Fetal Ventral Wall Defects
US/MR Evaluation

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I have no commercial disclosures
Ventral Wall Anomalies

Gastroschisis
Omphalocele
Bladder Extrophy
Ectopia Cordis
Limb Body Wall Complex
Embryology

5th week diverticula

7-11 weeks bowel retracts into abdomen

6th week growth and rotation of the midgut
Embryology

**Omphalocele**
- Failure of migration of mesodermal body folds
- Enlarged umbilical ring
- Umbilical membrane is the amnion, Wharton jelly and peritoneum

**Gastroschisis**
- Rupture of the abdominal wall due to:
  - Abnormal involution of the right umbilical vein and omphalomesenteric artery
  - Mensenchymal defect
Umbilical Cord Insertion

12 week of GA
Bowel Herniation

20 weeks GA
Omphalocele

• Incidence 1/4000

• Survival 80-90% if no associated anomalies

• Higher incidence of chromosomal anomalies when only bowel is herniated

• Higher incidence of prematurity
Associated Anomalies

- **Chromosomal anomalies 50%**
  - Trisomy 21, 13 and 18

- **Structural anomalies 60%**
  - Cardiac anomalies
    - Septal defects and tetralogy of Fallot
  - OEIS
  - Beckwith Wiedemann
  - Pentalogy of Cantrell
Edema of the Umbilical Cord

Postnatal diagnosis of Beckwith Wiedemann
22 weeks of GA
Giant Omphalocele

**Definition:**
- Exceeds 5 cm and contains liver in the herniated sac
- More of 75% of liver is herniated

**Complications:**
- Pulmonary hypoplasia due to thoracic deformity
- 20% of demise in neonates without associated anomalies
Giant Omphalocele - Outcome
Perforated Omphalocele
Gastroschisis
Cord Insertion
Gastroschisis

- Incidence: 1/5000 ~ increasing*
- Adolescent mothers 6 times more frequent
- Associated environmental effect
- No chromosomal anomalies – debate

Liver Herniation

- Approximately 6%
- Associated with other organs out
- Higher comorbidities (lung hypoplasia)
- Survival
  - 97% without liver herniation
  - 43% with liver herniation
Obstruction and Bowel Wall Thickening
Toxicity of Amniotic Fluid

- Increased urea and creatinine
- Decreased $\text{Na}^+$ and osmolarity
- Meconium, GI content products
- Inflammatory process: Il-6, ferritin
Management

• Steroids
• Amnioexchange

• Doppler of SMA correlates with length of stay in NICU*

Short Gut Syndrome in Utero
Closed Gastrochisis

32 weeks of GA
Postnatal Complications

• Dehydration, Infection, Bowel Ischemia
• Atresia (10%)
• Bowel obstruction, perforation
• Edema and poor peristalsis, malabsorption
• IUGR

• Survival
  – 1943 ~10 %
  – 2013 ~ 90%
Bladder Exstrophy

- Incidence: 1/30,000
- Failure of anterior body wall closure
- Spectrum of anomalies from epispadias to cloacal exstrophy

- Ultrasound
  - Absent bladder
- MRI
  - Evaluation of associated anomalies
OEIS
Omphalocele – Exstrophy – Imperforate Anus – Spinal Defect

• Incidence: 1/200 000 – 400 000

• Underlying cause is unknown

• Cloacal exstrophy when
  – Two hemibladders
  – Rudimental colon and prolapse of terminal ileum
Ectopia Cordis and Omphalocele
28 weeks of GA

After few days
Limb – Body – Wall Complex
19 weeks of GA
Limb – Body – Wall Complex

- Amelia, visceral, limb, craniofacial abnormalities
- Non fusion of amnion and chorion
- Incidence ~1/40 000
- DD:
  - Amniotic band syndrome
  - Pentalogy of Cantrell
  - Cloacal extrophy
3D Imaging

Courtesy Prof Jaramillo
Conclusions

• High resolution Ultrasound and MRI have improved the prenatal diagnosis of ventral wall anomalies

• Accurate diagnosis is essential for:
  – Parental counseling
  – Planning of surgical and neonatal management
Other Wall Defects

Berlin wall 1989