed Sequestration

Predicting Outcome for Fetal Lung Masses (CPAM)

- Small masses → No Hydrops → excellent prognosis
- Large masses → Hydrops fetalis (10% of cases, usually fatal), pulmonary hypoplasia
  - 40% of Masses grow in-utero
  - 15% of Masses stable or regress in-utero

Whereas hydrops is almost universally associated with a large lung mass, only a minority of large masses result in hydrops

Predicting Outcome for Fetal Lung Masses (CPAM)

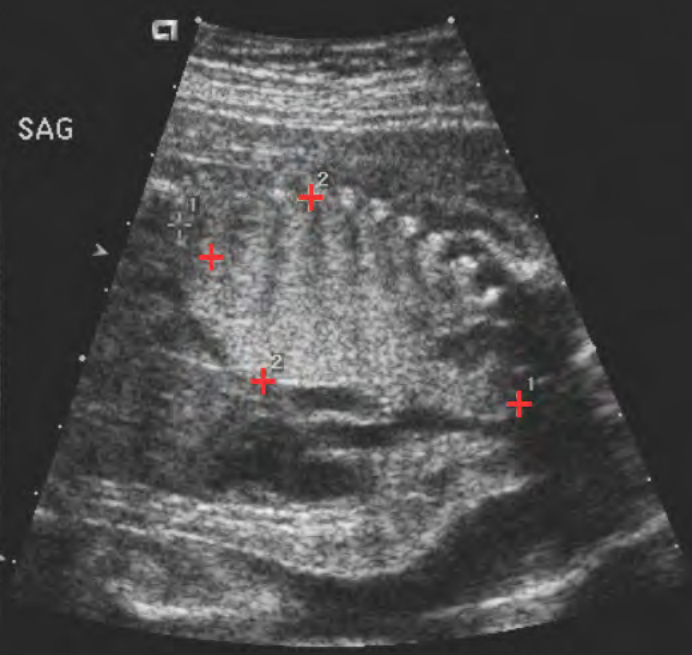
- Small masses → No Hydrops → excellent prognosis
- Large masses → Hydrops fetalis (10% of cases, usually fatal), pulmonary hypoplasia
  - 40% of Masses grow in-utero
  - 15% of Masses stable or regress in-utero

Whereas hydrops is almost universally associated with a large lung mass, only a minority of large masses result in hydrops

GROWING CPAM

18 wks

23 wks



23 wks

Congenital Pulmonary Airway Malformation

21 wks GA

Congenital Pulmonary Airway Malformation

21 wks GA

Newborn

Asymptomatic

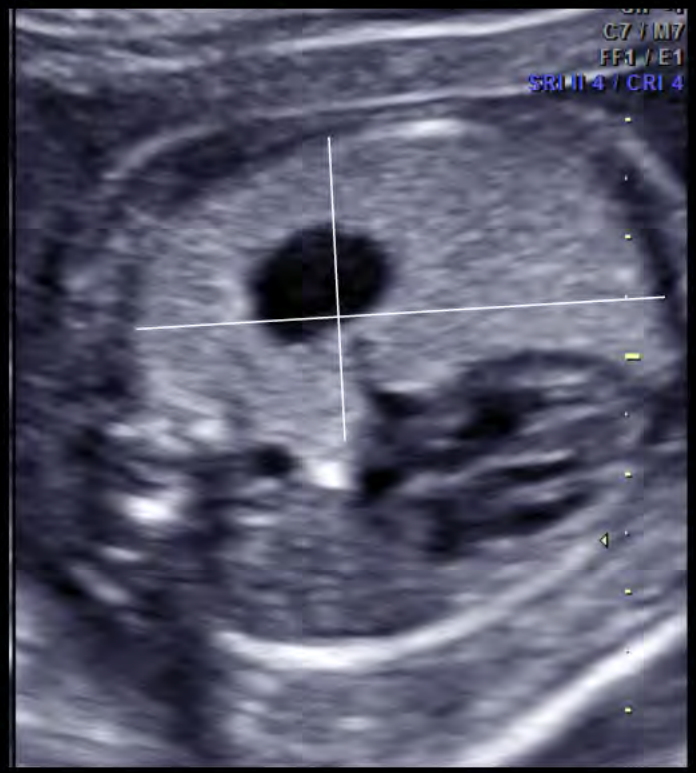
CPAM Volume Ratio (CVR) predicts Hydrops Risk 2° to CPAM*

- Volume Ratio (CVR) = CPAM Volume/Head Circumference
- CVR ≥ 1.6 → Increased Risk for Hydrops (75%)
- CVR ≤ 1.6 & No Dominant Cyst → < 3% Risk for Hydrops

*Crombleholme TM, et al J Ped Surg, 2002; 37: 331-338

CPAM Volume Ratio (CVR)
(CPAM Volume divided by Head Circumference)
CVR = L x H x W x 0.52 / HC

Sagittal



Transverse

CPAM Volume Ratio (CVR) predicts Hydrops Risk *

- Volume Ratio (CVR) = CPAM Volume/Head Circumference
- CVR $\geq$ 1.6 → Increased Risk for Hydrops (75%)
- CVR $\leq$ 1.6 & No Dominant Cyst → < 3% Risk for Hydrops

*Crombleholme TM, et al J Ped Surg, 2002; 37: 331-338

Congenital Pulmonary Airway Malformation

24 wks GA

CVR = 2.8

Congenital Pulmonary Airway Malformation

24 wks GA

CVR = 2.8

Prenatal Steroids for Microcystic CPAM (Data from 3 Centers)*

	Pts	CVR	Hydrops	Hydrops Resolved	Survival
UCSF	13	2.7	9 (69%)	7 (78%)	11 (85%)
CHOP	10	2.2	5 (50%)	4 (80%)	10 (100%)
Cincinnati	8	2.5	6 (75%)	5 (83%)	6 (75%)
Total	**31**	**2.5**	**20/31 (65%)**	**16/20 (80%)**	**27/31 (87%)**

