


## Ultrasound Evaluation of Fetal Chest

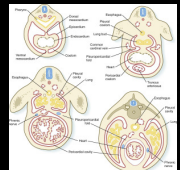



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 Texas Children's Hospital, Pavilion for Women  
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 &  
 Clinical Instructor  
 Thomas Jefferson University Hospital - Radiology Department  
 Philadelphia, Pennsylvania

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## Embryology & Pathogenesis

- Normal closure of pleuroperitoneal folds
  - 4th -10th wks GA
- Lung development
  - 3rd -16th wks GA

Fetal Chest

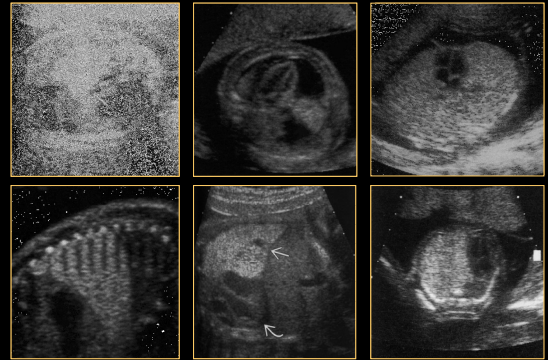
## Approach to the Fetal Chest



- Chest configuration
- Position of heart
- Size or growth of chest
- Presence of any masses
- Presence of fluid

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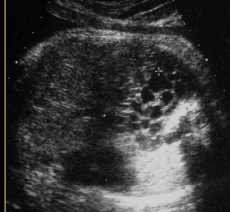


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## Fetal Lung Masses: DDX

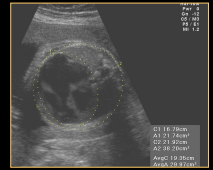
- Developmental Anomalies
  - Cystic adenomatoid malformation (CCAM)
  - Cystic pulmonary airway malformation (CPAM)
  - Bronchopulmonary sequestration (BPS)
    - Intralobar
    - Extralobar
  - “Hybrid” lesions
  - Bronchial atresia
  - Bronchogenic cyst
  - Lobar emphysema
- Neoplasms
  - Mesenchymal neoplasm
  - Pleuropulmonary blastoma



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## Fetal Chest

- Nomograms for thoracic circumference, length
- Chest size can be compared to head, abd, heart



Fetal Chest

TC / AC stable throughout pregnancy  
Normal > .80

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## Approach to the Fetal Chest Mass

- Is the chest normal in size?
- Is the axis of the heart deviated?
- Where is the stomach?
- Is the mass cystic or solid?
- If cystic, is it a simple cyst or a complex cystic mass?
- If solid, what does the Doppler show?
- Where is the mass?
- Does the mass extend beyond the chest wall?
- Are there other anomalies?
- Is there hydrops?
- What's the likelihood of pulmonary hypoplasia?

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## Congenital Diaphragmatic Hernia

- This is a developmental discontinuity of the diaphragm that allows abdominal viscera to herniate into the chest, resulting in
  - Pulmonary hypoplasia
  - Pulmonary hypotension



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## Sites of Herniation

- **Foramen of Morgagni**
  - Partial or complete absence of the central diaphragm
  - anteromedial retrosternal portion of diaphragm
- **Foramen of Bochdalek**
  - Posterolateral corner of the diaphragm fails to close
  - More common

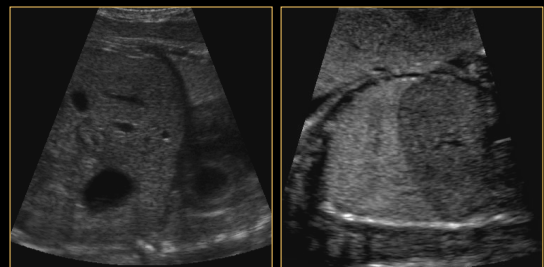


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## Diaphragm

*“imperative to view entire diaphragm”*



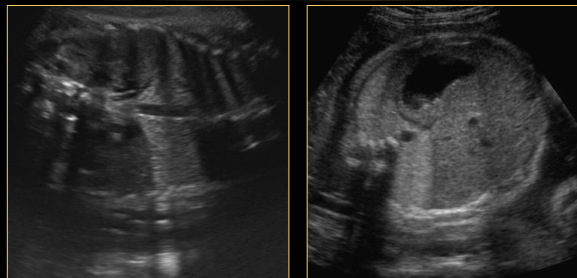
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Most CDHs are posterior so coronal view of anterior diaphragm may be normal

