Fetal Chest

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Disclosures

Christopher Cassady MD

Relevant Financial Relationships: NONE

Learning Objectives

After completing this presentation, the learner will be able to:

1. Understand the pathologies seen by ultrasound in the fetal chest;
2. Be able to recognize the basic differences among pathologies;
3. Offer a reasonable differential diagnosis for a fetal chest lesion; and
4. Have an understanding of the natural history for follow up.

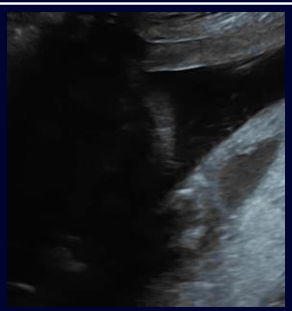
Introduction

1. Lung
2. Pleural space
3. Mediastinum
4. Chest Wall

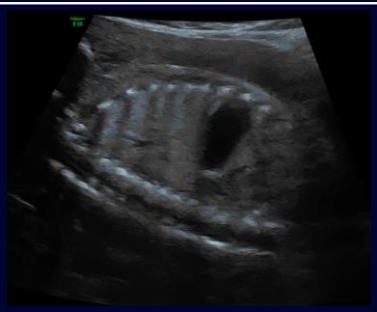
Fetal Chest

Normal

Fetal Chest



Fetal Chest



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Fetal Chest

1. Lung

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Fetal Chest

Congenital Lung Malformations

Neoplasm

Nutmeg lung

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Fetal Chest

Congenital Lung Malformations

- **Bronchial Atresia**
- **Sequestration**
- **CCAM**
- **Bronchogenic cyst**

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Fetal Chest: BA

Congenital Lung Malformations

- **Bronchial Atresia**
  key features: **mass effect**
  homogeneity
  pulmonary flow
  less conspicuous with time

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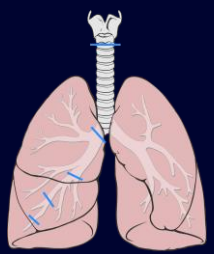
Fetal Chest: BA

Atresia can occur at any level from distal subsegmental to central mainstem and laryngotracheal.



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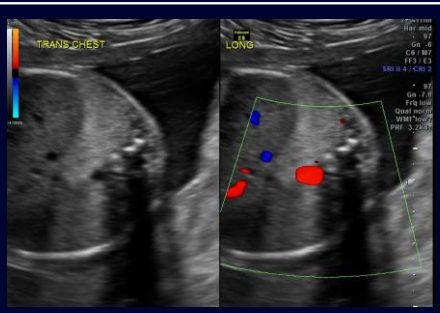
Fetal Chest: BA



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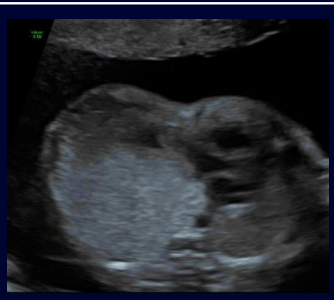
Fetal Chest: BA



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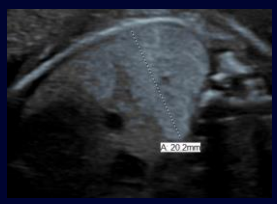
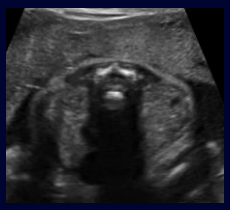
Fetal Chest: BA





21 weeks

29 weeks

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Fetal Chest: BA

Congenital Lung Malformations

central lesions stay large

- **Lobar Bronchial Atresia**
- **Congenital Lobar Overinflation**
- **CHAOS**

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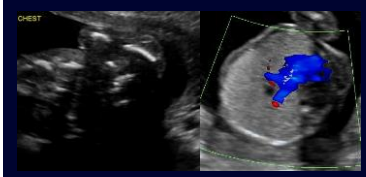
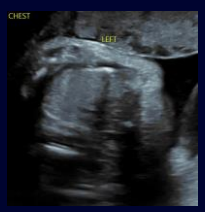
Fetal Chest: BA