*Adapted from Curran PF, et al; (2010), J Pediatr Surg, 45:145-150

Steroid Rx vs Fetal Surgery in 24 fetuses with CPAM and Fetal Hydrops*

	Steroid Rx	Surgical Rx
Mean GA Age	23 wks	24 wks
CVR	2.68 ± .29	2.95 ± 0.31
Survival to Delivery	12/13 (92%)	9/11 (82%)
Survival to Discharge	10/12 (83%)	5/9 (56%)

*Loh KC, et al. J Pediatr Surg. 2012; 47:36:39

Predicting Outcome for Fetal Lung Masses (CPAM)

- Small masses → No Hydrops → excellent prognosis
- Large masses → Hydrops fetalis (10% of cases, usually fatal), pulmonary hypoplasia
  - 40% of Masses grow in-utero
  - 15% of Masses stable or regress in-utero

Whereas hydrops is almost universally associated with a large lung mass, only a minority of large masses result in hydrops

Does MRI add value in Dx of Lung Masses ?

- Confirm Alternative/Additional Dx

  In 38% to 50% of fetuses MRI provided additional information c/w US*
- Helpful for Equivocal US

*Levine D, et al Radiology, 2003; 228: 379-388

*Hubbard AM Radiology, 1999; 212: 43-48

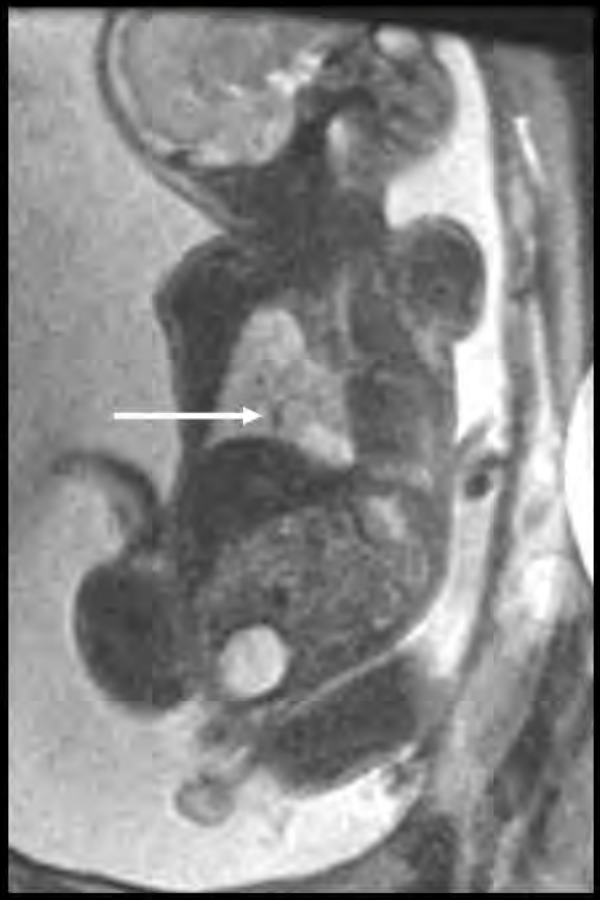
MRI of Congenital Lung Masses

26 weeks GA - Fluid sensitive sequence

Most masses exhibit higher signal than normal lung on fluid sensitive sequences (2nd Trimester)

MRI of Congenital Lung Masses

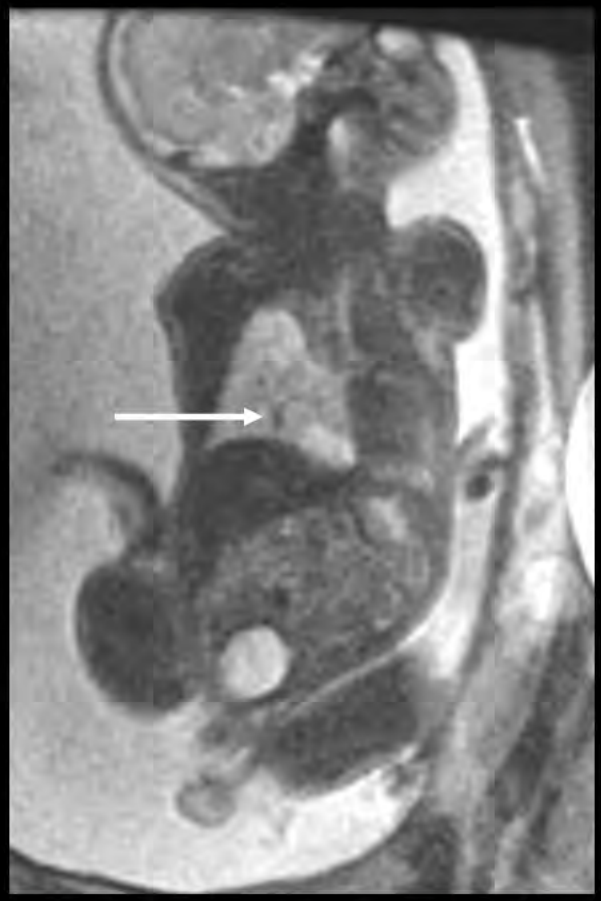
26 weeks GA - Fluid sensitive sequence



Most masses exhibit higher signal than normal lung on fluid sensitive sequences (2nd Trimester)

MRI of Congenital Lung Masses

26 weeks GA - Fluid sensitive sequence



Most masses exhibit higher signal than normal lung on fluid sensitive sequences (2nd Trimester)

