Fetal Ventral Wall Defects US/MR Evaluation

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Ventral Wall Anomalies

Outline

Gastroschisis

Omphalocele

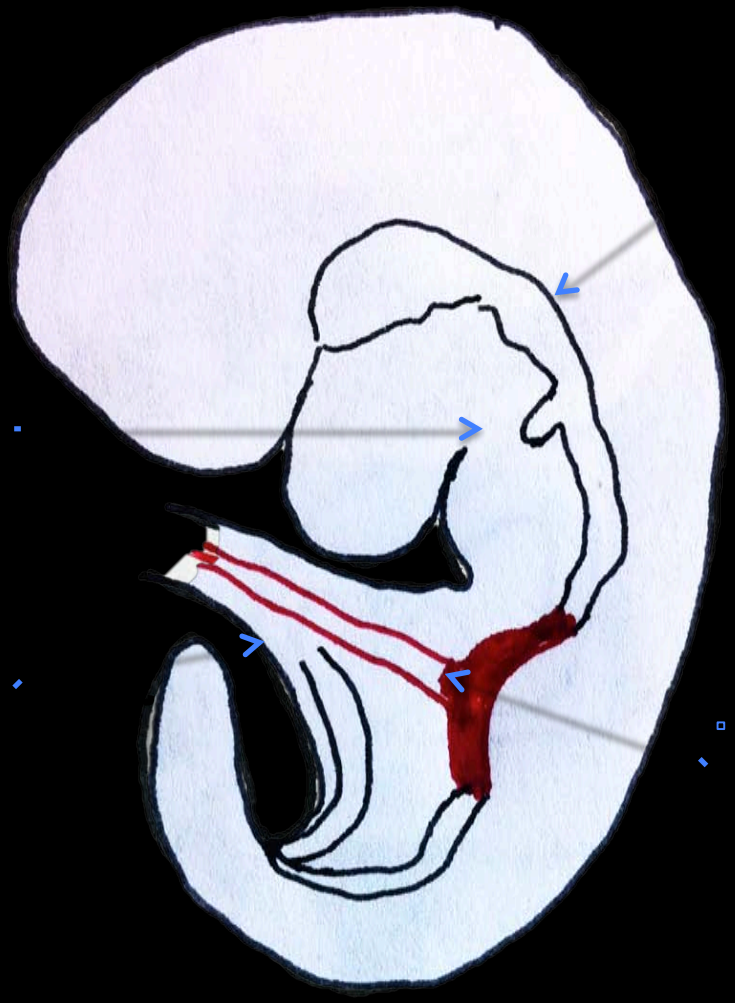
Bladder Extrophy

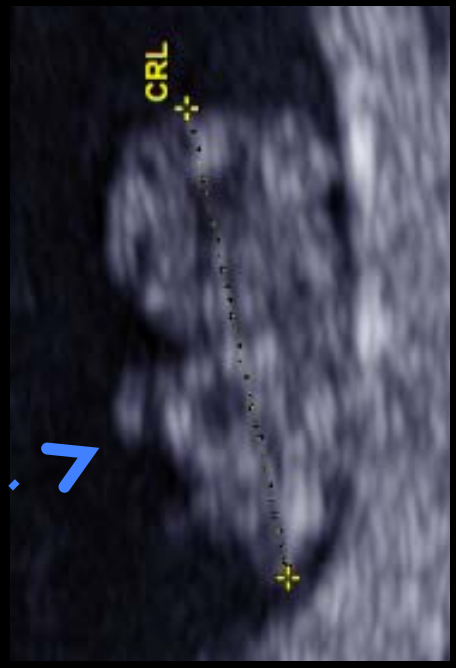
Ectopia Cordis

BWC

Embryology

- 5th week
  diverticula
- 7-11 weeks
  bowel retracts
  into abdomen





- 6th week
  growth and rotation
  of the midgut

Embryology

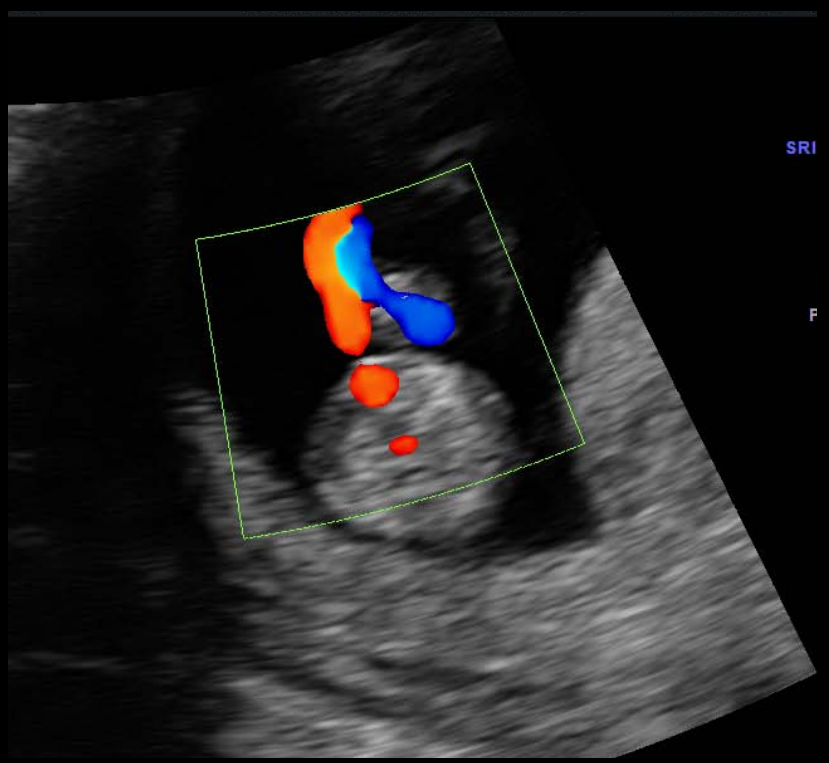
Omphalocele

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