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## Diaphragm

*“imperative to view entire diaphragm”*

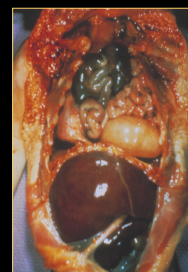


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## Congenital Diaphragmatic Hernia

- All infants have postnatal respiratory distress related to pulmonary hypoplasia
- If IUGR, 90% have other major anomaly
- 20% have cardiac & 30% have CNS anomalies
- Chromosomal abnormalities

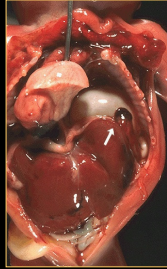


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## Congenital Diaphragmatic Hernia

- Left-sided 85%
  - Bowel, stomach, spleen
- Right-sided 15%
  - Bowel, liver, GB
- Pulmonary hypoplasia
  - Variable
  - Is worse for CDH than other chest masses of comparable size



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## Congenital Diaphragmatic Hernia

- Mediastinal shift
- Abdominal circumference small (< 5th percentile)
  - Liver, gallbladder, **stomach**, bowel or spleen in chest
  - May see peristalsis in the chest
- AFV: poly, oligo, normal



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## Congenital Diaphragmatic Hernia



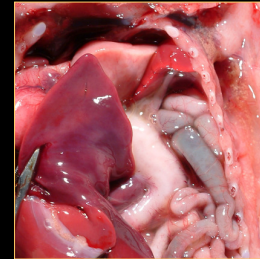
Clubbing of the hand and webbing at the wrist and elbow consistent with early onset of fetal akinesia

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## Congenital Diaphragmatic Hernia

- How does the residual lung look like?

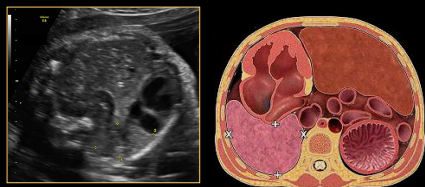


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## Congenital Diaphragmatic Hernia

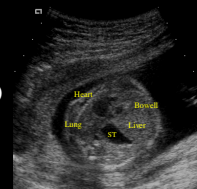
- Lung area to head circumference ratio < 1 is associated with a high rate of neonatal death due to pulmonary hypoplasia



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## CDH: Predictors of Outcome

- LHR: Lung-to-head ratio
  - >1.2 = 79% survival (30/38)
  - 0.9-1.2 = 59% survival (13/24)
  - < 0.9 = 4% survival (1/24)
- MRI volumetric assessment



Metkus AP, et al. Sonographic predictors of survival in fetal diaphragmatic hernia. J Pediatr Surg 31:148, 1996

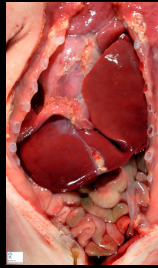
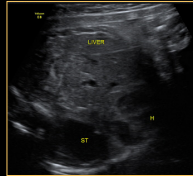
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## CDH: Predictors of Outcome

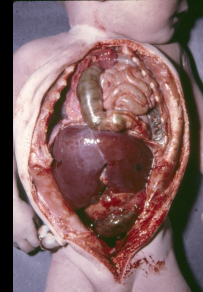
- Liver herniation as predictor of outcome
  - No: 79% survival
  - Yes: 41% survival

Walsh DS, et al. Am J Obstet Gynecol 18:1067, 2000



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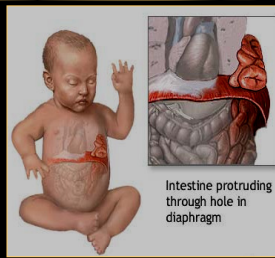
## How to Detect a Congenital Diaphragmatic Hernia



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## Congenital Diaphragmatic Hernia



Left-sided hernias more frequent than right-sided ones

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## Congenital Diaphragmatic Hernia



- Left Sided Hernia
  - Cystic or heterogenous mass in left side of chest
  - Absence of fluid-filled stomach
  - Deviation of heart toward right
  - Polyhydramnios
  - Hydrops uncommon unless associated malformation present

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## Congenital Diaphragmatic Hernia