MRI of Congenital Lung Masses

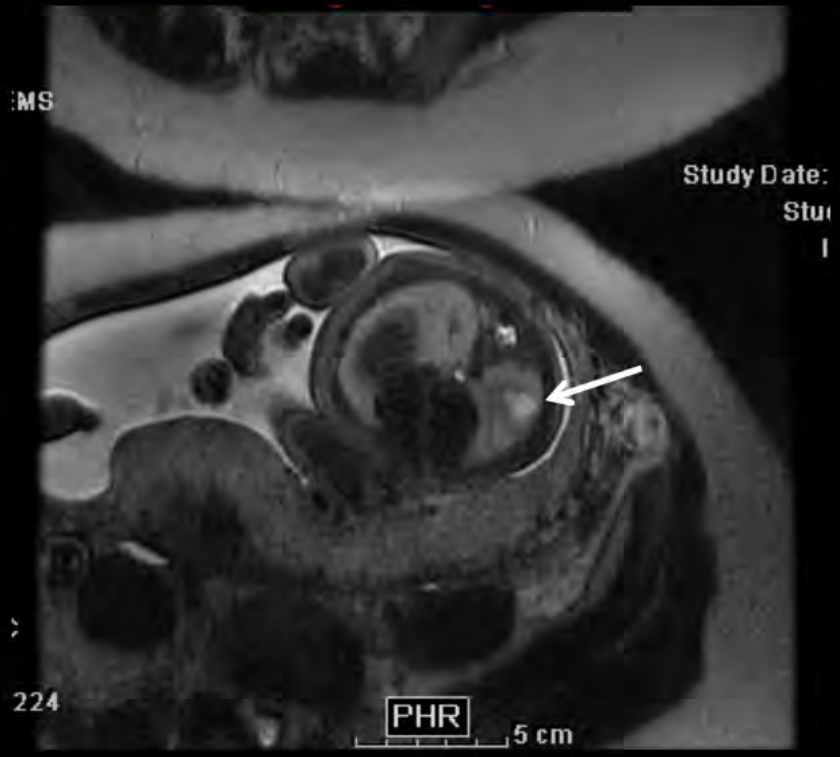
26 weeks GA - Fluid sensitive sequence

Most masses exhibit higher signal than normal lung on fluid sensitive sequences (2nd Trimester)