Gastroschisis

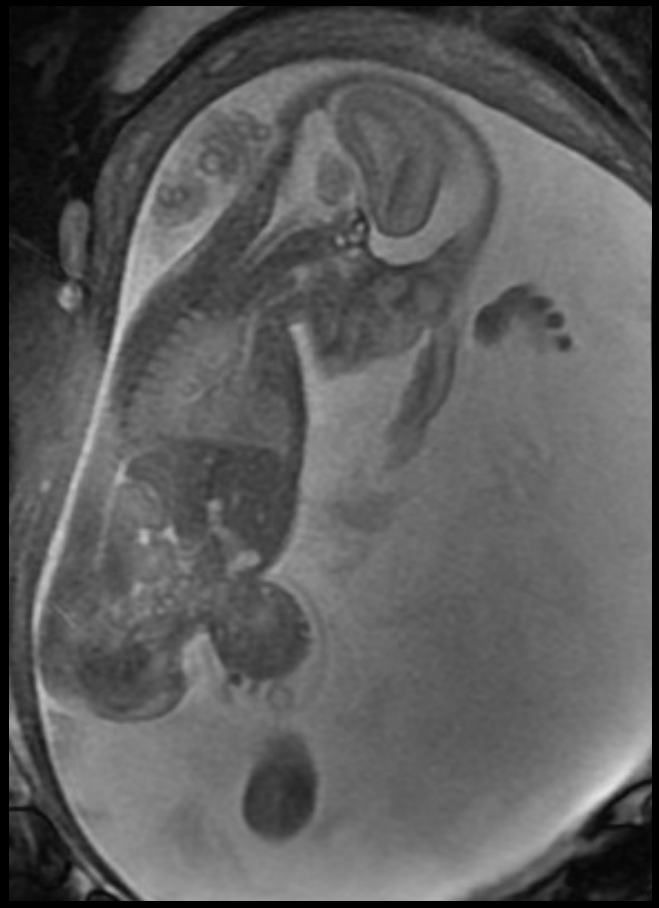
- Rupture of the abdominal wall due to:
  - Abnormal involution of the right umbilical vein and omphalomesenteric artery
  - Mensenchymal defect

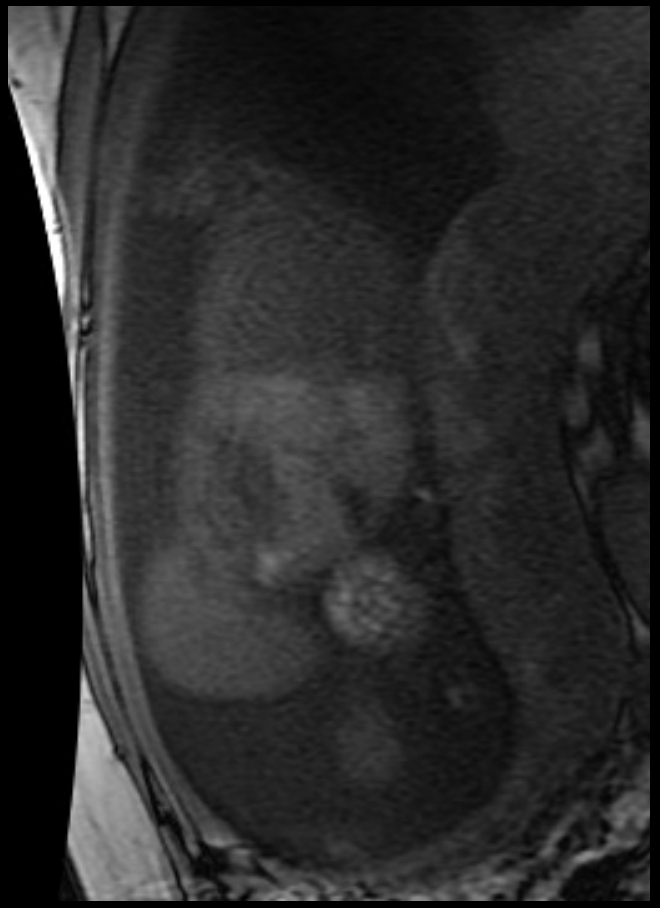
Umbilical Cord Insertion



12 week of GA

Bowel Herniation





20 weeks GA

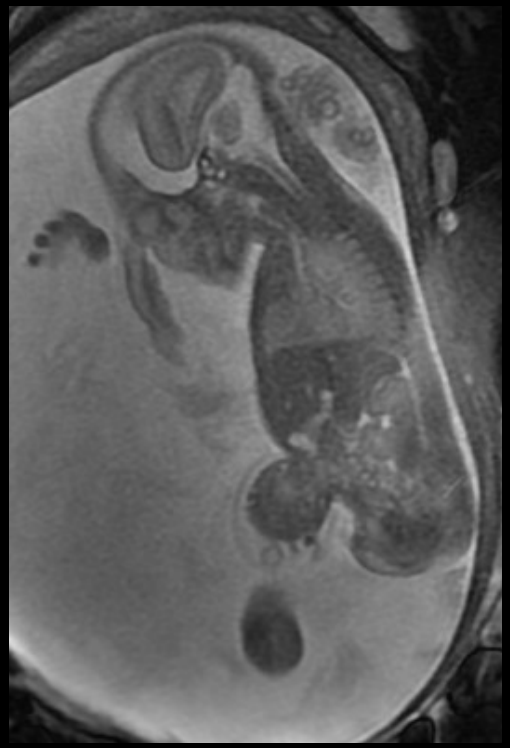
Omphalocele

- Incidence 1/4000
- Survival 80-90% if no associated anomalies
- Higher incidence of chromosomal anomalies when only bowel is herniated
- Higer incidence of prematurity