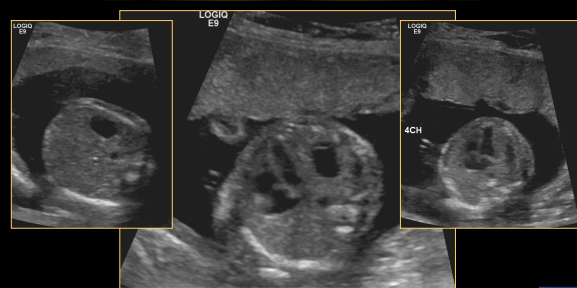


"If multiple ribs are seen, image is oblique"  
Incorrect scan plane may result in erroneous diagnosis

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## Congenital Diaphragmatic Hernia



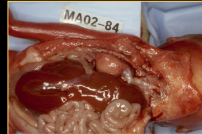
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## Congenital Diaphragmatic Hernia



- Right Sided Hernia
  - May be confused for chest mass
  - Contains liver and bowel
  - Stomach is below diaphragm
  - Gallbladder often herniated
  - Doppler will show portal & hepatic veins

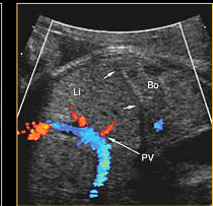
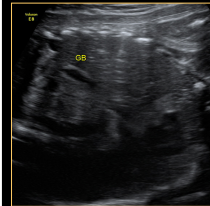


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## Congenital Diaphragmatic Hernia

- CDH with liver herniation
- Always use Doppler to evaluate for liver



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## Congenital Diaphragmatic Hernia

- Factors that has been associated with poor prognosis include:
  - Marked mediastinal shift associated with pulmonary hypoplasia
  - IUGR
  - Polyhydramnios
  - Detection prior to 24 weeks
  - Significant liver involvement



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## Extra Corporeal Membrane Oxygenation



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## Congenital Diaphragmatic Hernia

- Obstruction of trachea results in
  - Expansion of the fetal lungs by retained pulmonary secretions & lung tissue stretch, is associated with improved lung growth & development



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## Congenital Diaphragmatic Hernia

- Hysterotomy & fetal surgery
  - High maternal morbidity & no improved fetal survival
- Hysterotomy & endoscopically
  - Neck dissection & clipping of the trachea
  - High rate of preterm delivery
  - Irreversible damage to the laryngeal nerve & trachea

*J Pediatr Surg* 1997; 32: 1637-1642  
*Am J Obstet Gynecol* 2000; 183: 1059-1066  
*J Pediatr Surg* 2003; 38:1012-1020


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## Congenital Diaphragmatic Hernia

- Fetoscopic Endoluminal Tracheal Occlusion with balloon

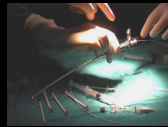
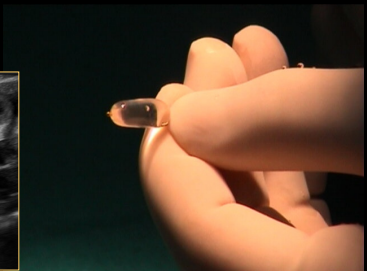
Ultrasound Obstet Gynecol 2004; 24:121-126  
Deprest, Belgium; Gratacos, Spain; Nicolaidis, England

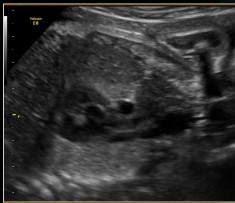




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## Tracheal Occlusion Fetoscopic endotracheal balloon


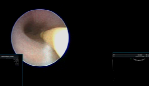
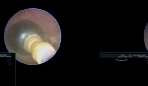


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## Fetoscopic Endoluminal Tracheal Occlusion


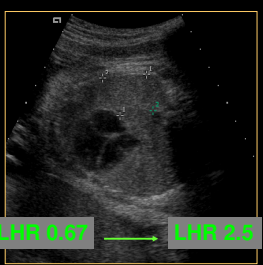
- Performed before 26-28 weeks gestational age on fetuses with poor prognosis
  - Liver up and LHR < 1.0
- Reverse occlusion at 34 weeks by fetoscopy or US guided balloon puncture
- EXIT – Surgery – ECMO (may be best strategy)

Ex Utero Intrapartum Treatment

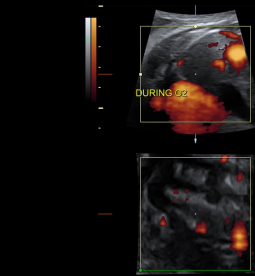
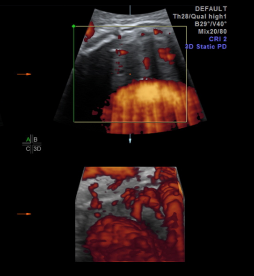
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## Severe CHD – 20 days post PLUG