Referral for Fetal Lung Mass
3rd Trimester - BPM's More Difficult to see on US

30 weeks Gestational Age

Referral for Fetal Lung Mass
3rd Trimester - BPM's More Difficult to see on US



FOV 26 cm, ST= 3mm

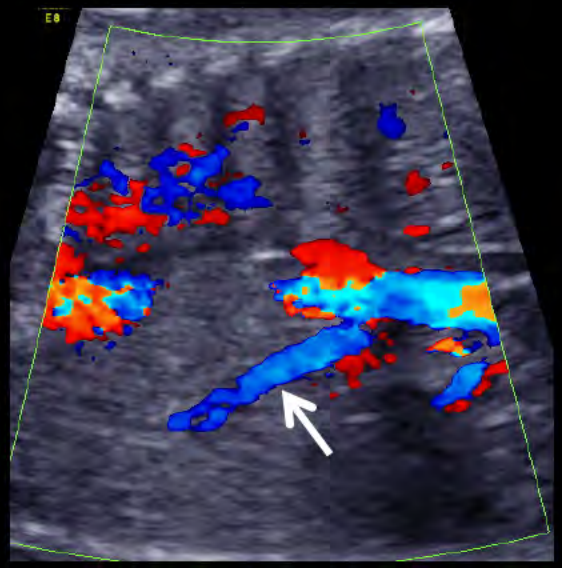
FOV 35 cm, ST= 4mm

Congenital Pulmonary Airway Malformation

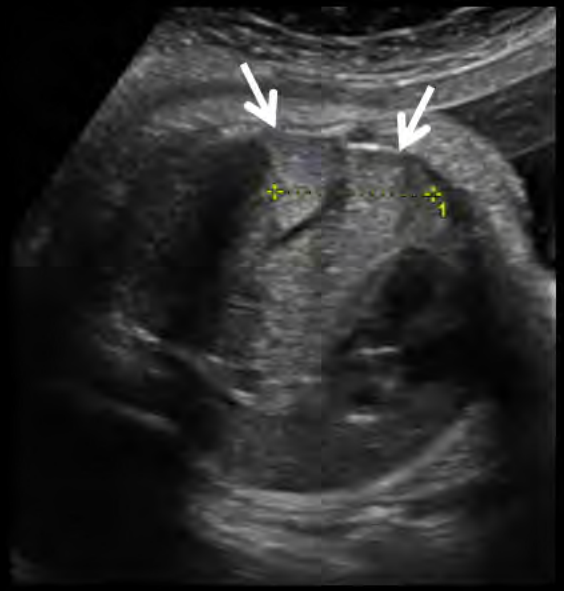
32 wks GA

2nd Lesion- BPS in Lower Lobe

Bronchopulmonary Sequestration-35 wks GA



Bronchopulmonary Sequestration-35 wks GA



Ultrasound

Incidental Lt CDH

T1 wt MRI

Hybrid Airway Malformation

24 wks GA

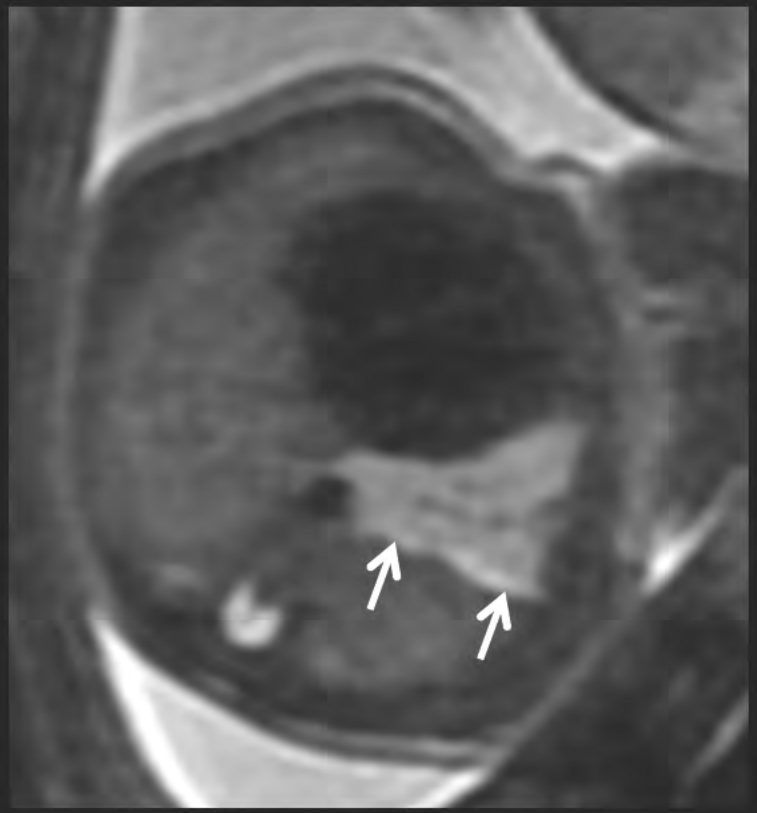
Heart

Aorta

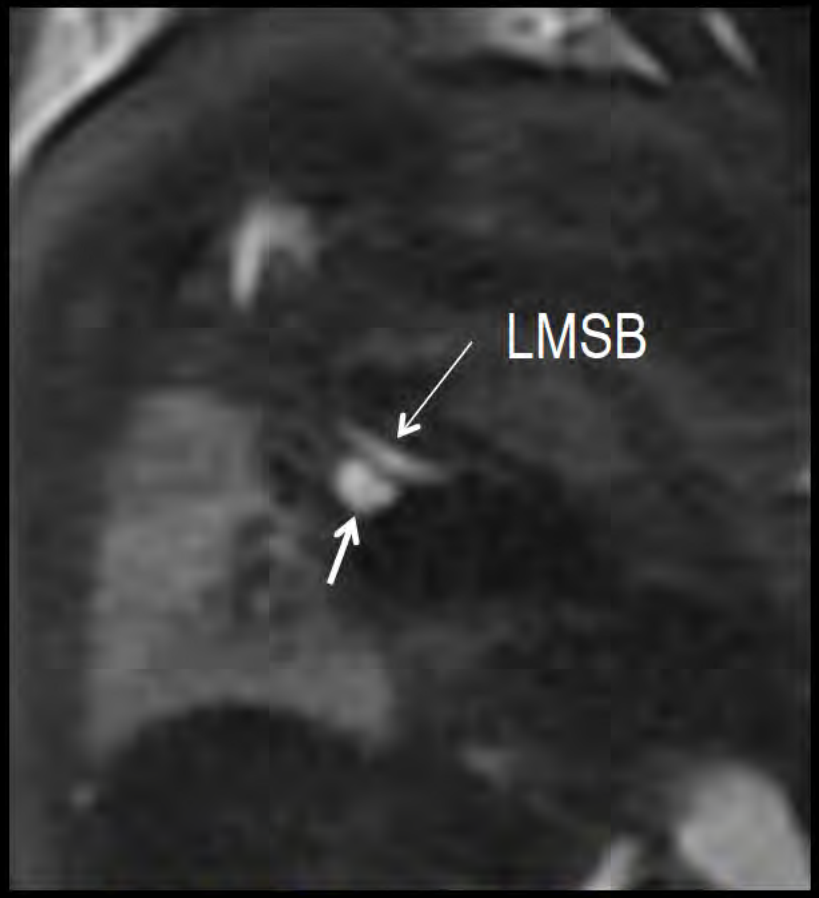
Solid

Cystic

Incidental Left Lung Mass at 23 weeks GA

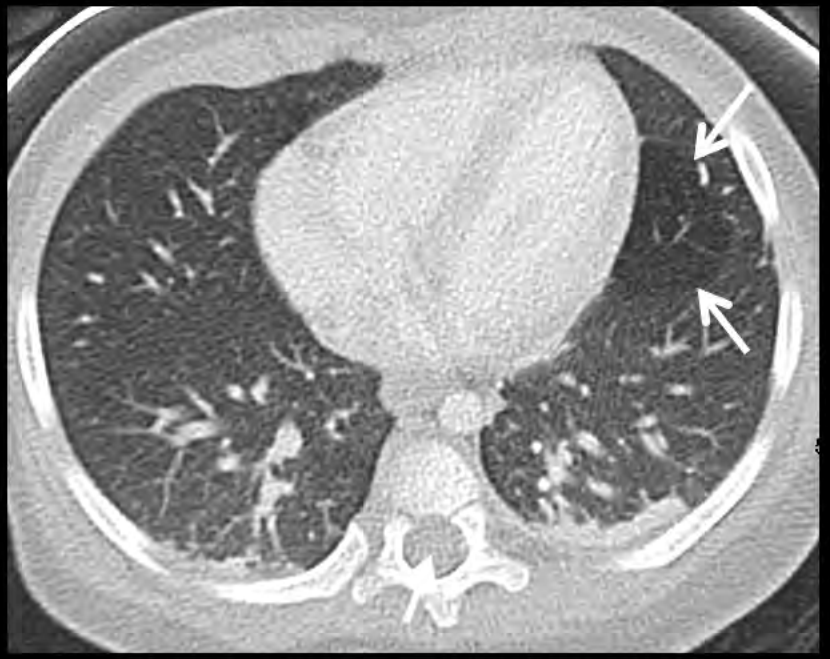


Incidental Left Lung Mass at 23 weeks GA



Additional Finding: Bronchogenic Cyst

Incidental Left Lung Mass at 23 weeks GA



CT Scan at 6 months

Hyperinflation 2° to Bronchogenic Cyst

Congenital Lobar Overinflation