Associated Anomalies

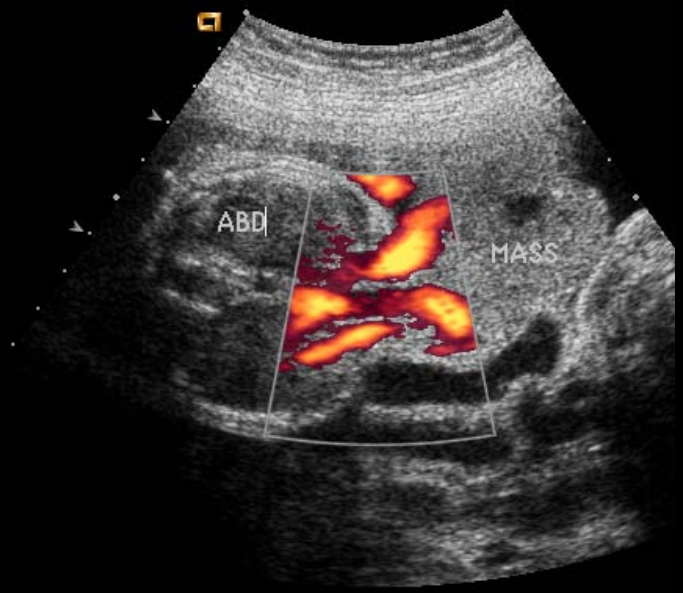
- *Chromosomal anomalies 50%*
  - Trisomy 21, 13 and 18
- *Structural anomalies 60%*
  - Cardiac anomalies
    - Septal defects and tetralogy of Fallot
  - OEIS
  - Beckwith Wiedemann
  - Pentalogy of Cantrell

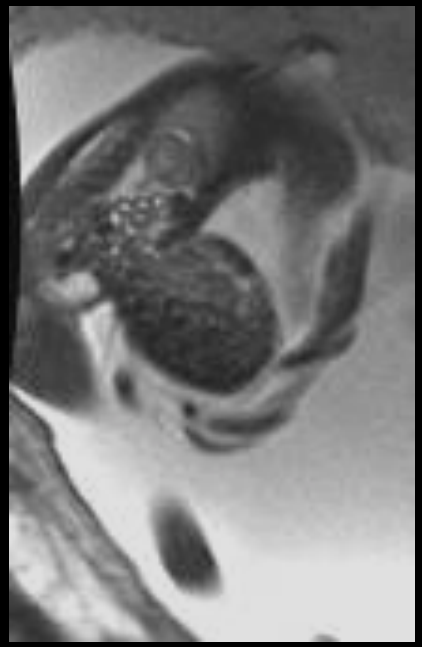
Edema of the Umbilical Cord

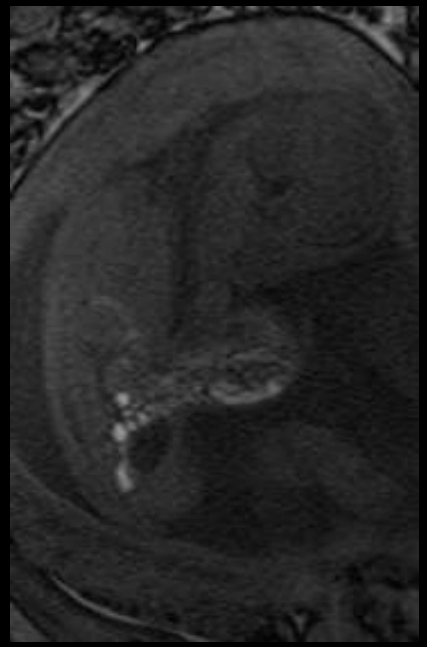


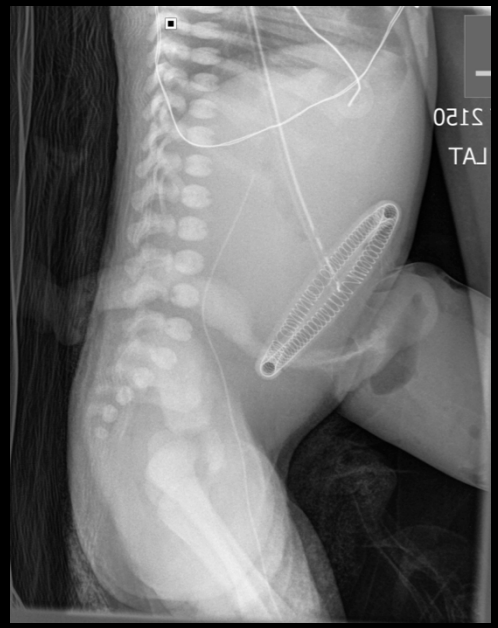
Postnatal diagnosis of Beckwith Wiedemann

22 weeks of GA









Giant Omphalocele

Definition:

