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Abdominal wall defects – diagnosis and management

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Classification

Gastroschisis Exomphalos

Complex abdominal wall defects

Pentalogy of Cantrell

Bladder/cloacal extrophy

Body stalk anomaly

Amniotic bands

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Gastroschisis

Prevalence: 1: 3,000 births

Diagnosis:

- Paraumbilical right abdominal wall defect
- Evisceration of bowel, floating freely in the amniotic fluid

Causes:

- Sporadic
- Young women
- Drug abuse

Complications:

- Bowel obstruction 20%
- Fetal growth restriction 50%
- Spontaneous preterm birth 30%
- Fetal death 3%

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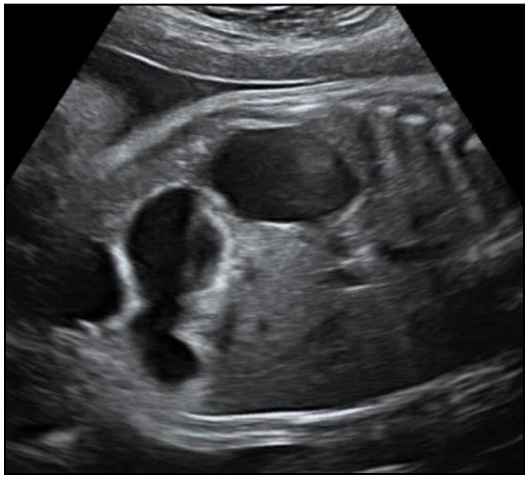
Gastroschisis

Follow-up : every 4 weeks
- Growth (Sieme formula)
- Well being (Doppler UA and MCA)
- Bowel dilatation

Delivery : vaginal, at 38 weeks, earlier if
- poor growth
- fetal hypoxia
- intra-abdominal bowel dilatation (>20 mm)

Prognosis : >90% survival
- Main cause of death: short bowel syndrome

Recurrence: 3%



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Exomphalos

Prevalence:

- **Only bowel in sac**
- 11 w 1 in 100
- 12 w 1 in 800 (90% resolve by 20 w)
- 13 w 1 in 2,000

- **Liver in sac**
- 11-13 w 1 in 3,500

Diagnosis:

Midline sac containing bowel / liver with umbilical cord at apex

Exomphalos

Associated abnormalities:

- Chromosomal defects (T18 and T13): 30-50%
- Genetic syndromes (Beckwith-Wiedemann syndrome): 10%
- Other defects (mainly cardiac): 30-50%

Management:

- Karyotyping and molecular testing for Beckwith-Wiedemann

Follow-up : every 4 weeks; growth (Sieme formula)

Delivery : vaginal, at 38 weeks

- Earlier if poor growth and hypoxia
- C-section: giant exomphalos (>75% of liver in sac)

Exomphalos

Prognosis :

Survival

- Isolated:
  - >90% for small / moderate
  - >80% for giant
- Non-isolated – depends of the associated defects

Recurrence:

- Isolated: no increased risk
- Part of trisomies: 1%
- Part of BWS up to 50%

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Bladder extrophy

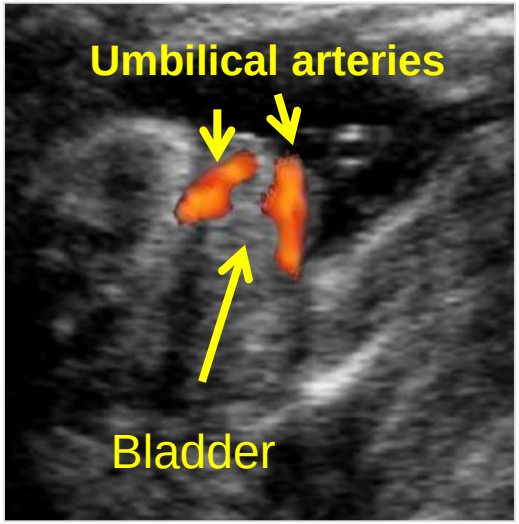
Prevalence:

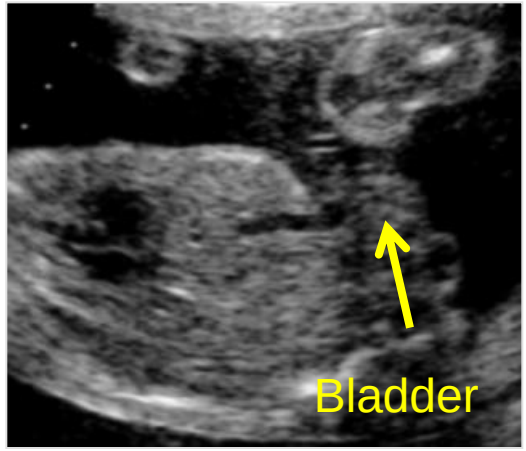
1 in 30,000 births

Etiology: Sporadic

Prenatal findings:

- Normal amniotic fluid
- Bladder not visible
- Suprapubic mass
- Low umbilical cord
- Splayed iliac crests
- Small penis, bifocal clitoris





Bladder extrophy

Management:

- Amniocentesis to determine the genetic sex of the fetus

Follow-up : standard

Delivery : vaginal, at 38 weeks

Prognosis :

- Survival > 95%
- Surgery for bladder closure
  urinary continence, epispadias

Recurrence:

- No increased risk

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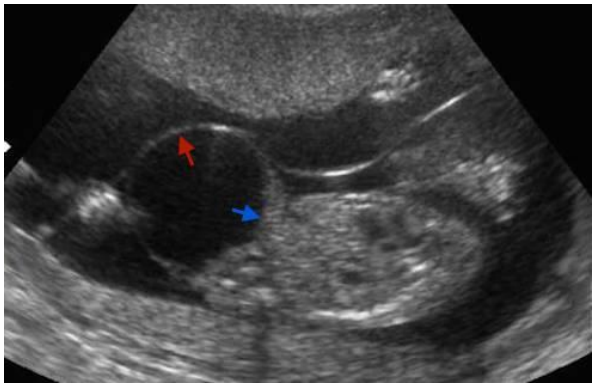
Cloacal extrophy

Omphalocele **E**xtrophy of the bladder **I**mperforate anus **S**pinal defects

Prevalence: 1 in 300,000 births

Etiology: Sporadic



Diagnosis:

- Low exomphalos
- Non-visible bladder
- Sacral spina bifida
- Normal volume of the amniotic fluid

Management:

Amniocentesis for genetic sexing

Normal delivery at term

Prognosis :

- Survival: >90% after extensive reconstructive surgery
- Normal lifestyle and fertility after surgery
- Some form of urinary tract diversion is required

Recurrence: No increased risk

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Body stalk anomaly

Prevalence: 1 in 10,000 births

Etiology: Sporadic

Diagnosis:

- Major abdominal wall defect
- Severe kyphoscoliosis
- Short or absent umbilical cord
- Liver directly attached to the placenta

Associated abnormalities:

- Exencephaly or encephalocoele, facial cleft, and limb amputations are common

Prognosis : Lethal - either in utero or in early neonatal period

Recurrence: No increased risk