*–Not an uncommon cause for a fetal chest mass**

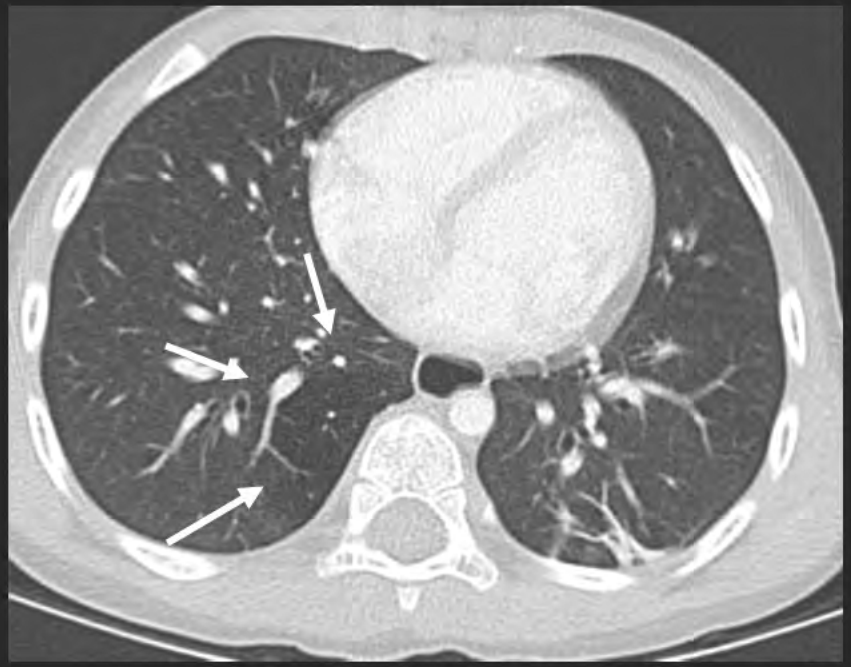
- CLO accounted for 29% of prenatally diagnosed chest masses
- Accurate Dx is important as these lesions may be managed conservatively

*Barth RA et al, 2010 ESPR

Prenatal Diagnosis: Right Lower Lobe CLO

Asymptomatic thru 6 years of age

22 weeks Gestational Age



4 years old

Right Lower Lobe CLO

Asymptomatic thru 36 months

36 weeks Gestational age

19 months old

Congenital Lobar Overinflation
*–Not an uncommon cause for a fetal chest mass**

- CLO accounted for 29% of prenatally diagnosed chest masses
- 9/10 cases asymptomatic (1- 6 years old)
- 1/10 cases symtomatic (Newborn)

*Barth RA et al, 2010 ESPR

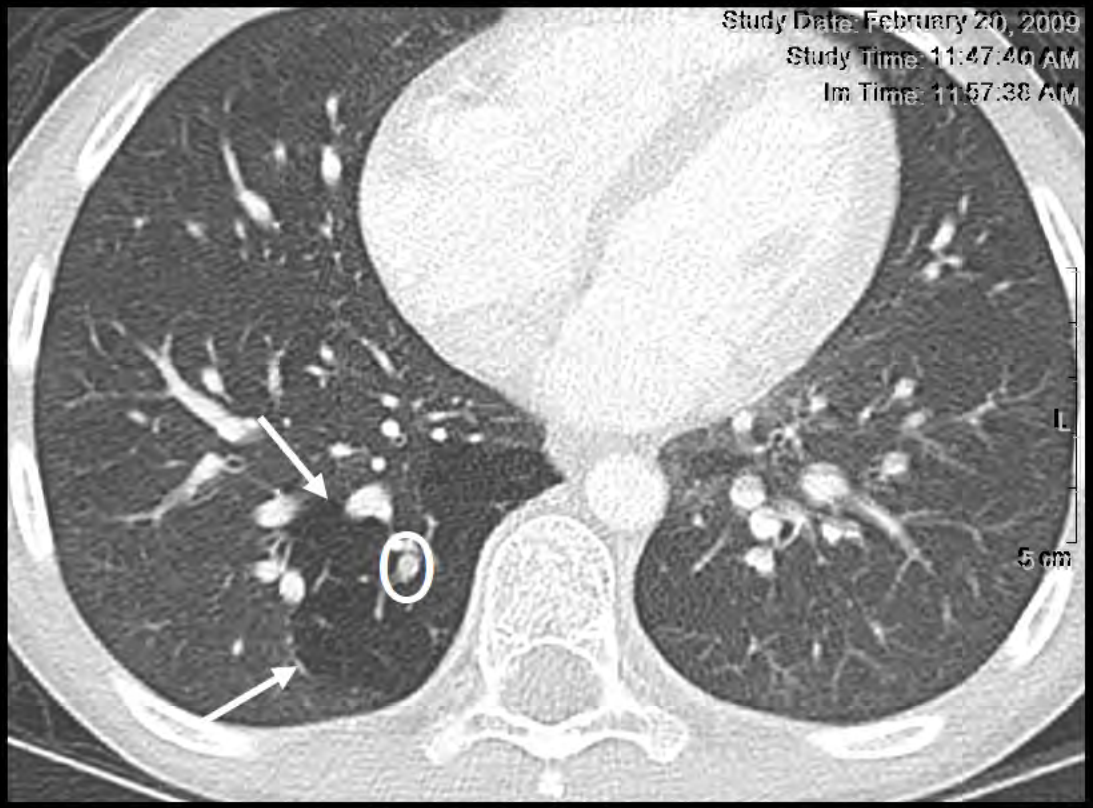
Postnatal Diagnosis: Right Lower Lobe CLO

Incidental CLO on CT for Appendicitis

Asymptomatic thru 9 years of age

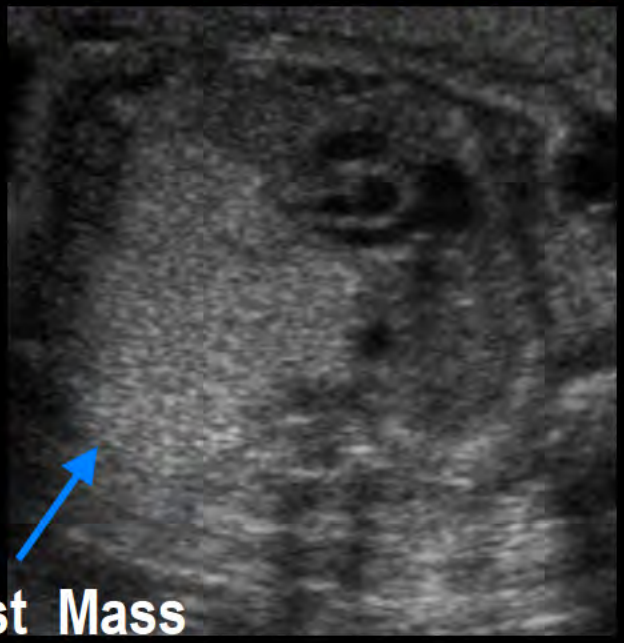
Postnatal Diagnosis: Right Lower Lobe CLO

