Fetal Chest
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Organs

- Heart
- Lungs
- Pleura
- Oesophagus
- Blood vessels
- Bony cavity
Normal lung anatomy is difficult to quantify at 18-20 weeks.

Normal lungs produce midrange echoes on U/S.

Note relative proportions of heart:lungs ie 1/3:2/3. This remains the same since lungs and heart grow at similar rates.

Gross lung anomalies eg pulmonary hamatoma can be seen at early gestation.
Measurements at this gestation are unrewarding.

Marked asymmetry and flattening of the fetal chest may indicate various skeletal dysplasias.

FBM are mainly intermittent

Fluid flux down the trachea may be seen by colour Doppler
Abnormalities

- Diaphragmatic hernia
- Congenital cystic adenomatous malformations
- Broncho-pulmonary sequestration
- Pleural effusions
- Pulmonary hypoplasia
- Congenital hydro/chylo thorax
- Pulmonary lymphangiectasia
- Chondrodysplasia
- Bronchogenic cysts and other tumours
Herniation of the abdominal viscera into the thorax at about 10-12 weeks, when the intestines return to the abdominal cavity from the umbilical cord. However, intrathoracic herniation of viscera may be delayed until the second or third trimester of pregnancy.
DIAPHRAGMATIC HERNIA

Displaced Heart
Stomach
Liver
Area of Diaphragmatic Defect
DIAPHRAGMATIC HERNIA

- Incidence- 0.29 per 1,000 live births
- Findings on U/S
  - Fluid filled thoracic mass caused by stomach or bowel
  - Solid organs such as liver and spleen in the chest
  - Abnormal cardiac position
  - Decreased abd circumfrence
  - Viscera can slide btn abd and chest.
Heart Displaced Toward Right Side

Abnormal Cardiac Position Due to Intra-thoracic Bowel

Right

Left

Liver

Bowel

Spine
DIAPHRAGMATIC HERNIA

- Differential diagnosis:
  - Eventration
  - CCAM
  - Bronchopulmonary sequestration
  - Tumour
DIAPHRAGMATIC HERNIA

- ¾ involve the left side due to foramen of Bochdalec
- Herniation may cause pulmonary hypoplasia
- May be associated with hydramnios
- Colour Doppler has a role in diagnosis since the liver and lungs have the same texture
- Absence of gastric bubble in upper abdomen may be a sign of DH
Outcome depends on presence of additional chromosomal and structural abn.

Reported survival rate of 42.5%

Prognosis depends on:
- Fetal hydrops
- Degree of pulmonary hypoplasia
- Presence and severity of ass. abn.
Endoscopic occlusion of the fetal trachea is now carried out in human fetuses with diaphragmatic hernia.
Fetal pleural effusions, which may be unilateral or bilateral, may be an isolated finding or they occur in association with generalized edema and ascites.
Chylothorax

Definition
Chylothorax is an accumulation of chyle in the pleural cavity.

Incidence
Chylothorax is a common cause of pleural effusion during the first days of neonatal life. Prevalence 1:10,000 deliveries. Male to Female ratio is 2:1.

Etiology
Accumulation of lymph within the pleural cavity can result from overproduction or impaired re absorption of lymph. The latter could be due to an obstruction.
Chylothorax occurs usually as a unilateral pleural effusion involving the right side of the lung in most instances. In rare cases, pleural effusions can be bilateral.

Unilateral pleural effusion can also shift the mediastinum, impair venous return, and lead to congestive heart failure and hydrops.
Chylothorax may be associated with trisomy 21.

Anomalies reported in association with chylothorax include:
- congenital pulmonary lymphangiectasis,
- tracheoesophageal fistula,
- extralobar lung sequestration,
- and a multiple malformation complex (anemia, tracheoesophageal fistula).
The differential diagnosis of congenital chylothorax is problematic:

- Isolated pleural effusions
- or non immune hydrops.

NB: Biochemical or cytological examination of the pleural fluid can permit a differential diagnosis between the effusion seen in congenital chylothorax and that seen in other causes of non immune hydrops.
Figure 5-1. Transverse scan of a patient with congenital chylothorax. A predominantly left chylothorax (*) deviates the heart (H) toward the right. L, Lung; Sp, spine.

Figure 5-2. Longitudinal scan of the fetus shown in Figure 5-1. The arrow points to the presence of ascites. *, chylothorax; LVR, liver.
Figure 5-3. Transverse scan of the chest in a fetus with bilateral chylothorax (C). H, heart.
Extremely difficult to provide prognostic figures for congenital chylothorax diagnosed in utero because of the limited experience with this condition and the uncertainty surrounding its diagnosis.
1. When a diagnosis is made before viability, the option of TOP can be offered.

2. Karyotyping is indicated, since this condition has been associated with chromosomal anomalies such as trisomy 21.

3. After viability, the management depends on the gestational age and the development of signs of hydrops or mediastinal shift.

4. In term infants, a thoracentesis should be considered before delivery to avoid respiratory failure due to a large pleural effusion.
**Congenital Cystic Adenomatoid Malformation of the Lung**

**Synonym**
Adenomatoid hamartoma.

**Definition**
Benign tumour of the lung characterized by disordered overgrowth of terminal bronchioles.
CCAML is a rare malformation of the lung
~200 cases have been reported in the literature to date.
There is no sex predilection.
Incidence of 1 per 10,000 births
U/S features

- Intrathoracic lung mass
- Abnormal cardiac position
- Hydramnois and hydrops
Classification of CCAML into three subtypes according to the size of the cysts:

- type I has large cysts,
- type II has multiple small cysts of less than 1.2 cm in diameter
- type III consists of a noncystic lesion producing mediastinal shift (Fig. 5-4).

