

# Abdominal wall defects – diagnosis and management

**Anca Panaiteescu**  
**Kypros H. Nicolaides**



## Gastroschisis Exomphalos



## Complex abdominal wall defects



**Pentalogy of Cantrell**

**Bladder/cloacal extrophy**

**Body stalk anomaly**

**Amniotic bands**

# 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%  |



## **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

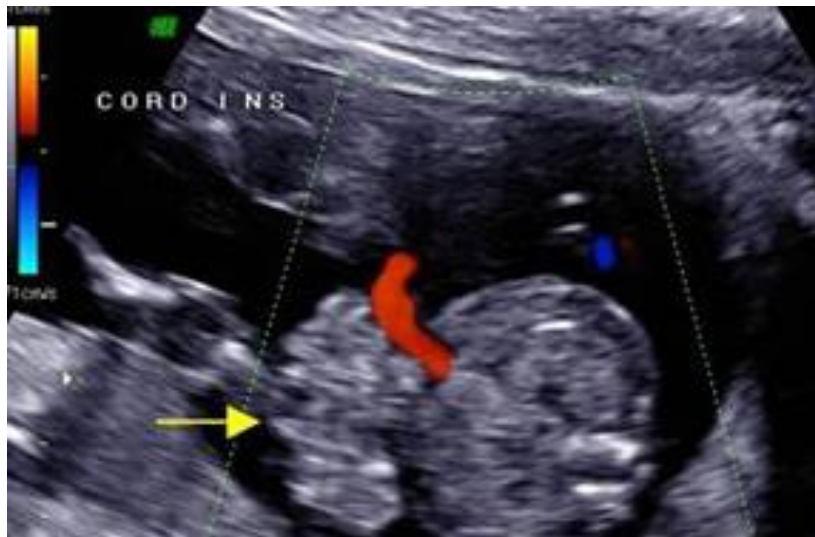
- Main cause of death: short bowel syndrome

## **Recurrence:** 3%





# Gastroschisis



## 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**



## 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)

## **Prognosis :**

### **Survival**

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

### **Recurrence:**

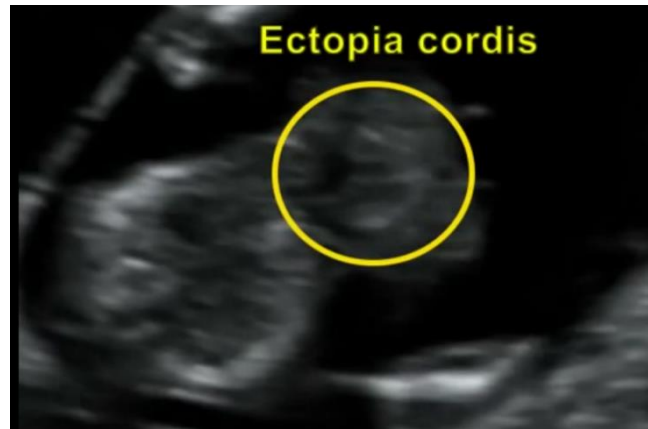
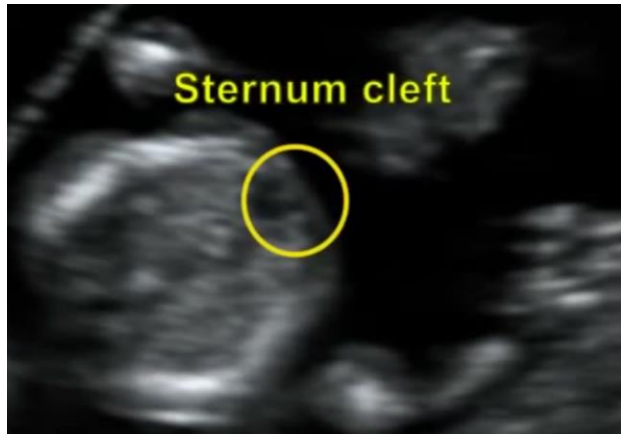
- **Isolated: no increased risk**
- **Part of trisomies: 1%**
- **Part of BWS up to 50%**



