Anomalies of the Fetal Thorax

Fetal Thorax

Upper half of trunk
Supported by spine and ribs
Separated from abdomen by diaphragm
Contains
- Heart
- Lungs
- Mediastinal structures

Thoracic Structure & Support

Ribs and Spine
Amniotic fluid

Both are necessary for adequate thoracic growth and pulmonary development

Ultrasound evaluation

- Are supporting structures normal?
- Are internal organs in normal position?
- Are internal organs normal?
- Any masses, cysts, or fluid?

Diaphragm

18 weeks

34 weeks
### Abnormal Thoracic Structure

Skeletal dysplasias with poor thoracic growth & support
- Thanatophoric dysplasia
- Osteogenesis imperfecta type 2
- Achondrogenesis

### Thoracic Wall Defect

- Isolated defect
- Ectopia cordis
- Pentalogy of Cantrell
- Amniotic Bands

### Ectopia Cordis

- Heart outside of thorax
- Heart often structurally abnormal
- Prognosis: poor
**Fetal Thorax**

Ultrasound evaluation

- Are supporting structures normal?
- Are internal organs in normal position?
- Are internal organs normal?
- Any masses, cysts, or fluid?

**Situs Inversus**

Complete (situs inversus totalis)

< 3% risk for cardiac anomalies

20% have Kartagener syndrome, bronchiectasis, nasal polyps

Partial situs inversus

40% have severe anomalies, cardiac, polysplenia, asplenia

**Complete Situs Inversus**

**Situs Ambiguous -- Heterotaxy**

Asplenia

- Bilateral right-sidedness
- Mortality 90 - 95%

Polysplenia

- Bilateral left-sidedness
- Mortality 80%

**Heterotaxy**

Ultrasound evaluation

- Are supporting structures normal?
- Are internal organs in normal position?
- Are internal organs normal?
- Any masses, cysts, or fluid?
### Pulmonary Dysplasias

**Congenital abnormalities of the lung**

- Normal lung parenchyma with systemic blood supply (classic sequestration)
- Abnormal lung parenchyma with systemic blood supply (CPAM + sequestration)
- Abnormal lung parenchyma with pulmonary blood supply (classic CPAM)

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### Pulmonary Dysplasias

**Congenital abnormalities of the lung**

- Assessment:
  - Lung parenchyma
    - Echogenicity
    - Are cysts present?
  - Arterial blood supply
    - Pulmonary or systemic
  - Venous return
    - Pulmonary veins to left atrium
    - or
    - Systemic veins to right atrium

### Congenital Pulmonary Airway Malformation

- **Type I** - Cyst or cysts > 2 cm
  - Prognosis: Good
- **Type II** - Multiple small cysts < 2 cm
  - Prognosis: Poor due to associated anomalies
- **Type III** - Large solid echogenic mass
  - Prognosis: Poor due to pulmonary hypoplasia

### Congenital Pulmonary Airway Malformation

- ~50% regress in utero
- Type I more often than II or III
- Poor prognosis if:
  - Hydrops
  - Associated anomalies
- Good prognosis otherwise
Kilgore CPAM type I

Descending aorta

Congenital Pulmonary Airway Malformation Type I

19 weeks

21 weeks

32 weeks

Evolving
Congenital Pulmonary Airway Malformation Type I

Mann CPAM II

Congenital Pulmonary Airway Malformation Type II

Pulmonary arterial supply & venous return

Garip CPAM II

18 weeks

27 weeks

30 weeks

Evolving
Congenital Pulmonary Airway Malformation Type II

33 weeks

Evolving
Congenital Pulmonary Airway Malformation Type II
Mixed Lesion: CPAM Type II & Sequestration
Abnormal lung & systemic blood supply