21 weeks

30 weeks

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Fetal Chest: CLO

Congenital Lung Malformations

central lesions stay large

- **Lobar Bronchial Atresia**
- **Congenital Lobar Overinflation**
- **CHAOS**

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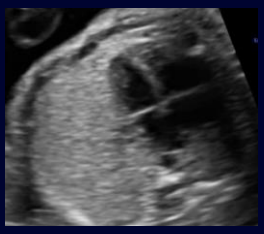
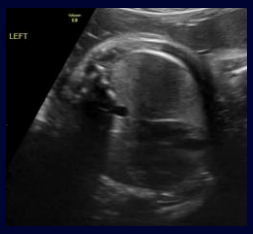
Fetal Chest: CLO



26 weeks



35 weeks

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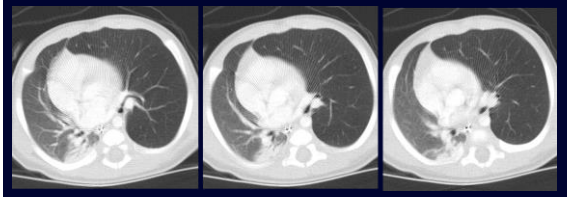
Fetal Chest: CLO



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Fetal Chest: CHAOS

Congenital Lung Malformations

central lesions stay large

- **Lobar Bronchial Atresia**
- **Congenital Lobar Overinflation**
- **CHAOS** **bilateral huge lungs, ascites**
  'unilateral CHAOS' = mainstem atresia

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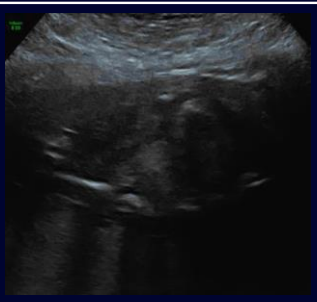
Fetal Chest: CHAOS



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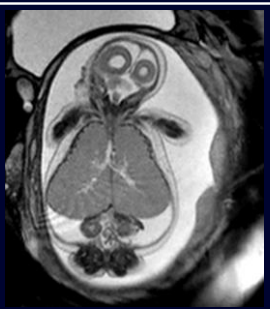
Fetal Chest: CHAOS



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Fetal Chest: Sequestration

Congenital Lung Malformations

- **Sequestration**
  key features: **mass effect**
  homogeneity
  systemic **flow**
  less conspicuous with time

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Fetal Chest: Sequestration

Congenital Lung Malformations

- **Sequestration**
  2 varieties:
  Intralobar venous return to heart
  Extralobar systemic venous return

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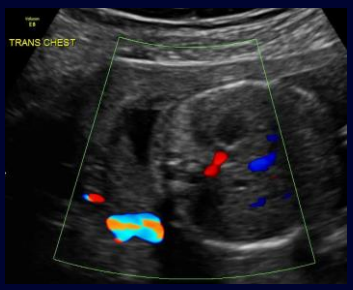
Fetal Chest: Sequestration



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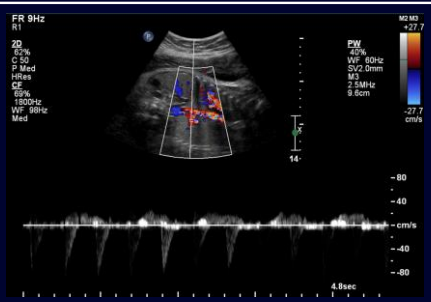
Fetal Chest: Sequestration



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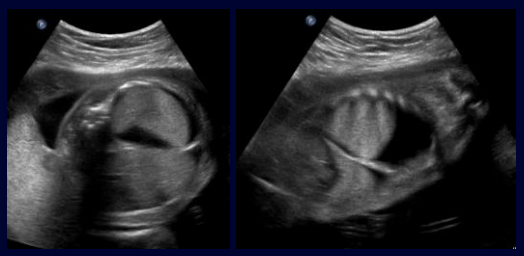
Fetal Chest: Sequestration



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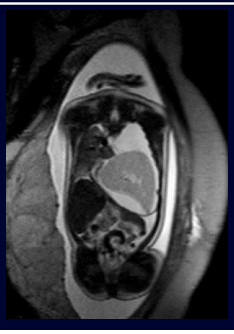
Fetal Chest: Sequestration



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Fetal Chest: CCAM

Congenital Lung Malformations

- **CCAM (CPAM)**
  key features: **mass effect**
  inhomogeneity
  cysts
  stabilize after 26 weeks

  divided into small or large (>2 cm) cyst types

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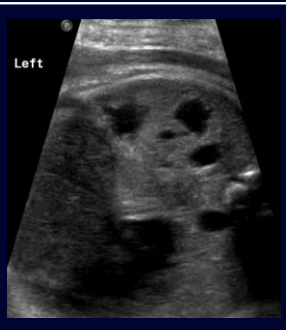
Fetal Chest: CCAM



