

تبييض محاضرة

Congenital Abdominal Wall Defects

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Done by :



* At the intrauterine life :

- the bowel is in contact (direct communication) with the amniotic fluid which causes irritation to the bowel → which makes bowel adherent to each others.

* The sac of omphalocele may devdop to skin → Then it may causes hernia → so, we repair the herria.

* case :

On US, if the herniation is seen at the week 7 is it omphalocele ?

- No!!

- The return of bowel to the abdomen cavity occure af 10th week., if herniation is seen after the 10th week → so, it's omphalocele



Congenital Abdominal Wall Defects

The two primary congenital abdominal wall defects are:

Omphalocele and Gastroschisis



Omphalocele → the defect is located **centrally**

Gastroschisis → most commonly of the **right side**, defect caused by **lateral folds** (THE MOST COMMON).

outer congenital anomalies such as ;

- umbilical hernia
- Epigastric hernia
- lumbar hernia



Introduction and Etiology

The abdominal wall forms during the fourth week of gestation.

→ Forms by 4 folds → 2 lateral , 1 superior , 1 inferior fold → fused together

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.

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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.



Etiology

→ All the layers of the abdomen are not present

→ the defect is **small**

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

→ the defect is **large**, SO ! More organs can be seen in

Omphalocele compared with gastroschisis



Possible causative factors

→ Chromosomal

→ Environmental

- Tobacco
- Environmental exposures (nitrosamines)
- Cyclooxygenase inhibitor use (aspirin and ibuprofen)
- Decongestants (pseudoephedrine and phenylpropanolamine)
- Low maternal age: younger than 21 years
- Prematurity → Causative agent for Gastroschisis more than omphalocele



Differentiating Characteristics Between Gastroschisis and Omphalocele

Characteristic	Omphalocele	Gastroschisis
Herniated viscera	Bowel ± liver Large Defect	Bowel only Due to small Defect
Sac	Present	Absent → (No peritoneum) (All the bowel is found outside)
Associated anomalies	Common (50%) Such as; cardiac anomalies ((m.c))	Uncommon (<10%) The most common is small bowel atresia
Location of defect	Umbilicus Centrally	Right of umbilicus
Mode of delivery	Vaginal/ <u>cesarean</u> ? To avoid rupture of cyst	Vaginal
Surgical management	<u>Nonurgent</u> Covered by hernia sac	<u>Urgent</u> Due to absent sac
Prognostic factors	Associated anomalies ① ② If the sac is ruptured or not	Condition of bowel



omphalocele is more poor prognosis due to the associated anomalies not due to the disease itself



	Gastroschisis	Omphalocele
PRESENTATION	Extrusion of abdominal contents through abdominal wall defect	Herniation of abdominal contents through umbilicus
COVERAGE	Not covered by peritoneum or amnion A ; “the guts come out of the gap (schism) in the letter G ”	Covered by peritoneum and amnion B (light gray shiny sac); “abdominal contents are sealed in the letter O ”
ASSOCIATIONS	Not associated with chromosome abnormalities; good prognosis	Associated with congenital anomalies (eg, trisomies 13 and 18, Beckwith-Wiedemann syndrome) and other structural abnormalities (eg, cardiac, GU, neural tube)



Diagnosis

((IUGR)) : when a baby in the womb (a fetus) does not grow as expected

- **Prenatal:**
 - **Ultrasound** → can detect both of them
 - **Intrauterine growth restriction (IUGR)**
 - **Maternal AFP: High**
 - ↪ Not specific for abdominal wall anomalies, it elevates also in meningocele , Sacrococcygeal teratoma
- **Postnatal: Clinical**



Gastroschisis

thick inflammatory peel :

is caused due to direct contact with amniotic

Gastroschisis fluid → inflammation → adhesions/perforation.

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 thick 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)

↪ Pre-natally , on US →

meconium is seen in the amniotic fluid



1• Pre-surgical intervention (stabilization) :

- NPO -IV fluid -IV antibiotic - Gastric decompression (NG tube)
- evacuate the stool (rectal stimulation).

- as much as we can decrease the distention → better closure of defect.

2• Closure approaches: Either ; primary closure or delayed closure

- Placement of Silastic® silo with staged reduction of the viscera over ^① several days followed by delayed facial closure with suture → Used to avoid ACS
- Complete reduction of the viscera shortly after birth and primary ^② facial closure with suture → if the defect is small, can be closed by suture
- Complete reduction of the viscera shortly after birth and covering of the defect with an umbilical cord flap (Sutureless) ^③



- silastic silo : allows the intestines to slowly move into the belly



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



Omphalocele

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

Cardiac defects are observed in 30 to 50 %. → makes the prognosis poorer
(Chromosomal)

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).