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## Severe CHD – 20 days post PLUG

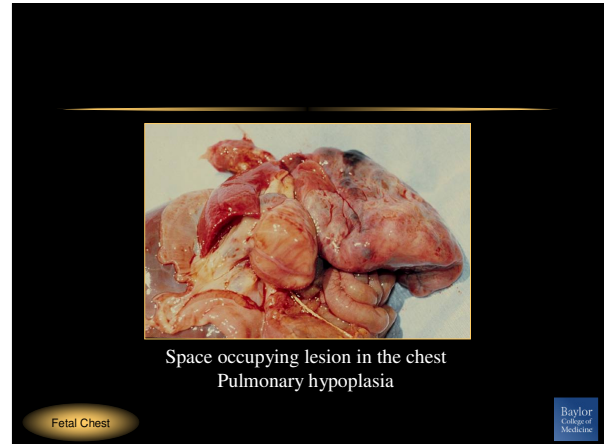
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## Congenital Diaphragmatic Hernia “Differential Diagnosis”

- Congenital cystic adenomatoid malformation
- bronchopulmonary sequestration
- Bronchogenic cyst
- Teratomas



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### Terminology

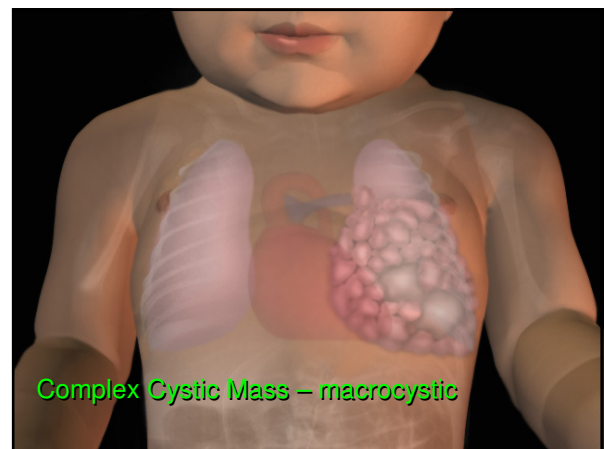
- **Congenital Cystic Adenomatoid Malformation**
  - Older name that reflects cystic and adenomatous histologic components of these masses
- **Congenital Pulmonary Airway Malformation**
  - Newer terminology reflects developmental disorder of pulmonary airway morphogenesis

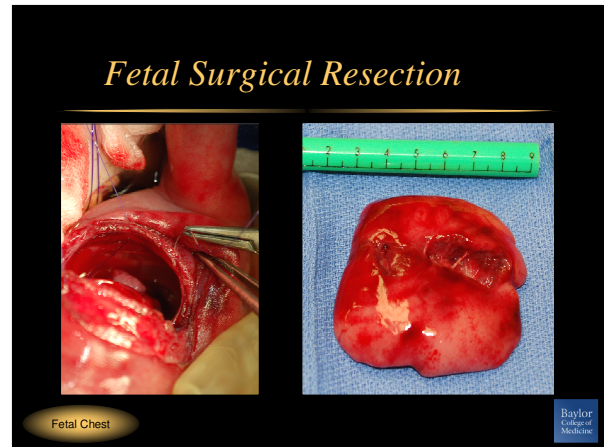
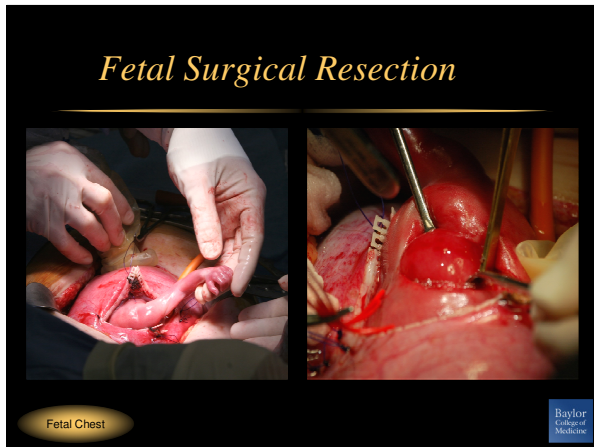
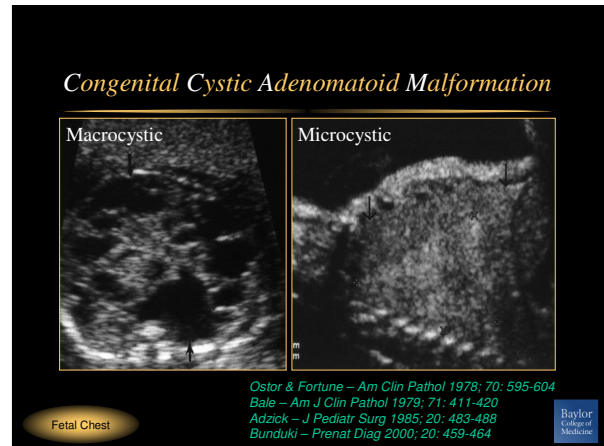
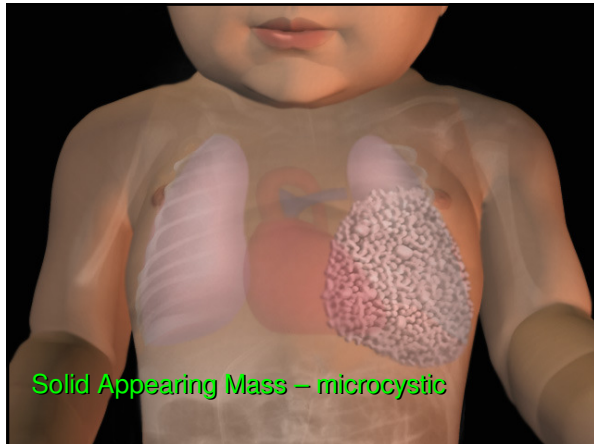
### Congenital Cystic Adenomatoid Malformation