Incidental CLO on CT for Appendicitis

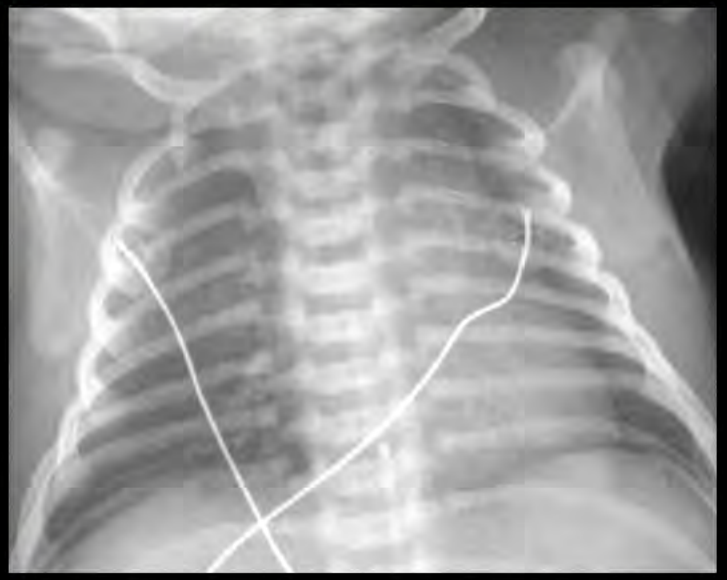


Asymptomatic thru 9 years of age

Historical Imaging Algorithm for Fetal Lung Malformations



Chest Mass



Normal CXR in Asx Newborn

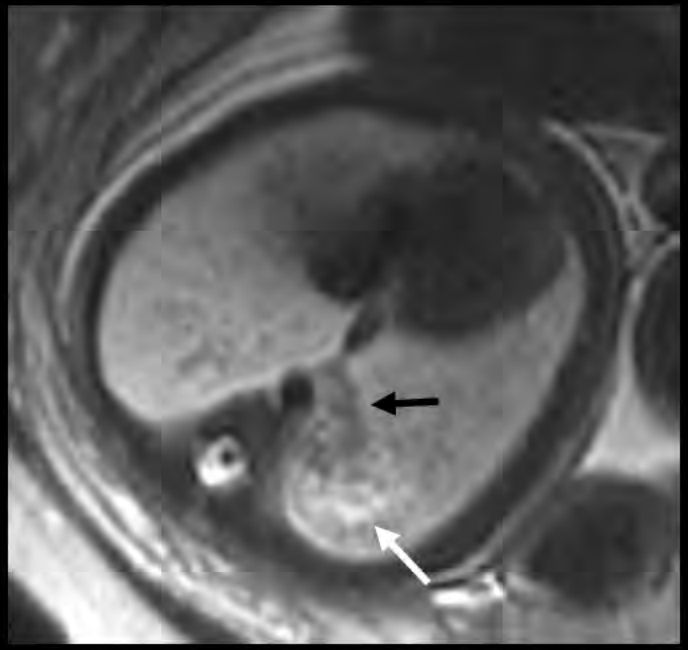
Newborn CT

3rd Trimester MRI correlates with Newborn CT*

(Lesion Visualization, Size, and Mass Effect)

MRI

CT



36 weeks GA

Newborn

Congenital Pulmonary Airway Malformation

*Rubesova E, 2007; SPR abstract

Equivocal Ultrasound for Chest Mass

Left Chest

22 wks GA

Equivocal Ultrasound for Chest Mass

Left Chest

22 wks GA

MR confirms RLL Lung Mass

23 wks GA

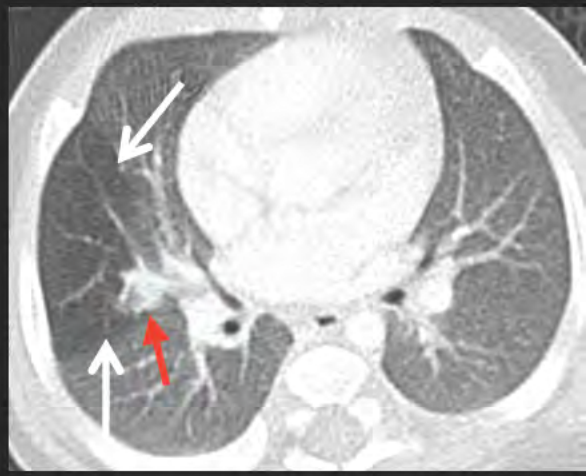
Right lower lobe CLO and Bronchial Atresia