# Exomphalos



# Pentalogy of Cantrell



# Bladder extrophy

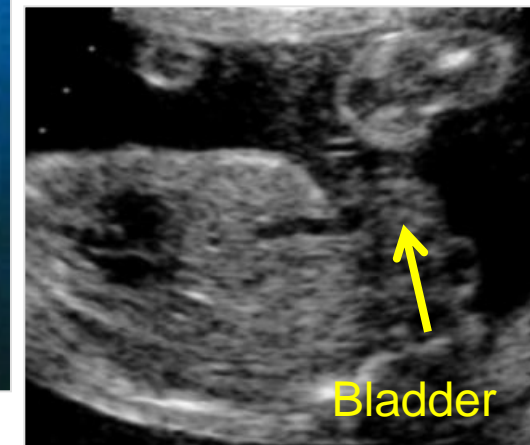
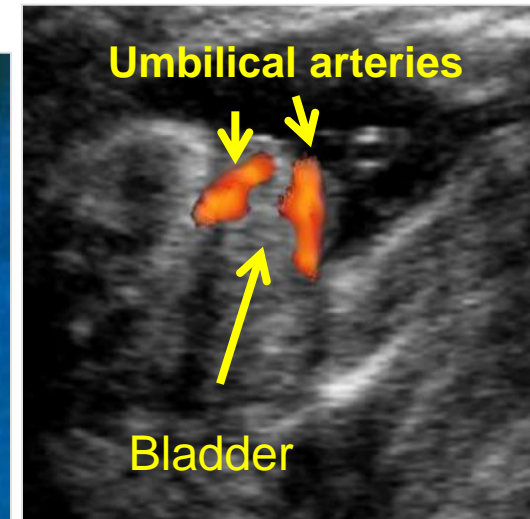
## Prevalence:

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





**O**mphalocele **E**xtrophy of the bladder **I**imperforate 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

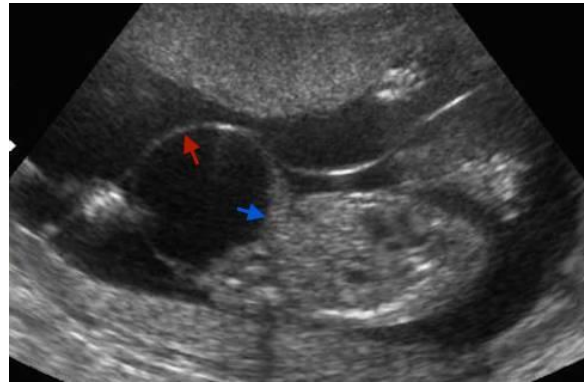
**Management:**

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



## 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

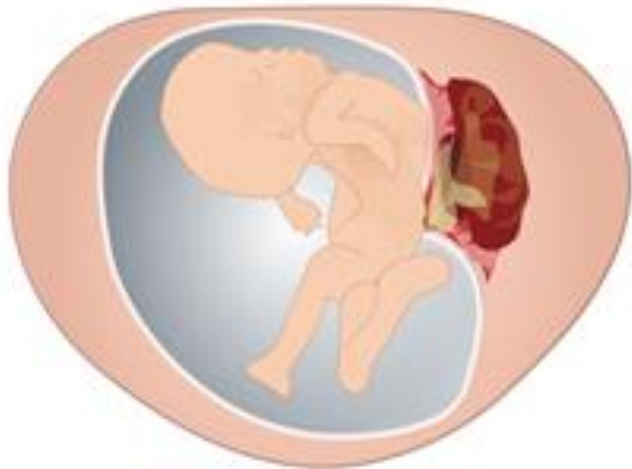
**Prognosis :** Lethal - either in utero or in early neonatal period