The worst prognosis is seen in type III lesions.
Figure 5-4. Classification of congenital cystic adenomatoid malformation. Type I is composed of a small number of large cysts with thick smooth muscle and elastic tissue walls. Relatively normal alveoli are seen between and adjacent to these cysts. Mucous glands may be present. Type II contains numerous smaller cysts (< 1 cm in diameter), with a thin muscular coat beneath the ciliated columnar epithelium. The area between the cysts is occupied by large alveoluslike structures. The lesion blends with the normal parenchyma. Type III occupies the entire lobe or lobes and is composed of regularly spaced bronchiolelike structures separated by masses of cuboidal epithelium-lined alveoluslike structures. (Reproduced with permission from Stocker et al.: Hum Pathol 8:155, 1977.)
Figure 5-5. Transverse scan of the chest of a fetus with congenital cystic adenomatoid malformation of the lung type 1. The heart is displaced toward the right hemithorax. Sp, spine; LV, left ventricle; RV, right ventricle; C, cysts; arrow heads point to pulmonary parenchyma.
Associated Anomalies

- bilateral renal agenesis,
- renal dysplasia, truncus arteriosus, tetralogy of Fallot,
- hydrocephalus, jejunal atresia,
- diaphragmatic hernia,
- deformity of clavicle and spine, and
- sirenomelia.
CYSTIC ADENOMATOID MALFORMATION (CAM)

<table>
<thead>
<tr>
<th>large cysts</th>
<th>small cysts</th>
<th>microcystic</th>
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<tbody>
<tr>
<td><img src="image1.png" alt="Large Cysts" /></td>
<td><img src="image2.png" alt="Small Cysts" /></td>
<td><img src="image3.png" alt="Microcystic" /></td>
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Figure 5-6. Longitudinal scan of the same fetus shown in Figure 5-5. Multiple large cysts (C) are visualized. Arrow points to pulmonary parenchyma. LVR, liver.

Figure 5-7. Longitudinal scan of an infant with congenital cystic malformation of the lung of the microcystic variety (type III). H, heart; LVR, liver; B, bladder; St, stomach; arrowheads point to the microcystic lesion.
Differential diagnosis

Diaphragmatic hernia
Bronchopulmonary sequestration
Eventration
Tumour
CAM: Prognosis

Depends on:
- presence of hydrops
- karyotype
- variety
- pulmonary hypoplasia
If this diagnosis is made before viability, TOP should be offered. Management and prognosis depend on the presence of associated hydrops.
Treatment

Lobectomy rather than segmentectomy
Lung Sequestration

**Synonyms**
Bronchopulmonary sequestration and accessory lung.

**Definition**
Congenital anomaly in which a mass of pulmonary parenchyma is separated from the normal lung.

*It usually does not communicate with an airway and receives its blood supply from the systemic circulation.*
Pathology

Classified into two types: intralobar and extralobar.

- In the intralobar variety, the sequestered lung and the normal lung share a common pleura.
- In the extralobar variety (most common) the sequestered lung is covered by its own visceral pleura in a way similar to an accessory lobe.
Figure 5-8. Autopsy specimen from a case of extralobar pulmonary sequestration. RL, right lung; LL, left lung; SL, sequestered lung. (Reproduced with permission from Romero et al: J Ultrasound Med 1:131, 1982.)
In intralobar lung sequestration, either side is involved with similar frequency. The lower lobes are the most commonly affected (98 percent).
Incidence

Rare anomaly without familial predispositions
In the extralobar variety, male to female 3:1.
Figure 5-9. The blood supply to the sequestered lung (SL) is derived from the systemic circulation. The arrow points to the vascular supply to an extralobar sequestered lung.
Associated Anomalies

tracheoesophageal fistula, esophageal duplications, neurenteric cysts, esophageal diverticulum, esophageal cysts, and bronchogenic cysts.
Diagnosis

The sequestered lung appears as an echogenic, intra-thoracic, or intraabdominal mass (Figs. 5-10, 5-11) usually with hydrops.
Figure 5-10. Longitudinal scan of an infant with extralobar pulmonary sequestration and nonimmune hydrops. Hydrothorax (HT) is present. SL, sequestered lung. (Reproduced with permission from Romero et al.: J Ultrasound Med 1:131, 1982.)

Figure 5-11. Transverse scan of the thorax demonstrates bilateral hydrothorax (ht), the right lung (L), and the sequestered lung (SL). (Reproduced with permission from Romero et al.: J Ultrasound Med 1:131, 1982.)
Only 1 of the 4 patients with lung sequestration detected antenatally survived. The other 3 had nonimmune hydrops and intrathoracic sequestration.
Before viability, the option of pregnancy termination should be offered.
After viability, prognosis is probably related to the development of hydrops.

*Delivery in a tertiary center is recommended.*
*Immediate respiratory support may be required.*
Bronchogenic Cyst

Definition
A bronchogenic cyst is a cystic structure lined by bronchial epithelium.

Incidence
The incidence of bronchogenic cysts is unknown, since a large number of them are asymptomatic. They are extremely rare in the neonatal period.
The other bronchopulmonary foregut malformations include:
- tracheoesophageal fistula,
- esophageal diverticulum,
- esophageal cysts,
- and lung sequestration.

Vertebral abnormalities (hemivertebrae) are often associated with bronchogenic cysts of mediastinal origin.
Figure 5-12. A fetus with two bronchogenic cysts. One is located in the apex of the right lung (B1), and the other (B2) is located in the opposite lung.
Prognosis

The treatment is surgical extirpation.

Large cysts may require lobectomy and even pneumonectomy.