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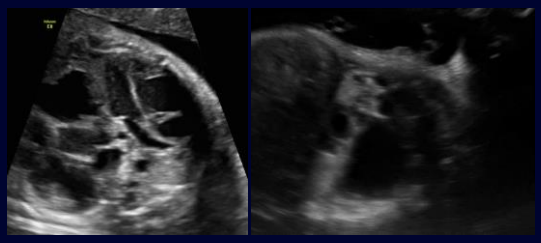
Fetal Chest: CCAM



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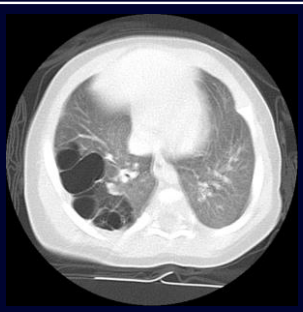
Fetal Chest: CCAM



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Fetal Chest: CCAM

Congenital Lung Malformations

- **CCAM (CPAM)**
  key features: mass effect
  inhomogeneity
  solid?

Stocker type III "CPAM": controversial. Many now believe this is a central atresia because the pathology is identical to CHAOS. Large lesion that looks like a lobar atresia on imaging.

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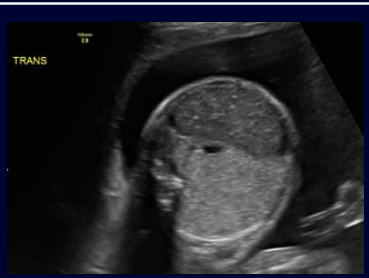
Fetal Chest: CCAM



So-called "type III (microcystic)" CCAM

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Fetal Chest: BC

Congenital Lung Malformations

- **Bronchogenic cyst**
  key features: single cyst, often central
  +/- distal obstruction
  no change in size over time

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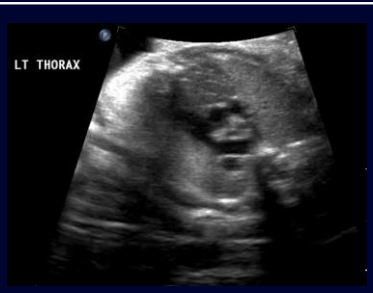
Fetal Chest: BC



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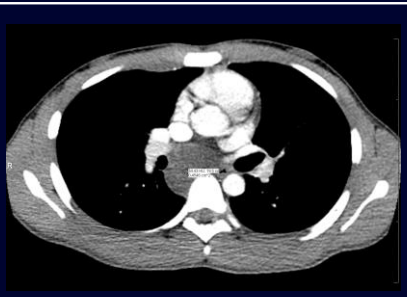
Fetal Chest: BC



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Fetal Chest: Neoplasm

Lung Neoplasms

- Pleuropulmonary blastoma
- Fetal Lung Interstitial Tumor
- Congenital Peribronchial Myofibroblastic Tumor

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Fetal Chest: Neoplasm

Lung Neoplasms

- Pleuropulmonary blastoma — cystic
- Fetal Lung Interstitial Tumor — solid
- Congenital Peribronchial Myofibroblastic Tumor — solid

key feature: growth after 26-28 weeks

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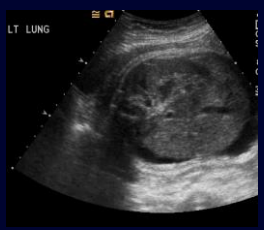
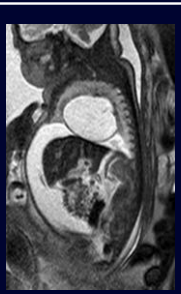
Fetal Chest: Neoplasm





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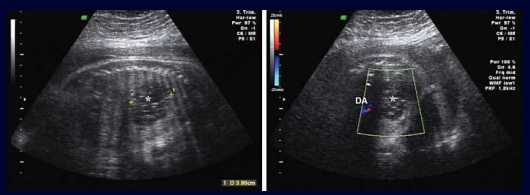
Fetal Chest: Neoplasm



Coleman A, Kline-Fath B, Stanek J, Lim FY. Pleuropulmonary Blastoma in a Neonate Diagnosed Prenatally as Congenital Pulmonary Airway Malformation. Fetal Diagn Ther. 2015 Jul 24. [Epub ahead of print]

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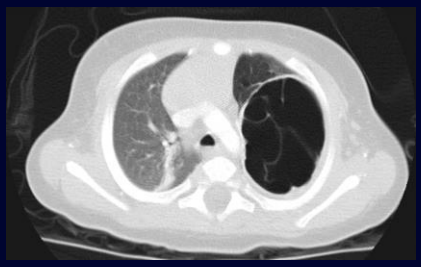
Fetal Chest: Neoplasm



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Fetal Chest: CVR

Why does any of this make a difference?