- Exceeds 5cm and contains liver in the herniated sac

or

- More of 75% of liver is herniated

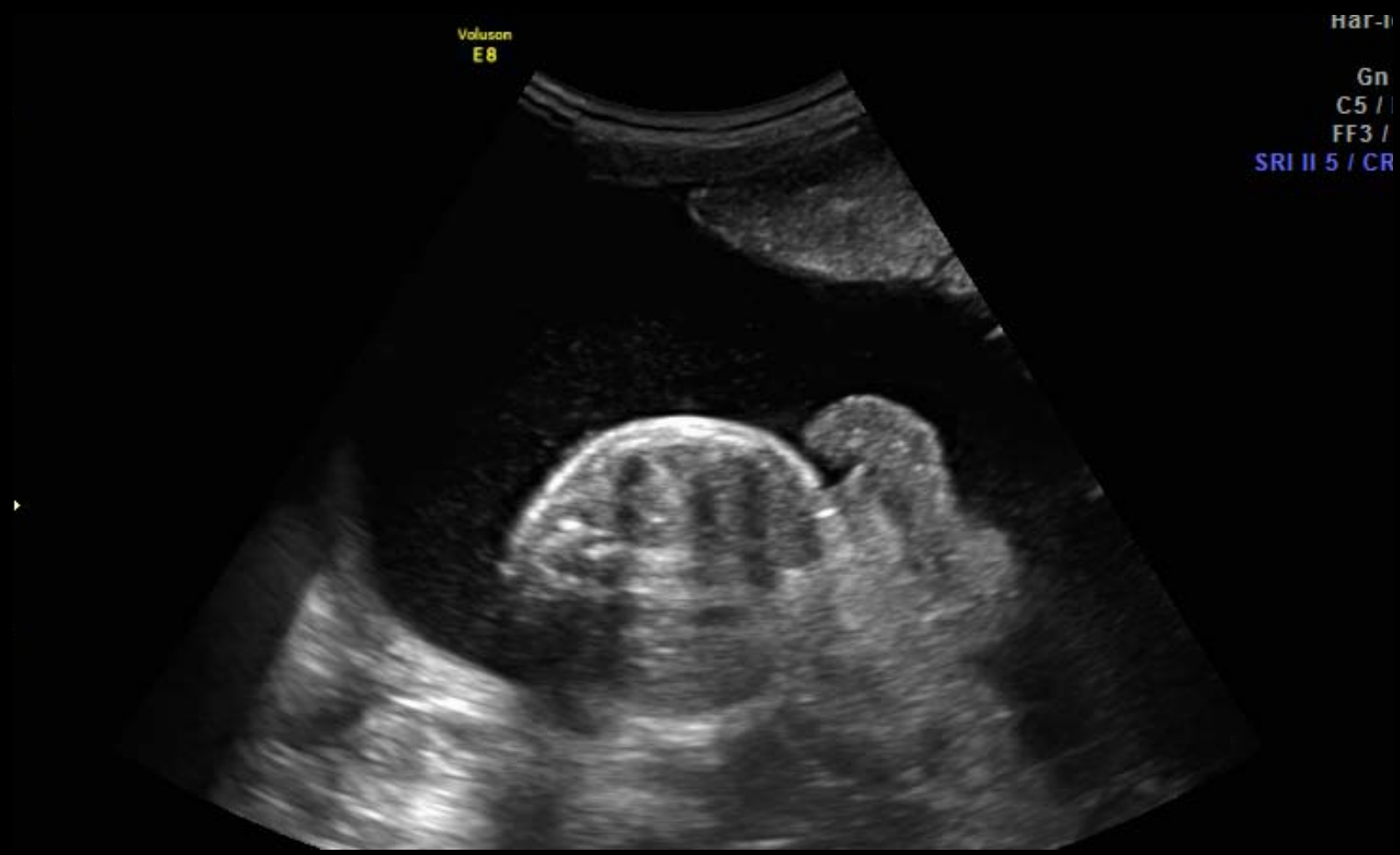
Complications:

- Pulmonary hypoplasia due to thoracic deformity
- 20% of demise in neonates without associated anomalies

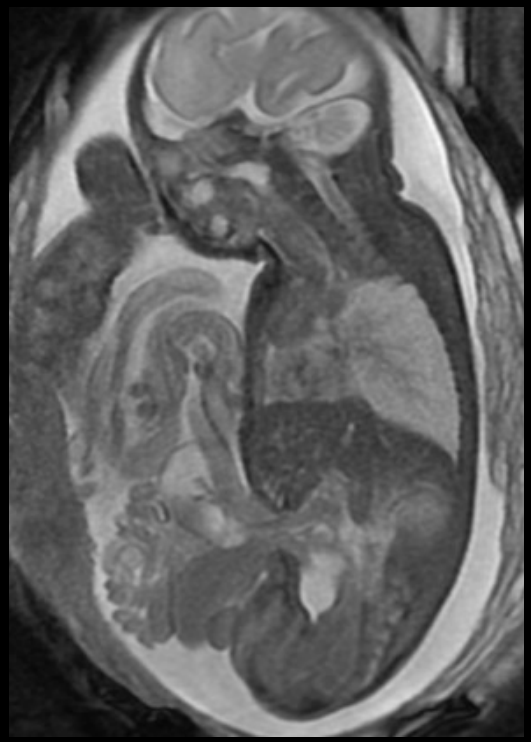
Giant Omphalocele - Outcome


Voluson
E8
95
Gn 3
C5 / M5
FF3 / E2
SRI II 5 / CRI 4

Perforated Omphalocele



Gastroschisis



Cord Insertion

Gastroschisis

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

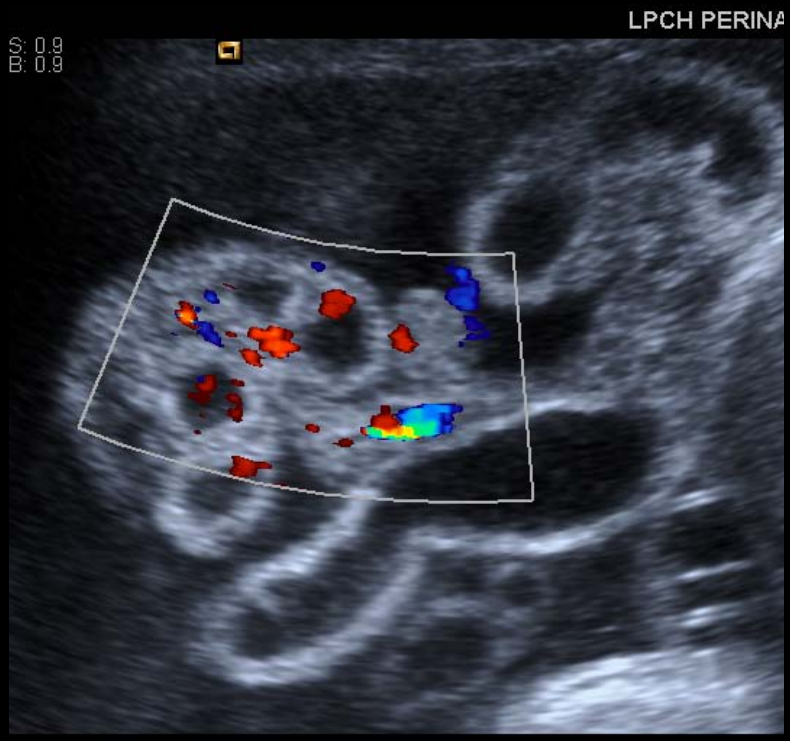
*Curry et al. Br J Obstet Gynaecol, Nov (107) (2000) 1339–1346