Asymptomatic thru 2 years of age

Left Chest

22 wks GA

23 wks GA



3 months old

Cardiac Dextroposition

22 Weeks GA

Cardiac Dextroposition 2° to Hypoplastic Rt Lung

22 Weeks GA

22 wks GA

Cardiac Dextroposition 2° to Hypoplastic Rt Lung

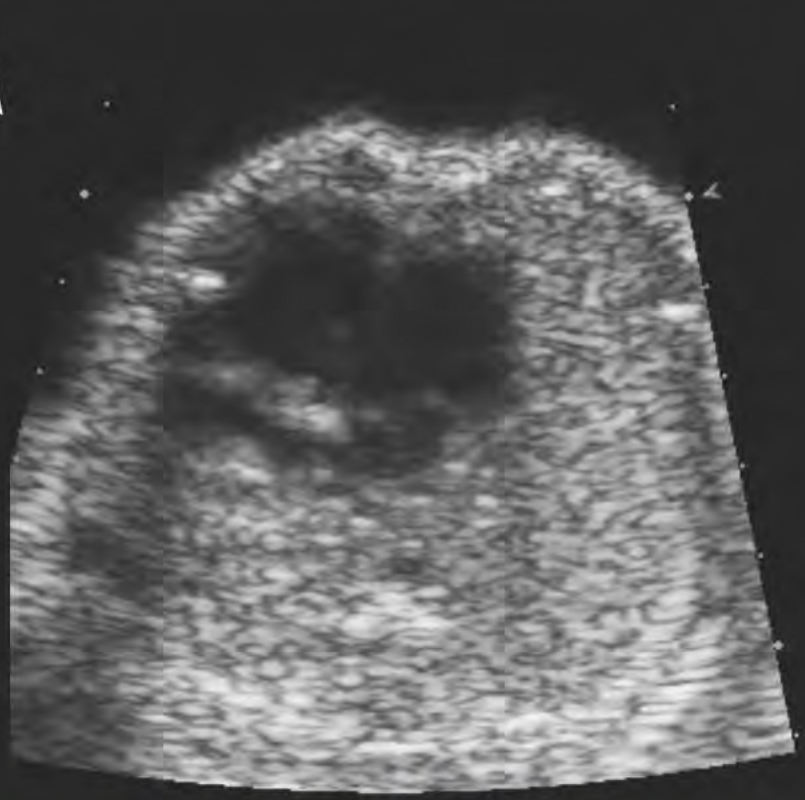
22 Weeks GA

22 wks GA

3 Months Old

Cardiac Dextroposition

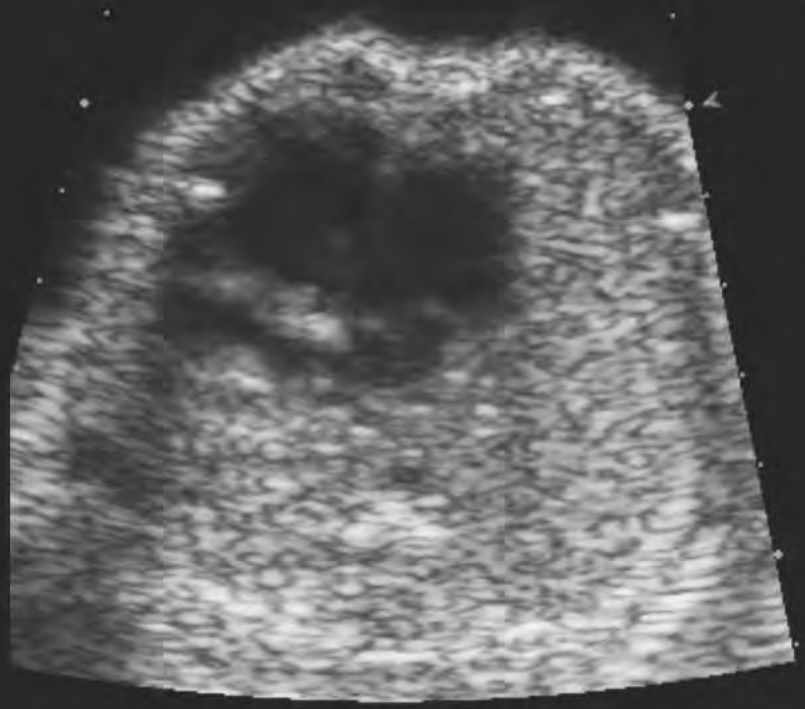
Rt Chest



Equivocal Ultrasound for Chest Mass

Cardiac Dextroposition 2° to Right Lung Hypoplasia

Rt Chest



Equivocal Ultrasound for Chest Mass

Horseshoe Lung & Right Lung Hypoplasia

Rt Chest

32 wks GA

Horseshoe Lung & Right Lung Hypoplasia

Rt Chest

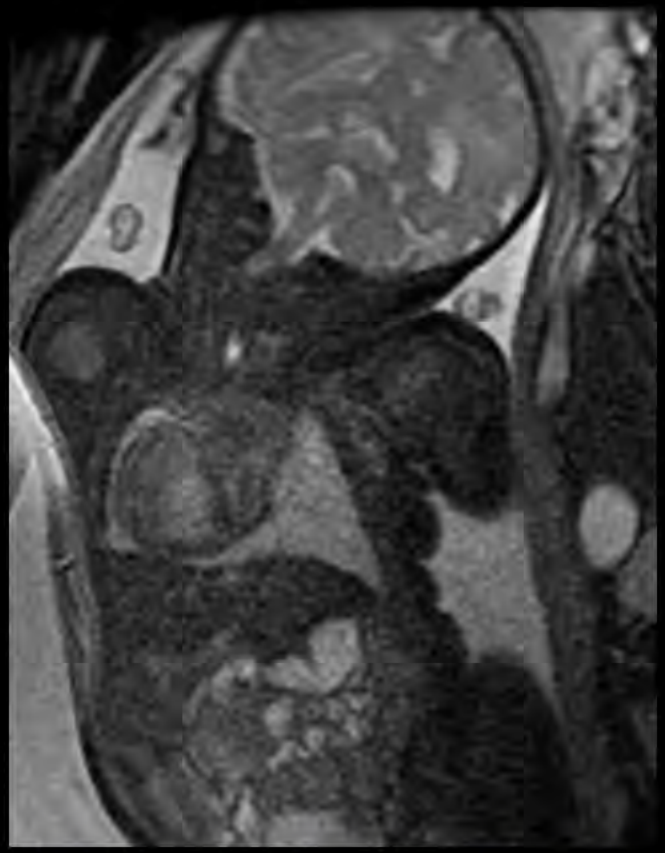
Newborn

Rt Chest

32 wks GA

Equivocal Ultrasound for Chest Mass
Cardiac Dextroposition

Rt



Lt

Rt

Lt

Horseshoe Lung & Right Lung Hypoplasia

Scimitar Syndrome & Right Lung Hypoplasia

Rt

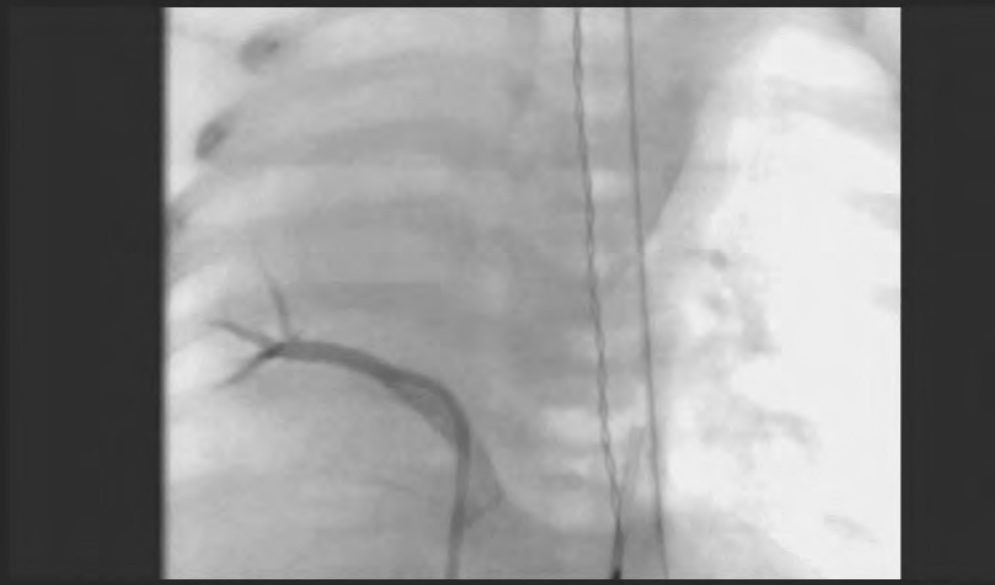
Lt

Rt

Lt

Scimitar Vein