Mixed Lesion: CPAM Type II & Sequestration
Abnormal lung & systemic blood supply

Mixed Lesion: CPAM Type II & Sequestration
Abnormal lung & systemic blood supply

Systemic arterial supply

Pulmonary venous return

Classic
Pulmonary Sequestration
Segment of lung with
Systemic arterial blood supply
No tracheobronchial communication
Intrapulmonary or extralobar
(within lung pleura or its own pleura)
Above or below the diaphragm
Left lower lobe posterior most common
(90%)
May decrease in size during gestation
Pulmonary (25%) or systemic (75%)
venous drainage
Sequestration & Congenital Pulmonary Airway Malformation

Prenatal

Sequestration & Congenital Pulmonary Airway Malformation

Extends below diaphragm

Tracheal Atresia

Ultrasound findings
- Enlarged echogenic lungs
- Polyhydramnios
- Hydrops

Fetal Thorax

Ultrasound evaluation
- Are supporting structures normal?
- Are internal organs in normal position?
- Are internal organs normal?
- Any masses, cysts, or fluid?
**Diaphragmatic Hernia**

Defect at foramen of Bochdalek
- Left — Stomach & bowel → thorax
- Right — Liver → thorax

Defect at foramen of Morgagni (rare)
- Anterior midline — Stomach, bowel & liver → thorax

Ultrasound findings associated with worse prognosis:
- Polyhydramnios (35%)
- Associated anomalies
- Early diagnosis
- Large volume herniated
- Mediastinal shift
- Right diaphragmatic hernia

Left diaphragmatic hernia with liver in thorax
- 37 weeks
EXIT to ECMO for Left Diaphragmatic Hernias

Improved survival to ~ 95%
Mortality often due to other anomalies particularly cardiac anomalies

Left diaphragmatic hernia – EXIT to ECMO

Right diaphragmatic hernia
Paradoxical motion with breathing movements

Left diaphragmatic hernia – EXIT to ECMO
Left diaphragmatic hernia – EXIT to ECMO

Left diaphragmatic hernia – Prenatal

Left diaphragmatic hernia – EXIT to ECMO
Left diaphragmatic hernia – EXIT to ECMO

Right diaphragmatic Hernia
EXIT to ECMO
Right diaphragmatic hernia delivered by EXIT to ECMO

Internal jugular vein Catheter to right atrium
Internal carotid artery Catheter to ascending aorta

Mediastinal Teratoma

Ultrasound findings
Complex intrathoracic mass
Solid, cystic, +/- calcifications
Cardiac displacement
+/- Hydrops

Neck teratoma extending into mediastinum

Neck teratoma extending into mediastinum

Neck teratoma extending into mediastinum
EXIT procedure & resuscitation

Right lung

Right lung
Neck teratoma extending into mediastinum

Pleural Effusions
Unilateral
   Early hydrops
   Chylous
Bilateral
   Hydrops
Small – May resolve
Large
   Hydrops
   Pulmonary hypoplasia

Pleural Effusions
Prognosis depends on
   Degree of pulmonary hypoplasia
   Underlying cause
   Presence of hydrops
   Aneuploidy ~ 12% (when isolated)
Outcome
   (~10% IUFD)
   ~20% Neonatal death
   syndromes, pulm. hypoplasia
   ~30% Survive with morbidity
   ~50% Normal

Bilateral pleural effusions 32 weeks
Trisomy 21

Pleural effusion & hydrops
Treated with thoracentesis

Pleural Effusions
Treatments options
   Thoracentesis
      Reverse the hydrops
      Allow for lung expansion
   Thoraco-amniotic shunt
      If reaccumulation
      after thoracentesis
**Thoracoamniotic Shunt**

Place pigtail catheter in pleural space and amniotic cavity
For continuous drainage of pleural effusions
Allow lungs to expand
Improve blood return to the heart to decrease hydrops

24 weeks

After drainage

Recurrent large pleural effusion – 27 weeks

Thoracoamniotic shunt 27 weeks
Thoracoamniotic shunt
27 weeks

After procedure

Thoracoamniotic shunt
At birth