- Pulmonary lesion due to abnormal development of lung tissue
  - Cystic dilatation of abnormal bronchi & maldevelopment of associated alveoli

### Congenital Cystic Adenomatoid Malformation

- This disorder is **NOT GENETIC**
- Can be accompanied by hydrops & mediastinal shift
- Almost exclusively **unilateral - left**
- Supplied by the **pulmonary artery**





### Congenital Cystic Adenomatoid Malformation

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History:

- First described by Chin & Tang (1949)

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### Congenital Cystic Adenomatoid Malformation

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- Classic 3 types;
- Type 1: Large cysts 3-10 cm
- Type 2: Cysts 0.5-2 cm
- Type 3: Solid

} Macrocystic

} Microcystic

Stocker 1977

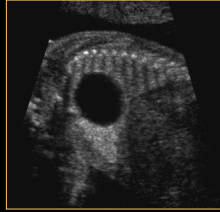
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## Congenital Cystic Adenomatoid Malformation

### Type I

- Macrocytic: cysts > 2cm
- Good prognosis after resection



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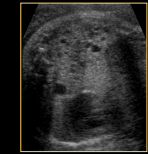
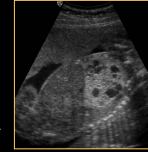
## Congenital Cystic Adenomatoid Malformation

### Type I

- Macrocytic: cysts > 2cm
- Good prognosis after resection

### Type II

- Medium-sized cysts: cysts < .5-2cm
- High incidence of anomalies, poor prognosis



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## Congenital Cystic Adenomatoid Malformation

### Type I

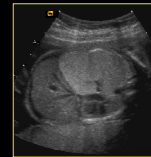
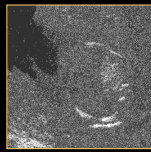
- Macrocytic: cysts > 2cm
- Good prognosis after resection

### Type II

- Medium-sized cysts: cysts < .5-2 cm
- High incidence of anomalies, poor prognosis

### Type III

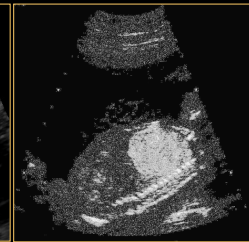
- Microcystic: cysts < 0.5cm
- Hyperechoic, looks solid
- Poor prognosis



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## CCAM vs. Normal Lung

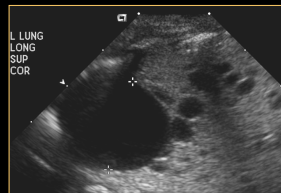
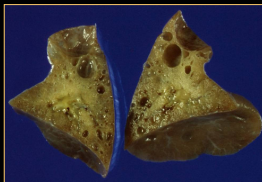


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## Congenital Cystic Adenomatoid Malformation

- May coexist with sequestration (50% ELS)

