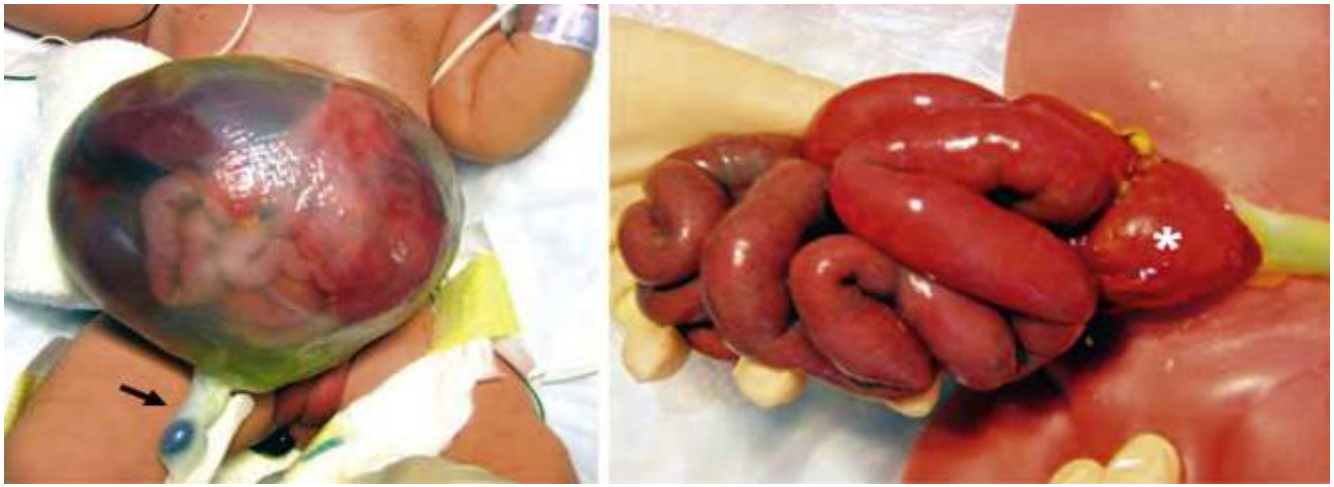
Congenital Abdominal Wall Defects

The two primary congenital abdominal wall defects are:

Omphalocele and Gastroschisis



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Introduction and Etiology

The abdominal wall forms during the fourth week of gestation.

During the sixth week, rapid intestinal and liver growth leads to herniation of the midgut into the umbilical cord. Elongation and rotation of the midgut occurs over the ensuing 4 weeks.

By week 10, the midgut returns to the abdominal cavity, where the first, second, and third portions of the duodenum and the ascending and descending colon assume their fixed, retroperitoneal positions.

An abdominal wall defect involves an interruption of these embryologic processes.

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Etiology

Gastroschisis: currently, the ventral body folds theory, which suggests failure of migration of the lateral folds (more frequent on the right side), is most widely accepted.

Omphalocele: develops due to a failure of the viscera to return to the abdominal cavity

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Possible causative factors

- Tobacco
- Environmental exposures (nitrosamines)
- Cyclooxygenase inhibitor use (aspirin and ibuprofen)
- Decongestants (pseudoephedrine and phenylpropanolamine)
- Low maternal age: younger than 21 years
- Prematurity

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Differentiating Characteristics Between Gastroschisis and Omphalocele

Characteristic	Omphalocele	Gastroschisis
Herniated viscera	Bowel ± liver	Bowel only
Sac	Present	Absent
Associated anomalies	Common (50%)	Uncommon (<10%)
Location of defect	Umbilicus	Right of umbilicus
Mode of delivery	Vaginal/cesarean	Vaginal
Surgical management	Nonurgent	Urgent
Prognostic factors	Associated anomalies	Condition of bowel

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Diagnosis

- **Prenatal:**
  - **Ultrasound**
  - **Intrauterine growth restriction (IUGR)**
  - **Maternal AFP: High**

- **Postnatal: Clinical**

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Gastroschisis

Incidence: 1 in 4000 live births.

Intestinal atresia is the most common associated anomaly.
Malrotation occurs in nearly every patient with gastroschisis but midgut volvulus is not commonly seen.

The exposed viscera may be covered with a think inflammatory peel. Bowel loops may be densely adherent making it impossible to distinguish one loop of bowel from another.

(Emergency repair, the more you wait, the more inflammation and the more hypothermia)

Classification:

- *Simple* gastroschisis (i.e. viable bowel, no atresias)
- Complicated gastroschisis (i.e. Perforated bowel, atresia)

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Closure approaches:

- Placement of Silastic® silo with staged reduction of the viscera over several days followed by delayed facial closure with suture

- Complete reduction of the viscera shortly after birth and primary facial closure with suture

- Complete reduction of the viscera shortly after birth and covering of the defect with an umbilical cord flap (Sutureless)

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Complications

- Abdominal compartment syndrome (ACS) can occur within the first few days following reduction and closure of a gastroschisis defect. ACS may manifest as increasing pulmonary pressures in ventilated patients, low urine output, differential cyanosis (bluish appearing legs caused by impaired venous return) and hypotension.

- Ten to 15% of patients with gastroschisis develop necrotizing enterocolitis (NEC).

- Intestinal atresia

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Omphalocele

Incidence: one to three per 10,000 live births.

Cardiac defects are observed in 30 to 50 %.

Karyotype abnormalities occur in 30 % of cases with trisomy 13, 18 and 21 being most common.

Many syndromes are associated with omphalocele:

- Beckwith-Wiedemann syndrome (congenital abdominal wall defect, macroglossia, hypoglycemia, and propensity for development of abdominal tumors)
- Pentalogy of Cantrell (epigastric omphalocele, diaphragmatic defect, pericardial defect, sternal cleft, and cardiac defect).

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Closure approaches:

- *Primary closure* if small

- Giant omphaloceles, however, are managed by allowing epithelialization of the sac with topical application of silver sulfadiazine, serial reductions and elective repair at six to 24 months (as a ventral hernia)

- Ruptured omphaloceles have very poor prognosis and require silo placement with delayed primary closure.

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Umbilical and Other Abdominal Wall Hernias

Umbilical Hernia:

- Failure of closure of umbilical ring
- The hernia sac is peritoneum
- The extent of skin protrusion is not always indicative of the size of the fascia defect.
- Umbilical hernias are present in 15–25% of newborns
- Premature and low birth weight infants have a higher incidence
- Umbilical hernias usually close spontaneously.
- It is very safe to observe the hernia until ages 4–5 years to allow closure to occur.

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- **Epigastric Hernia:**

- Hernias of the abdominal wall through the midline linea alba, with a location between the umbilicus and xiphoid process

- Incidence up to 5%

- These hernias present as either a small painless mass, which becomes painful with activity, or a small painful incarcerated mass.

- Typical contents are preperitoneal fat

- Epigastric hernias do not resolve and should be repaired

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Spigelian Hernia: (Spigelian Triangle)

- Quite rare in children and can be difficult to detect and diagnose.
- The actual defect occurs at the intersection of the linea semicircularis, linea semilunaris, and the lateral border of the rectus abdominis muscle
- More frequently in girls
- A tension-free closure is important to prevent recurrence

Lumbar Hernia:

- Bulge in the area bordered by the 12th rib, sacrospinalis muscle, and internal oblique muscle
- The bulge is usually due to herniated preperitoneal fat.
- Physical findings include a soft mass that is easily reducible.
- Repair is advisable because the defect never resolves spontaneously and incarceration is possible.

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