Liver Herniation

- Approximately 6% *
- Associated with other organs out
- Higher comorbidities (lung hypoplasia)
- Survival
  - 97% without liver herniation
  - 43% with liver herniation

**Mc Clellan et al. J Ped Surg 2011 (46) 2115-2118*

Obstruction and Bowel Wall Thickening



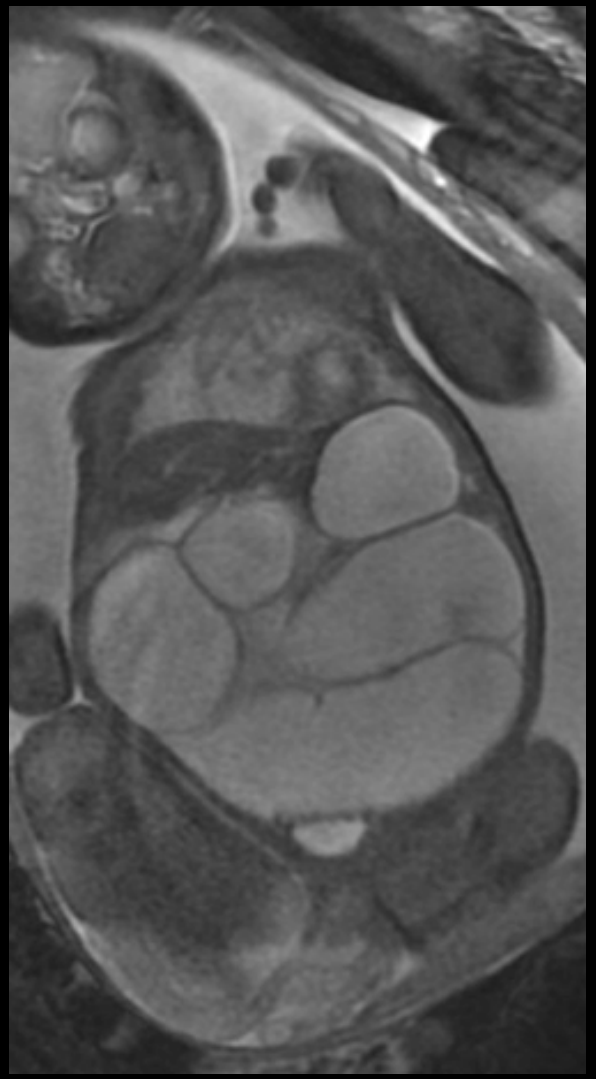
Postnatal Complications

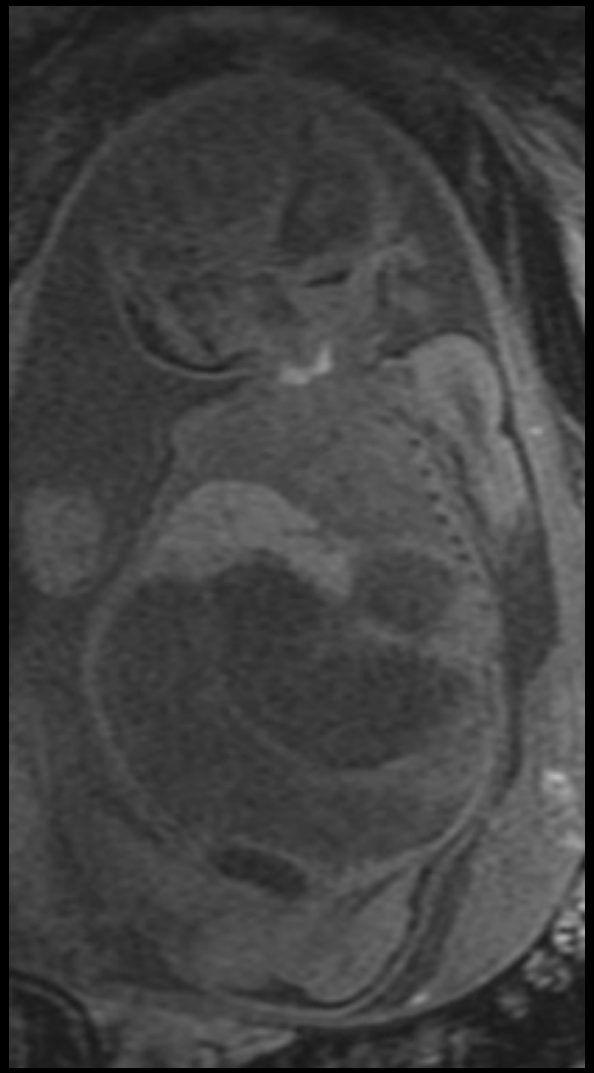
- Dehydration, Infection, Bowel Ischemia
- Atresia (10%)
- Bowel obstruction, perforation
- Edema and poor peristalsis, malabsorption
- IUGR

- Survival
  - 1943 ~10 %
  - 2013 ~ 90%

Short Gut Syndrome in Utero
Closed Gastroschisis

32 weeks of GA





Toxicity of Amniotic Fluid

- Increased urea and creatinin
- Decreased Na and osmolarity
- Maconium, GI content products*
- Inflamatory process Il-6