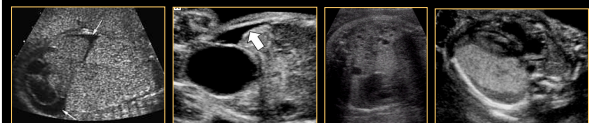


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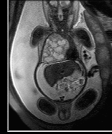
## Congenital Cystic Adenomatoid Malformation

- Heart is displaced
- Stomach is normal location
- Hydrops
  - Most important predictor of outcome
  - Dismal prognosis



## Lung Masses – Prognostic Indicators

- Calculate **CCAM Volume Ratio (CVR)**
  - CCAM volume calculated by measuring all 3 dimensions x 0.52
  - CCAM volume is then divided by HC
- CVR > 1.6 indicates increased risk of developing hydrops and fetal demise



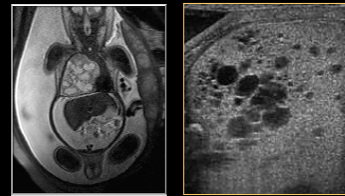
Crombleholme, et al. J Pediatr Surg 2002 37:331

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## Congenital Cystic Adenomatoid Malformation

- Look at the size of CCAM
- Look for evidence of heart failure
- Look for other anomalies - rare



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## Congenital Cystic Adenomatoid Malformation “Differential Diagnosis”

- Pulmonary sequestration
- Bronchogenic cyst
- Diaphragmatic hernia
- Mediastinal lesions
  - Enterogenous cyst
  - Neurenteric cyst
  - Cystic teratoma

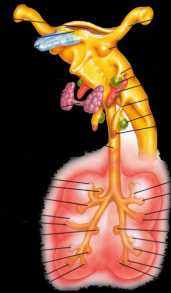


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## Bronchogenic Cyst

- Result from an abnormal development in the budding or branching of the tracheobronchial tree



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## Bronchogenic Cyst

- A singular cyst usually 1.5-2.0 cm in diameter
- Can be intrapulmonary or lie in the posterior mediastinum
- The cyst does not displace the heart
- May have mass effect and compress esophagus
  - polyhydramnios
- Rarely associated with hydrops



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## Pulmonary Sequestration

- Nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree

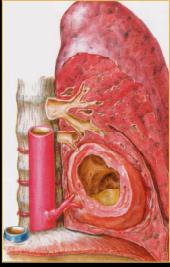
Intralobar pulmonary sequestration – 75%  
Extralobar pulmonary sequestration – 25%

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## Pulmonary Sequestration

- Intralobar sequestration
  - Located within the substance of a lung lobe



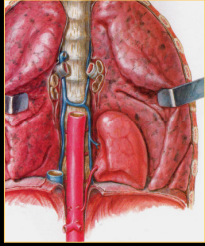
Other anomalies 14%

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## Pulmonary Sequestration

- Extralobar sequestration
  - Have separate pleural covering
  - Venous return to pulmonary veins
  - Arterial supply from thoracic or abdominal aorta \*



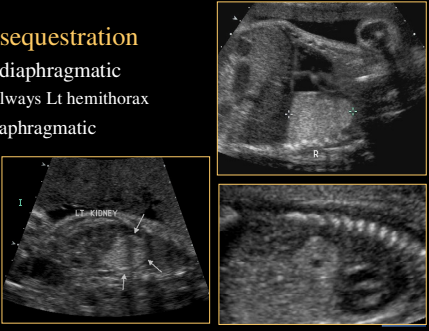
Other anomalies 60%

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## Pulmonary Sequestration

- Extralobar sequestration
  - 90% supradiaphragmatic
    - Almost always Lt hemithorax
  - 10% Subdiaphragmatic



R

LT KIDNEY

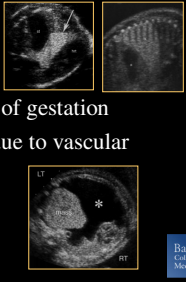
RT

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## Pulmonary Sequestration

- Appears as a well circumscribed uniformly echogenic mass
  - Triangular or lobar shape
- Mediastinal shift often is noted
- Lesion regress during the course of gestation
- Hydrops occasionally develops due to vascular compression





RT

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## Diagnostic Challenge

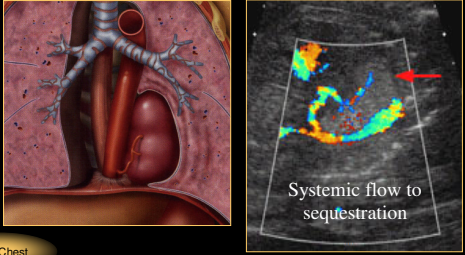
Pulmonary Sequestration  
Vs.  
Cystic Adenomatoid Malformation