CVR **"CCAM Volume Ratio" = CLM volume ratio**

$$\frac{\text{length x width x height x 0.52}}{\text{head circumference}}$$

>1.6

Crombleholme TM et al. Cystic adenomatoid malformation volume ratio predicts outcome in prenatally diagnosed cystic adenomatoid malformation of the lung. J Pediatr Surg 2002;37(3):331-338.

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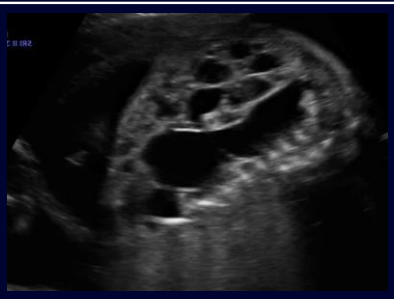
Fetal Chest: CVR



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Fetal Chest: CLM Imaging

IMAGING ALGORITHM

- ***Characterize lesion on US***
  - **"CVR" (CLM/Mass Volume Ratio)**
  - **Primarily solid or primarily cystic (solitary, or multiple?)**
  - **Systemic or pulmonary artery, vein**
  - **Hydrops (get echocardiogram for cardiac function)**
- ***MRI/echocardiogram***
- ***Antenatal imaging follow up*** **closely until 28 weeks (growth plateau); 34 weeks: √ mediastinal shift, confirm dx**
- ***Postnatal Chest CT***

Beydon N et al. Comparison between US and MRI in the prenatal assessment of lung malformations. Pediatr Radiol 2013;43:685-696

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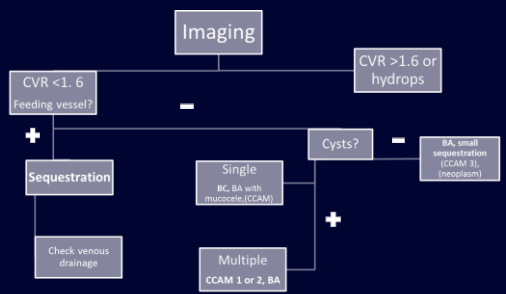
Fetal Chest: CLM Imaging



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Fetal Chest: CLM Treatment

Treatment

Prenatal: cardiac dysfunction **steroids (CVR >1.6)**
shunt dominant cyst
fetal surgery

Postnatally, they are almost all excised eventually (except subsegmental BA)

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Fetal Chest: Nutmeg Lung

Nutmeg Lung

key features: enlarged, heterogeneous, symmetric
typically small effusions
Classically from HLHS
if no structural cardiac abnormality,
suspect primary lymphangiectasia

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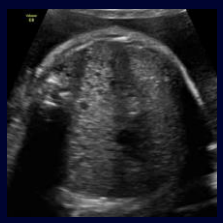
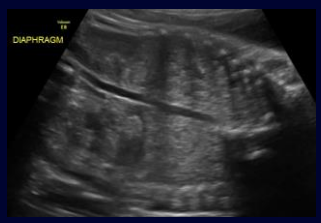
Fetal Chest: Nutmeg Lung





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Fetal Chest

2. Pleural Space

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Fetal Chest

Pleural Space

- Intrapleural CDH
- Isolated pleural effusion

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Fetal Chest: CDH

Congenital Diaphragmatic Hernia

Key features: heart displaced (mass effect)
heterogeneity to chest on side of hernia
+/- stomach in chest (if left iCDH)
?liver up?
if stomach not to front ribs, yes
if right iCDH, yes
use color Doppler to help measure LHR

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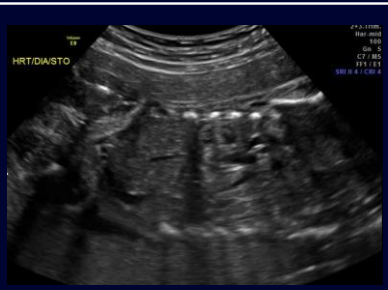
Fetal Chest: CDH