Closure approaches:

- **Primary closure** if small → if contents are **less** in amount
- **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) **→ The hernia itself**
 - when the sac develops to skin then form hernia.
 - contents are **high** in amount
 - Urgent case
- **Ruptured omphaloceles** have very poor prognosis and require silo placement with **delayed primary closure**.



Ventral wall defect ::

Skin bulging : umbilical hernia

Sac bulging: omphalocele

No skin no sac : gastrochiasis (no liver bec the defect is small)

If the defect <4cm mc anomaly is small bowel atresia

>4cm mc anomaly is cardiac

Umbilical and Other Abdominal Wall Hernias

low risk of
strangulation due to
the wide neck

Umbilical Hernia: to differentiate between amphalocele & umbilical hernia we consider the skin → if it's found → 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.



• 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** → usually symptomatic
- Epigastric hernias do not resolve and should be repaired



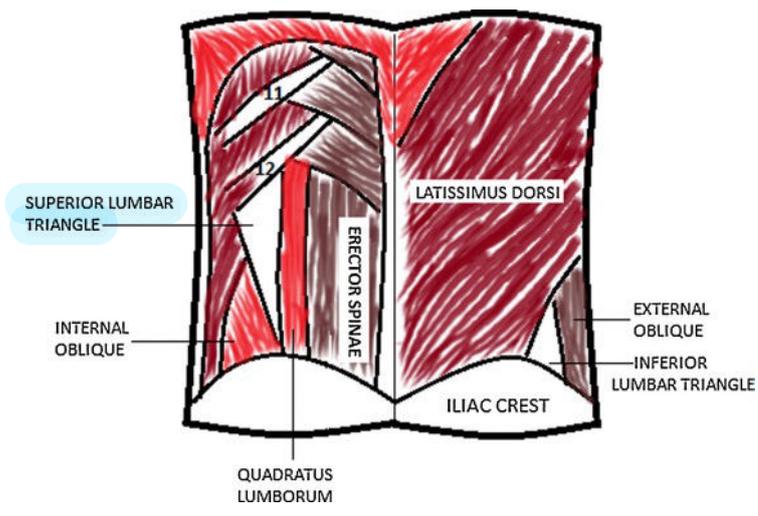
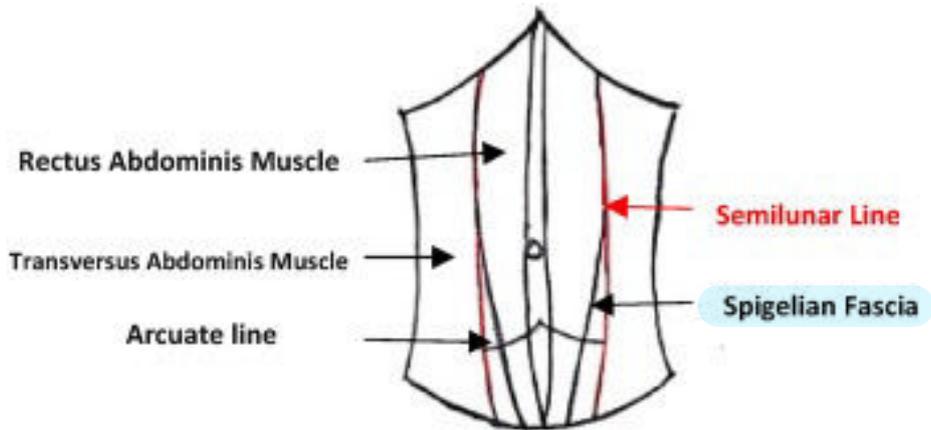
Spigelian Hernia: (Spigelian Triangle) → Lateral abdominal hernia

- 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
 - contents : pre-peritoneal fat

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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* All the following statements are true of infant with Anterior abdominal wall defects except:-

- A- It is associated with malrotation.
- B- There is prolonged adynamic ileus following repair.
- C- Staged procedure is performed by silo and delayed closure.
- D- Myo-cutaneous mobilization flap as operative option.
- E- The gastroschisis has a sac that may rupture during delivery

* Which of the following statements is FALSE regarding gastroschisis?

- a. It is associated with malrotation.
- b. There is a low incidence of associated anomalies.
- c. There is a prolonged adynamic ileus following repair.
- d. It is complicated by intestinal atresia in 10% to 15% of cases.
- e. It is associated with chromosomal syndromes.

* All the following statements are true of neonate with gastroschisis except :

- a. it is associated with malrotation
- b. there is prolonged a dynamic ileus following repair
- c. staged procedure is performed by silo and delayed closure
- d. Myo-cutaneous mobilization flap as operative option
- e. the umbilical cord attached to the right side of the defect

فَشَدُّ أَرْزِي بِتَوْفِيقٍ وَ مَقْدَرَةٍ