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Fetal Chest

## Pulmonary Sequestration

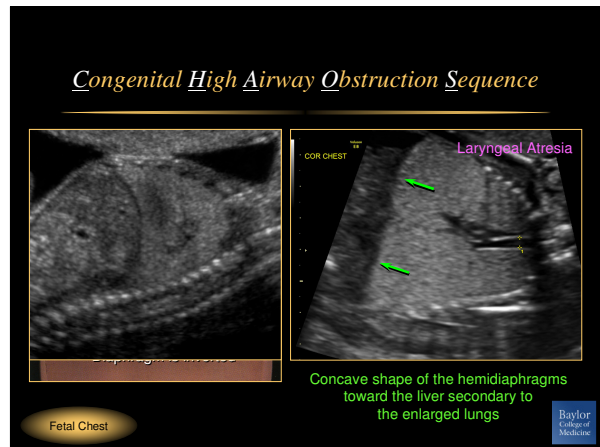
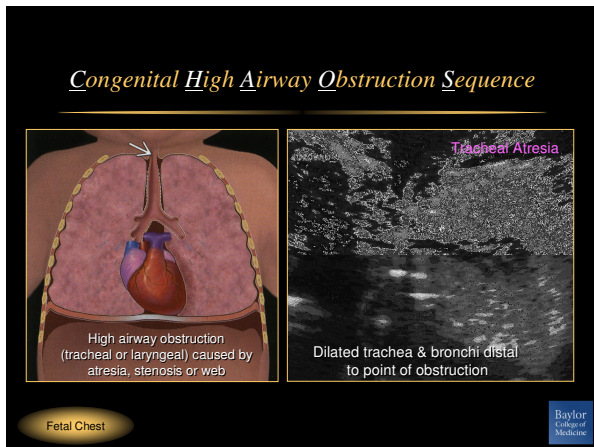
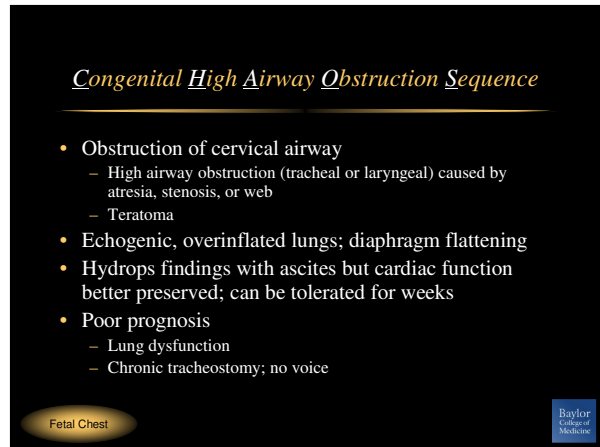
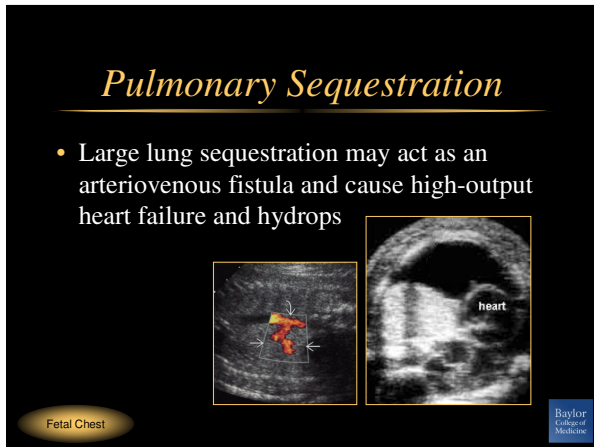
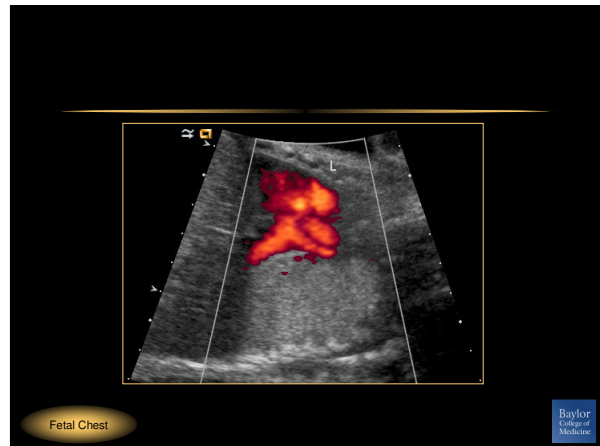
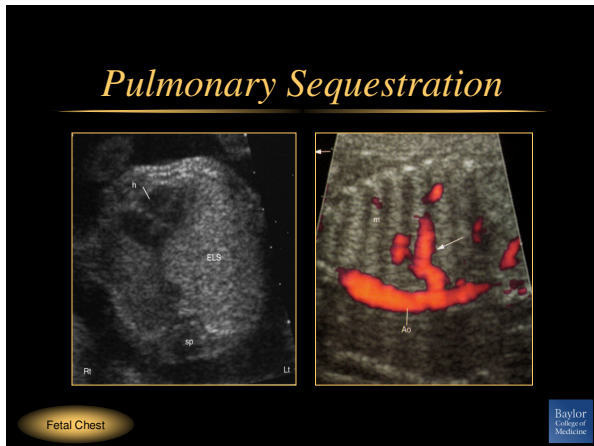
- Supplied by arteries arising from descending aorta