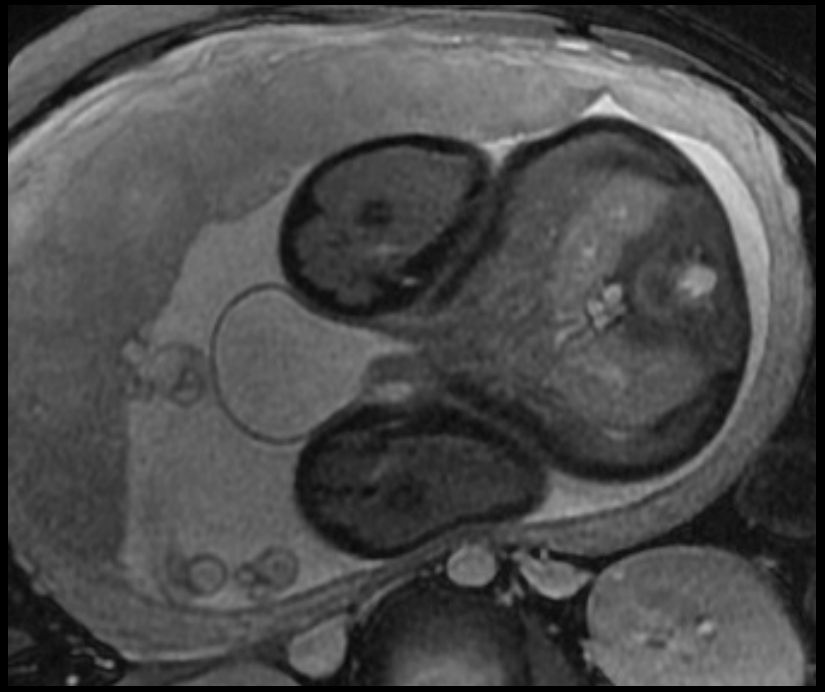
**Olguner, et al. J Pediatr Surg 35 (2000), pp. 458–461*