**Recurrence:** No increased risk

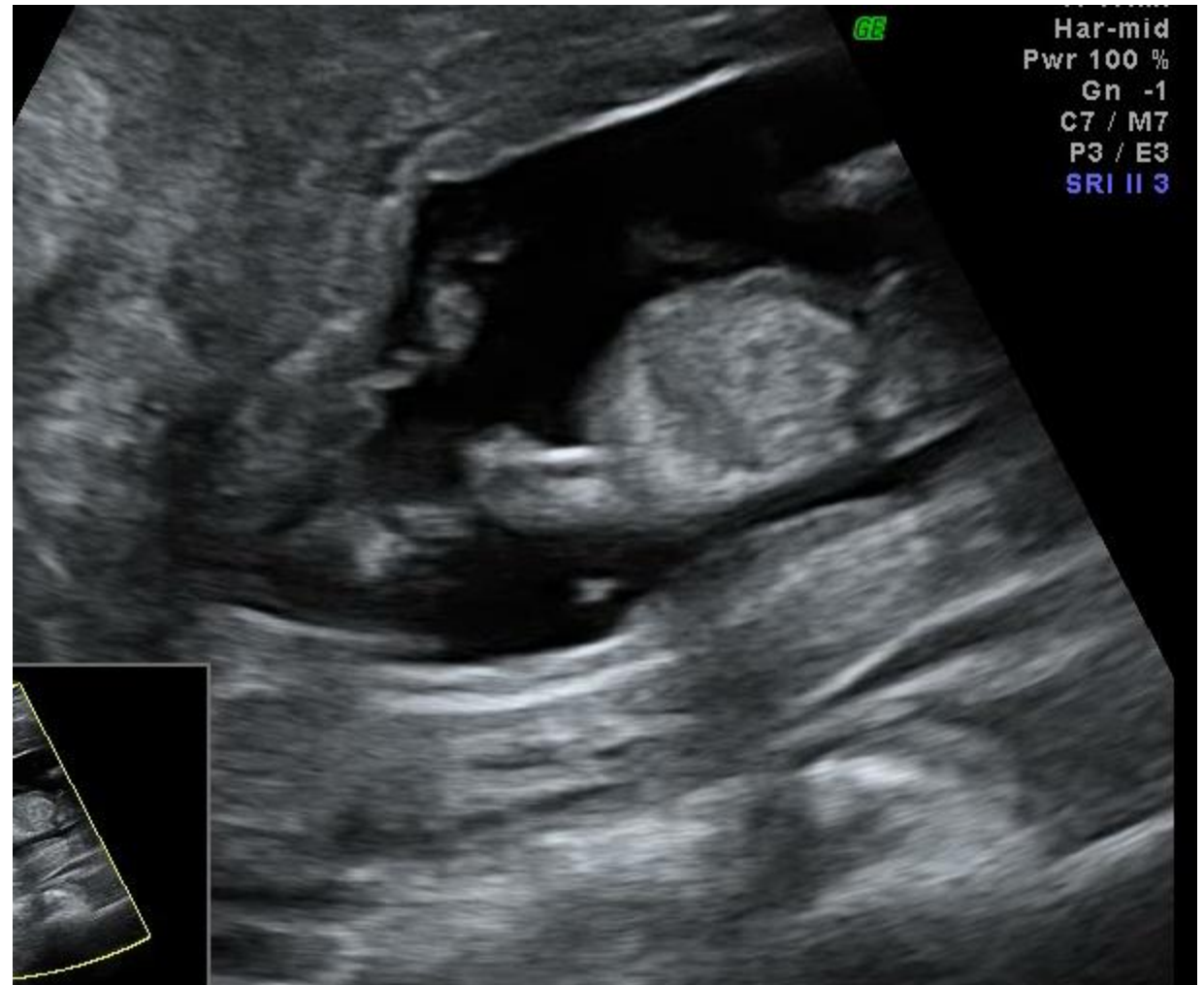




# Body stalk anomaly



## Amniotic bands syndrome





**Table 2.** Differential diagnosis of abdominal wall defects in the first trimester of pregnancy

Defect	Herniated viscera	Herniation site	Umbilical cord	Amniotic membrane	Coelomic space	Fetal mobility	Spine
Exomphalos	Liver, bowel	Base of umbilical cord	Free-floating	Continuous, fused with chorion	Obliterated	Normal	Normal/ kyphoscoliosis
Gastroschisis	Bowel	Amniotic cavity	Free-floating	Continuous, fused with chorion	Obliterated	Normal	Normal
Pentalogy of Cantrell	Heart, liver, bowel	Amniotic cavity	Free-floating	Continuous, fused with chorion	Obliterated	Normal	Normal
Cloacal extrophy/ OEIS complex	Cloaca	Amniotic cavity	Free-floating	Continuous, fused with chorion	Obliterated	Normal	Normal/ kyphoscoliosis
BSA	Liver, bowel	Coelomic cavity	Absent	Interrupted at the level of herniated abdominal organs	Contains abdominal organs	Stuck through abdominal-placental attachment	Kyphoscoliosis
Abdominoschisis with amniotic bands	Liver, bowel	Amniotic cavity	Free-floating	Ruptured	Obliterated	Normal	Normal

Thank you !