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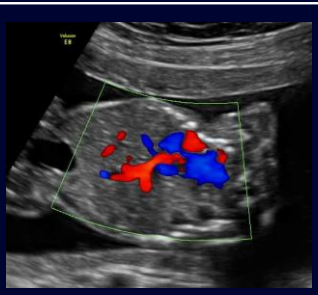
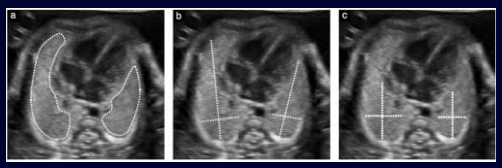
Fetal Chest: CDH



Cassady

Fetal Chest: CDH



The Lung:Head ratio

Peralta CFA, Cavoretto P, Csapo B, Vandecruys H, Nicolaides KH. Assessment of lung area in normal fetuses at 12-32 weeks. Utlrasound Obstet Gynecol 2005;26:718-724.

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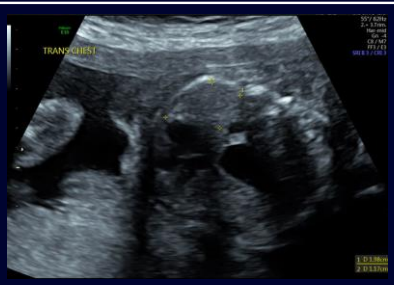
Fetal Chest: CDH



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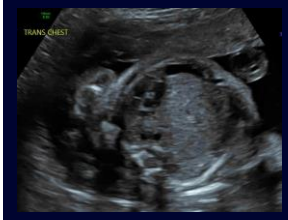
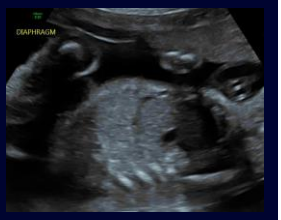
Fetal Chest: CDH





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Fetal Chest: Effusion

Pleural effusion

Key features: simple fluid surrounds lung

exclude chromosomal/structural cause
(including cardiac, trisomies)
then ddx = chylous, infection, hydrops

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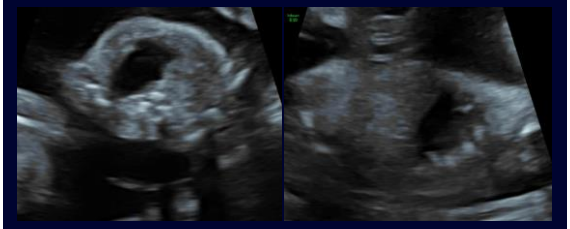
Fetal Chest: Effusion



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Fetal Chest

3. Mediastinum

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Fetal Chest

Mediastinum

- Mediastinal CDH
- Teratoma
- (Neural tumor)
- (Vascular malformation)

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Fetal Chest: CDH

Mediastinal CDH

Key features: lung volumes good
anterior: displaces heart back
middle: stomach up on right,
but not liver

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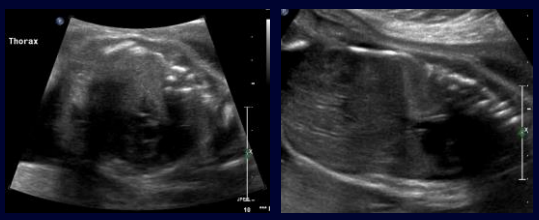
Fetal Chest: CDH



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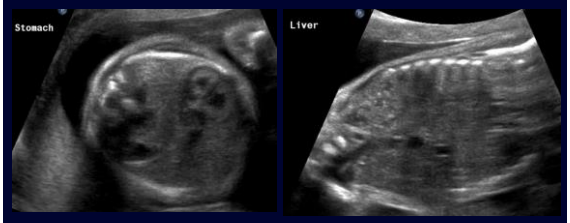
Fetal Chest: CDH



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Fetal Chest: Teratoma

Teratoma

Key features: heterogenous mass
cystic and solid
calcifications
vascular on Doppler

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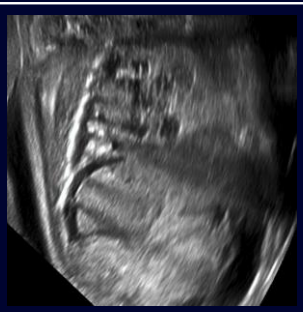
Fetal Chest: Teratoma