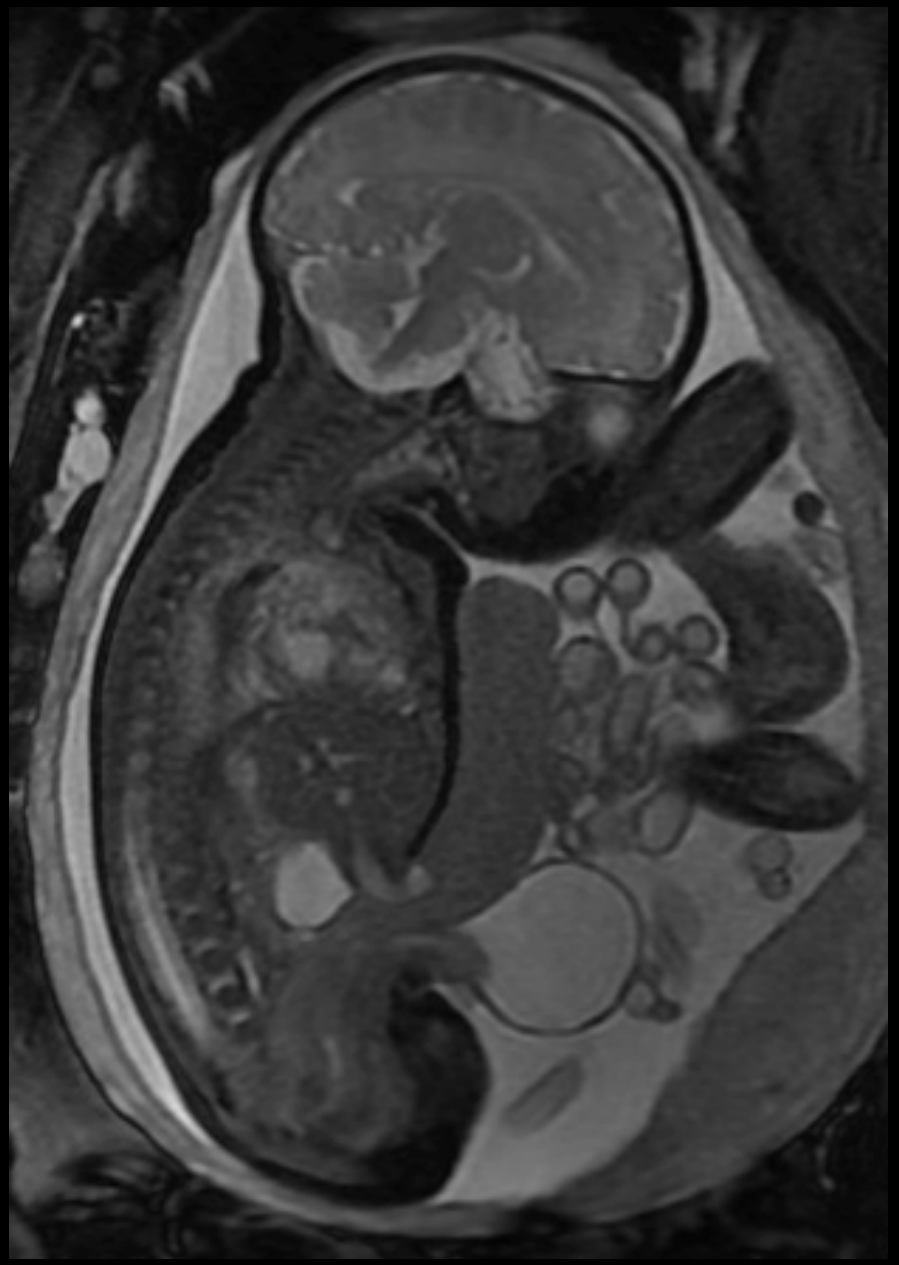
Management

- Steroids
- Amnioexchange

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

** Abuhamad et al. Am J Obstet Gynecol 176 (5) (1997) 985–990*



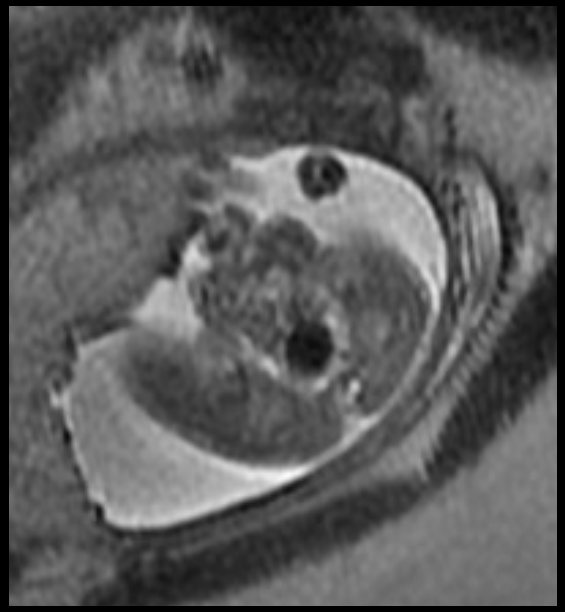


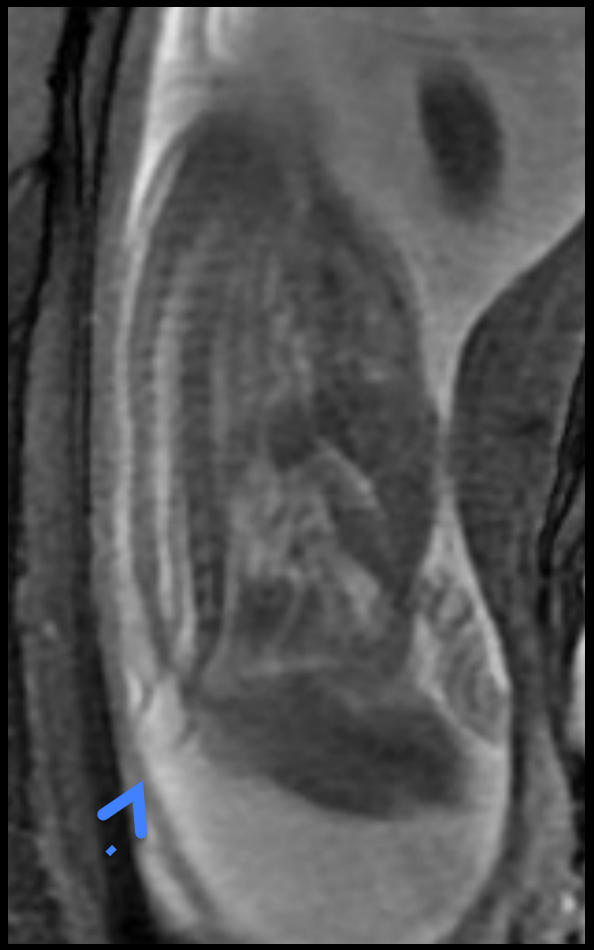
Bladder Exstrophy

- 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





OEIS

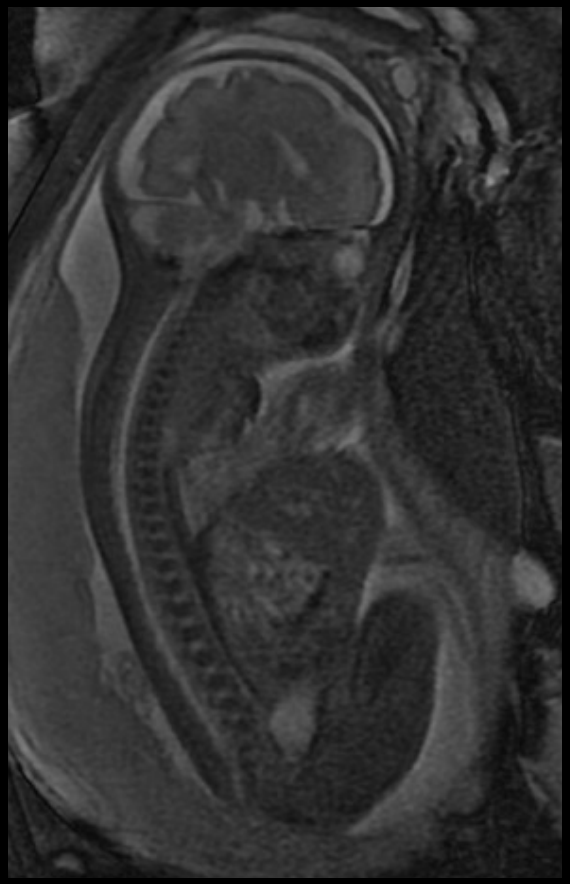
Omphalocele – Exstrophy – Imperforate Anus – Spinal Defect

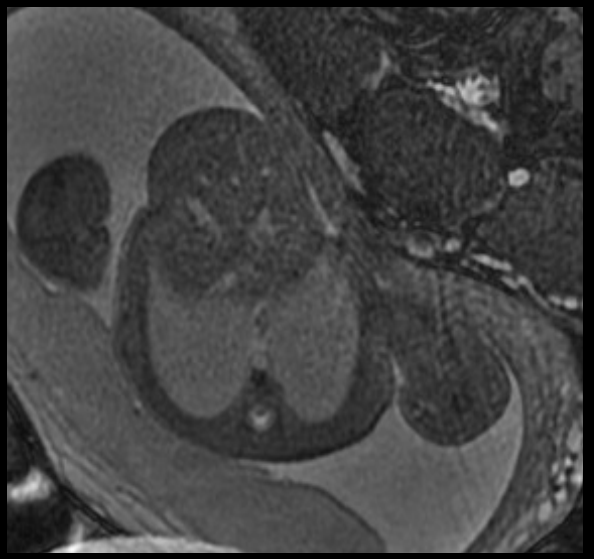
- Incidence: 1/200 000 – 400 000
- Underlying cause is unknown
- Cloacal exstrophy when
  - Two hemibladders
  - Rudimental colon and prolapse of terminal ileum


