Obstetrics Content Outline

Obstetrics - Fetal Abnormalities

Effective February 2007

10 – 16%

Abdominal wall

Sonographic Evaluation of the Fetal Abdominal Wall

• image the cord insertion site and the fetal anterior abdominal wall
  – evaluate for the presence of defects
• Questions to ask if defect is found
  – Is a limiting membrane present?
  – What is the relation of the umbilical cord to the defect?
  – Which organs are eviscerated?
  – Is the bowel normal in appearance?
  – Are other fetal malformations evident?

Abdominal wall

Anterior Abdominal Wall Abnormalities

• common abdominal wall defects:
  – Omphalocele
  – umbilical hernia (a form of omphalocele)
  – gastroschisis.
• Rare defects include
  – ectopia cordis
  – pentalogy of Cantrell
  – limb–body wall complex
  – amnion rupture sequence
  – bladder and cloacal extrophy.

Abdominal wall

Omphalocele

• omphalocele - a midline defect of
  – abdominal muscles, fascia, and skin - results in herniation of intraabdominal structures into the base of the umbilical cord.
  – This herniation is covered by a membrane
• 8th to 12th weeks fetal bowel normally migrates into the umbilical cord
• normally return by the 12th week of pregnancy.

Abdominal wall

Omphalocele

• characterized as two types:
  – Those that contain liver within the sac
  – Those that contain a variable amount of bowel, but no liver - a higher risk for chromosomal abnormalities

Abdominal wall

Distinguishing an omphalocele from gastroschisis

– Look for the presence of a membrane - gastroschisis does not have one.
– Look at the umbilical cord; the cord goes through the omphalocele—it’s usually is to the right of the gastroschisis
– Determine which organs are eviscerated
– Look for other anomalies; omphalocele has a high percentage of chromosome abnormalities.
Abdominal wall
Sonographic signs of an omphalocele
- Central abdominal wall defect
  - evisceration of the bowel or a combination of liver and bowel into the base of the umbilical cord
- Membrane composed of peritoneum and amnion forms the omphalocele sac
- Ascites may coexist with omphalocele.
- Hydramnios found in one third of fetuses

Abdominal wall
Gastroschisis
- opening in the layers of the abdominal wall with evisceration of the bowel
  - infrequently, the stomach and genitourinary organs.
- the defect is positioned to the right of the umbilical cord.

Abdominal wall
Gastroschisis
- Small bowel is always found in the herniation
- AFP levels are significantly higher
- sonographer may be able to detect after 12 weeks of gestation.

Abdominal wall
Amniotic Band Syndrome
- the rupture of the amnion
  - leads to entrapment or entanglement of the fetal parts by the "sticky" chorion.
- Late entrapment leads to amputations or limb restrictions

Abdominal wall
Amniotic Band Syndrome
- Early entrapment by the bands may lead to severe craniofacial defects and internal malformations.
- Associated anomalies include
  - anomalies of the limbs, cranium, face, thorax, spine, abdominal wall.
**Abdominal wall**

**Beckwith-Wiedemann Syndrome**
- a group of disorders
  - Omphalocele
  - Macroglossia
  - visceromegaly.

**Pentalogy of Cantrell, Ectopic Cords, and Cleft Sternum**
- association of two major defects
  - an omphalocele and an ectopic heart
- The three other anomalies are the result of these defects
  - defect in the lower sternum
  - anterior diaphragm
  - diaphragmatic pericardium.

**On ultrasound, the heart may be seen to lie outside the normal thoracic cavity or bulge through the defective sternum.**

**Abdominal wall**

**Limb–Body Wall Complex**
- occurs with the fusion of the amnion and chorion
  - the amnion does not cover the umbilical cord normally
- associated with large cranial defects
  - exencephaly or encephalocele
  - facial cleft
  - body-wall defect - thorax, abdomen
  - limb defects
  - scoliosis and various internal malformations.
micronathia

Gastroschisis

twins

Thoracic

• normal shape
  – symmetrically bell shaped
  – ribs forming the lateral margins
  – clavicles the upper margins
  – diaphragm the lower margin
• thorax is normally slightly smaller than the abdominal cavity
• Chest circumference
  – made in the transverse plane at the level of the four-chamber view of the heart
**Thoracic**
- central portion of the thorax is occupied by the mediastinum
- the majority of the heart is positioned in the midline and left chest
  - apex should be directed toward the spleen
  - base lies horizontal to the diaphragm
- abnormal heart position may indicate
  - presence of a chest mass, pleural effusion, or cardiac malformation.

**Texture**
- fetal lungs appear on sonography as homogeneous and moderately echogenic
- lungs are similar to or slightly less echogenic than the liver in Early in gestation
- latter pulmonary echogenicity is increased relative to the liver.
- Ultrasound cannot assess lung maturity.Ψ

**Abnormalities of the Thoracic Cavity**
- lungs
  - separate from the heart and above the diaphragm
  - Lesions may be cystic, solid, or complex
  - Pleural effusions are commonly found in conjunction with lung masses
  - A mass may have detrimental effects on lung development
    - pulmonary hypoplasia

**Pulmonary Hypoplasia**
- reduction in lung volume resulting in small, inadequately developed lungs
- commonly results from
  - prolonged oligohydramnios
  - secondary to a small thoracic cavity (ex. chromosomal abnormalities)
- Poor prognosis - 80% die after birth.

**Additional causes**
- diaphragmatic hernia Ψ
- extreme reduction in amniotic fluid volume.
- Kidney abnormalities
- severe intrauterine growth retardation
- early rupture of the membranes
- pleural effusion
- cystic adenomatoid malformation of the lung
- bronchopulmonary sequestration

**Pleural Effusion**
- accumulations of fluid within the pleural cavity
- common reason
  - chylothorax right-sided unilateral collection of fluid secondary to a malformed thoracic duct.
Pleural Effusion

- immune (e.g., Rh disease)
- congestive heart failure.
- chromosomal abnormalities (e.g., trisomy 21)
- Compression of lung parenchyma may cause pulmonary hypoplasia

Solid Lung Masses

Bronchopulmonary Sequestration

- extra pulmonary tissue is present within the pleural lung sac
- This extra lung tissue is nonfunctional
- receives its blood supply from systemic circulation.
- prognosis for intralobar sequestration is very favorable
- extralobar sequestration carries a poor prognosis

Congenital Cystic Adenomatoid Malformation

CCAM

- abnormality in the formation of the bronchial tree
  Three forms
  - type I one or several large cysts replace normal lung tissue
  - Type II lesions consist of multiple small cysts
  - Type III bulky, large, noncystic lesions appearing as echo-dense masses of the entire lung lobe.

Congenital Diaphragmatic Hernia

- opening in the pleuroperitoneal membrane
- most common type (>90%) posteriorly and laterally in the diaphragm (through foramen of Bochdalek).

Abnormalities of the Diaphragm

- Normal - appear as a curvilinear structure coursing anteriorly to posteriorly
- Fetal stomach and liver should be identified caudal to the diaphragm
- the lungs and heart positioned cephalad.
Congenital Diaphragmatic Hernia

- organs shift the heart and mediastinal structures to the right side of the chest
- stomach is in the chest near the heart, instead of below the diaphragm.