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Fetal Chest

4. Chest Wall

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Fetal Chest

Chest Wall

- **Thoracic Hypoplasia**
- **Lymphatic malformation**
- **Hamartoma**

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Fetal Chest: Hypoplasia

Thoracic hypoplasia

Oligo/anhydramnios
Rib constriction: skeletal dysplasia
Lung hypoplasia: masses (CDH, cardiac);
increased abdominal pressure;
abnormal diaphragm function
(eg. giant omphalocele)

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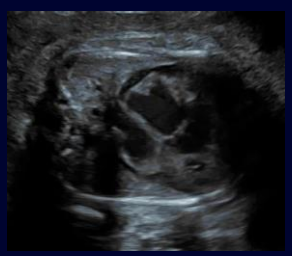
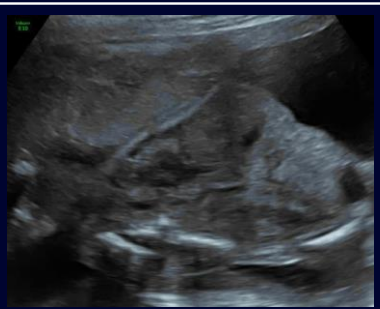
Fetal Chest: Hypoplasia



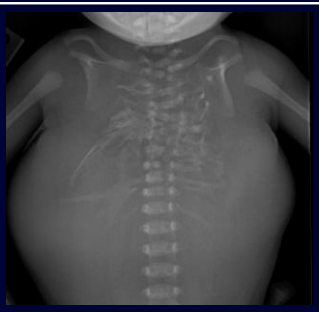
Cassady

Fetal Chest: Hypoplasia



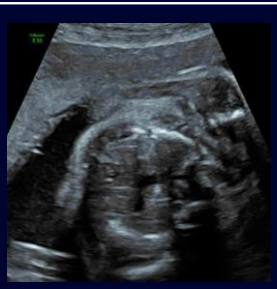
Cassady

Fetal Chest: Hypoplasia



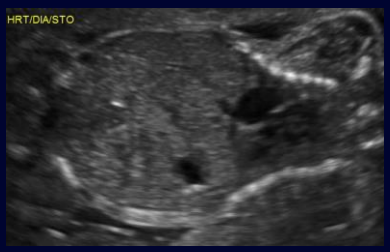
Cassady

Fetal Chest: Hypoplasia



Cassady

Fetal Chest: Hypoplasia



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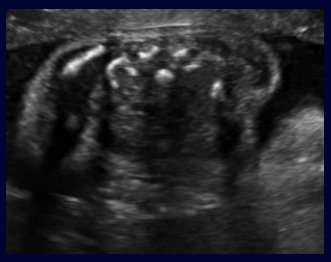
Fetal Chest: Hypoplasia



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Fetal Chest: LM

Lymphatic malformation

Key features: **multicystic infiltrating mass typically from neck or axilla**

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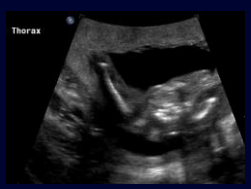
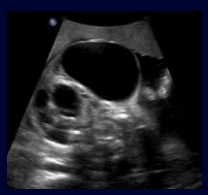
Fetal Chest: LM





21 weeks **32 weeks**

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Fetal Chest: Hamartoma

Mesenchymal hamartoma

Key features: heterogeneous
calcification of margins
deforms local ribs

benign. very rare.

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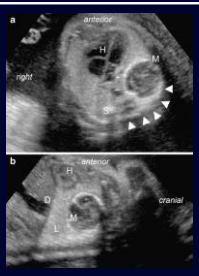
Fetal Chest: Hamartoma



Chu L, Seed M, Howse E, Ryan G, Grosse-Wortmann L. Mesenchymal hamartoma: prenatal diagnosis by MRI. Pediatr Radiol 2011 Jun;41(6):781-4

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Fetal Chest: MRI

Consider MRI:

1. if intervention is possible
2. when the fetal airway may be compromised
3. to assess pulmonary hypoplasia
4. when there are still unanswered questions important for safe delivery planning

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Conclusions

LUNG:	congenital lung malformations rarely neoplasm, lymphangiectasia
PLEURAL SPACE:	congenital diaphragmatic hernia effusion
MEDIASTINUM:	diaphragmatic hernia rarely neoplasm
HEART:	structural CHD neoplasm, effusion
CHEST WALL:	anhydramnios, skeletal dysplasia lymphangioma, hamartoma

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Key References

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