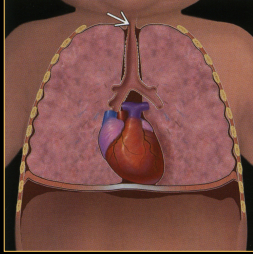
Systemic flow to sequestration

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### *Congenital High Airway Obstruction Sequence*



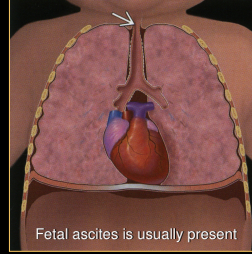
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Bilaterally enlarged echogenic lungs  
Heart appears small & ML in position



### *Congenital High Airway Obstruction Sequence*



Fetal Chest

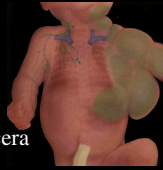


Fetal ascites is usually present



### *Cystic Lymphangioma*

- A localized collection of lymphatic vessels resulting in a nodule or mass
  - Cheek & parotid
  - Around the neck – 75%
  - Axilla & hemithorax – 20%
  - Retroperitoneum & abdominal viscera
  - Lambar region
  - Limbs



Fetal Chest



### *Cystic Lymphangioma*

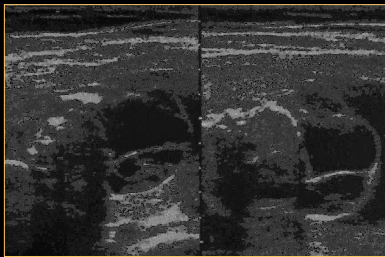
- Benign tumors of the lymphatic system
- Uni- or multilocular cystic masses



Fetal Chest



### *Cystic Hygromas*



Cystic hygromas can extend to the scalp, axilla or chest

Fetal Chest



### *Cystic Lymphangioma*

- Determination of the karyotype is recommended in all cases
- Serial sonograms
  - Assess the growth
  - Monitoring for the development of hydrops

Fetal Chest



## Pleural Effusions

- Fluid in the pleural cavity
- Unilateral or bilateral sonolucency in fetal thoracic cage
- Shifting mediastinum or 4 chambered cardiac apex
- Shrunken lung parenchyma & fluttering with cardiac rhythm



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## Pleural Effusions – Common Causes

- Hydrops fetalis (*Bilateral*)
  - Immune
  - Non-immune
  - Poor prognosis
- Chromosomal, most often T21
- Sporadic syndromes, as well as some associated anomalies
- Chylothorax (*Unilateral*)
  - Accumulation of lymphatic fluid
  - Malformation in the thoracic duct
    - Leakage from lower portion
      - Result in rt. Pleural effusion
    - Leakage from upper portion
      - Result in lt. Pleural effusion

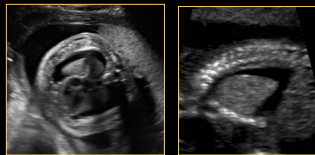
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## Pleural Effusions

### Outcome:

- Depends on degree of pulmonary hypoplasia underlying causes and chromosomal abnormalities
- If polyhydramnios develops, the prognosis is poor



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## Pleural Effusions “Protocol Advice”

- Look for hydrops
  - Skin edema
  - Ascites
  - Pericardial effusion
- Look carefully at fetal heart
  - Structural defects
  - Tachycardia
- Look for sign of fetal infection
  - Brain, liver, spleen calcifications
  - Intracranial hemorrhage
  - IUGR



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