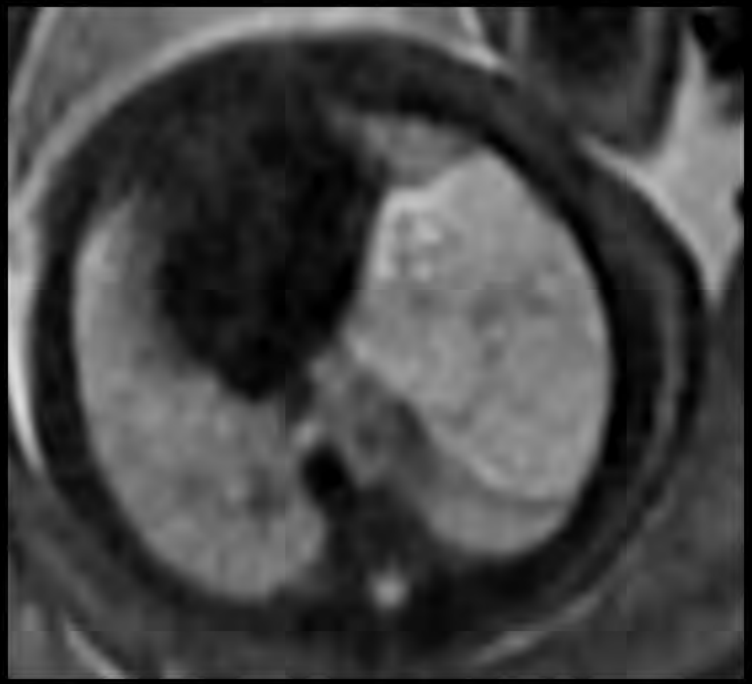
Scimitar Syndrome & Right Lung Hypoplasia



Other Causes of Lung Masses

Cystic Pleuropulmonary Blastoma

Fetal Ultrasound

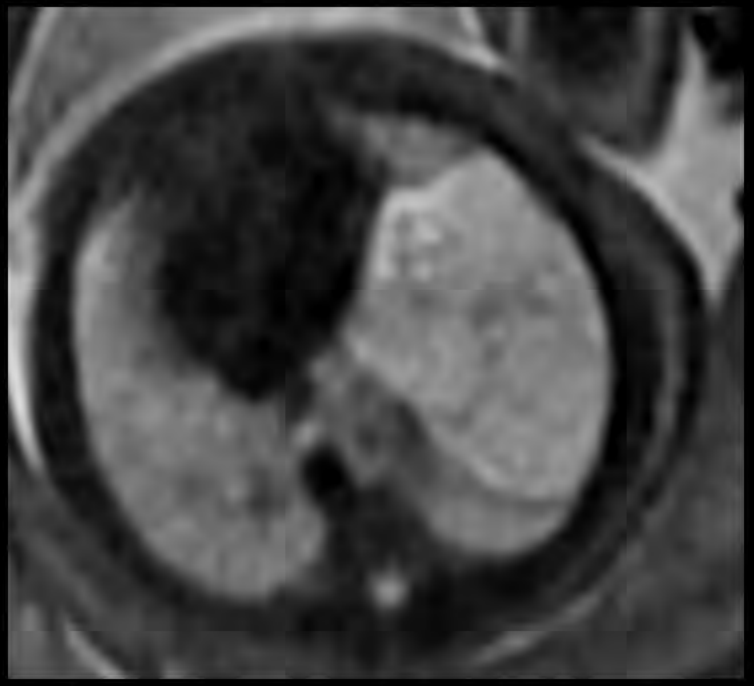


Fetal MRI

*Courtesy of Beth Kline-Fath, MD, Cincinnati Children's Hospital

Cystic Pleuropulmonary Blastoma

Newborn CT



Fetal MRI

*Courtesy of Beth Kline-Fath, MD, Cincinnati Children's Hospital

Cystic Pleuropulmonary Blastoma (PPB)

- Rare malignant lung tumor in children
  - aka pulmonary blastoma, pulmonary sarcoma, pulmonary rhabdomyosarcoma
- Thought to arise from pleuropulmonary germ cells
- Cystic PPB has been detected prenatally and is indistinguishable from macrocystic CPAM

Summary

- Ultrasound is primary modality for Dx and Predicting Outcome of Fetal Lung Mass
- Most complements
  - -Additional Findings
  - -Equivocal Cases
- Lobar Overinflation-Not and Uncommon Dx
- MRI may change Postnatal Imaging algorithm