Voluson
E8
BARB ANATOMY
Har-mid
95
Gn -2
C4 / M7
FF3 / E2
SRI II 5 / CRI 3

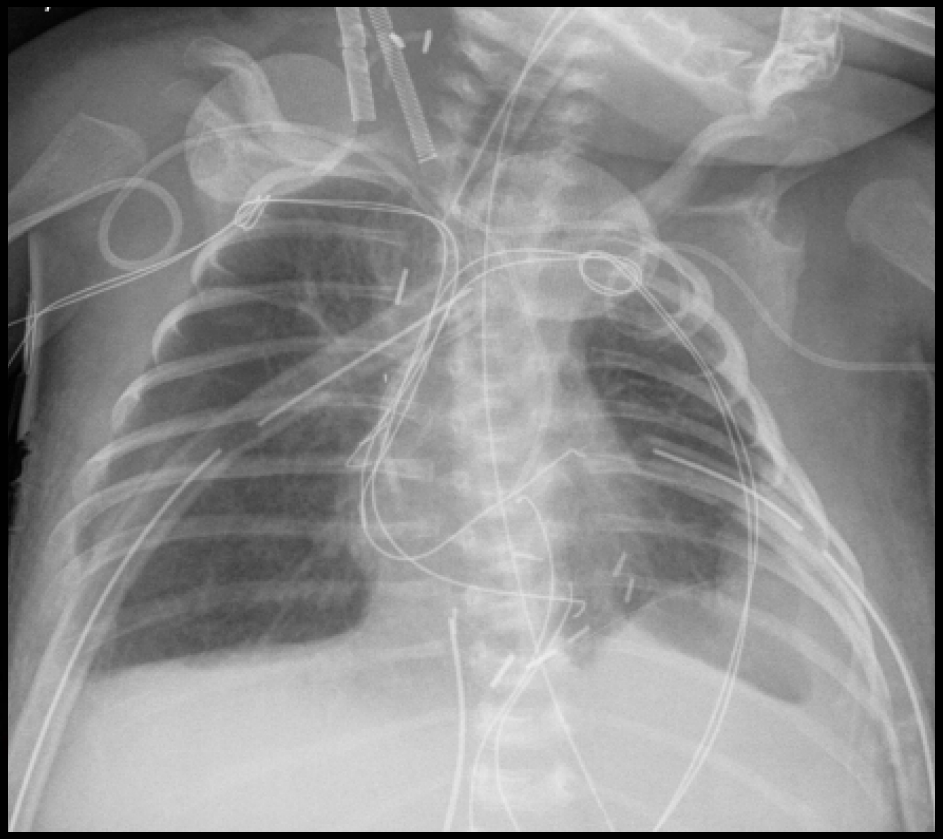
Ectopia Cordis and Omphalocele


ARM
HRT
LIVER





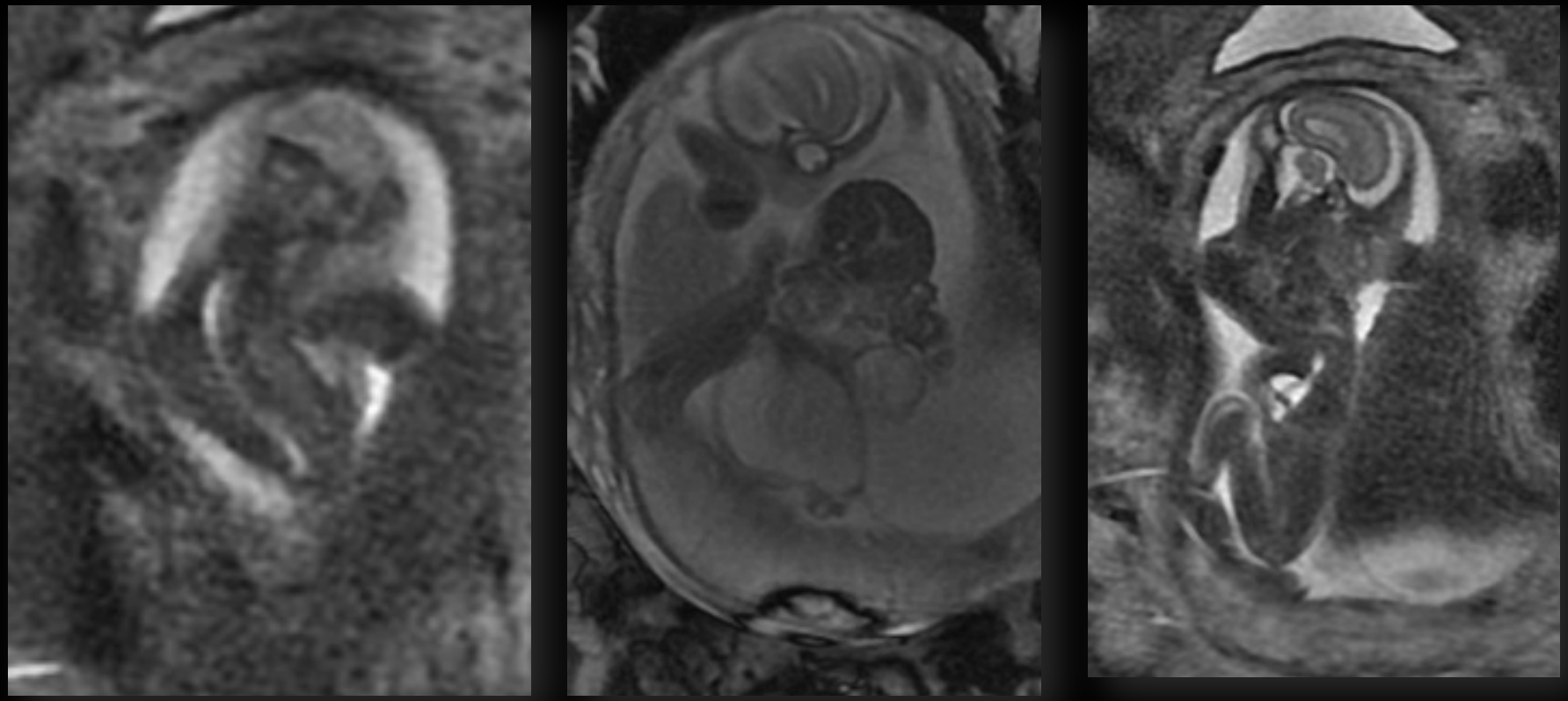
28 weeks of GA



After birth

At 2 months

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