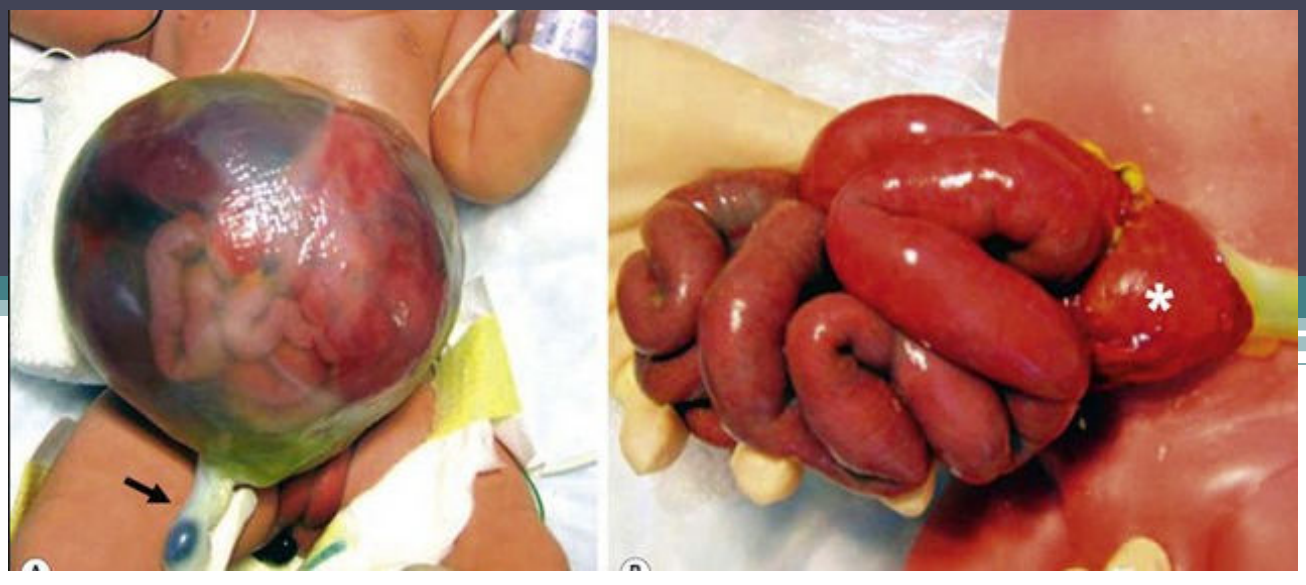
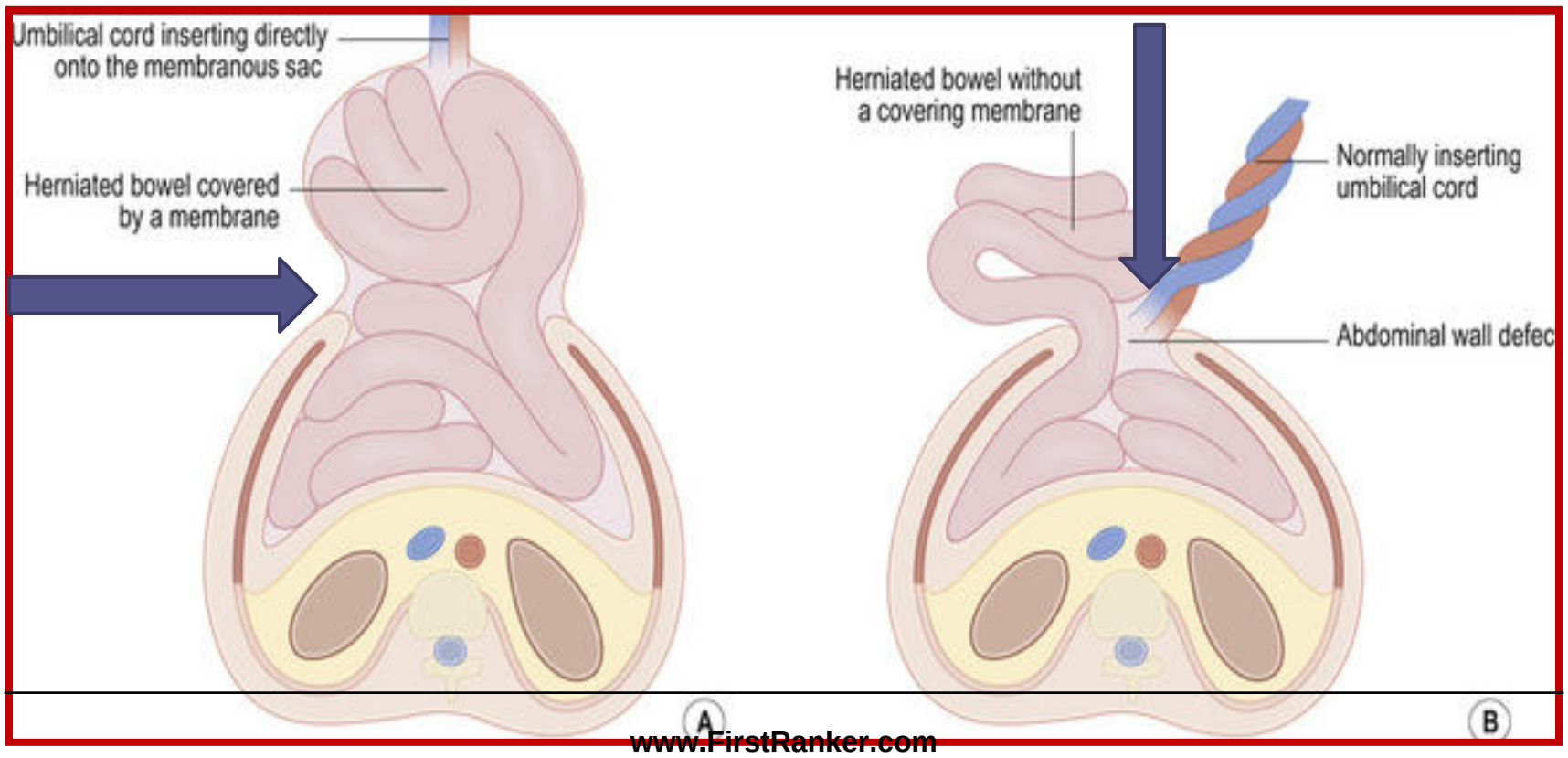
ABDOMINAL WALL DEFECTS : OMPHALOCELE AND GASTROSCHISIS



ABDOMINAL WALL DEFECTS

- A type of **congenital defect** that allows the abdominal organs to protrude through an **unusual opening (blue arrows)**that forms on the abdomen.



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- Types
- Gastroschisis
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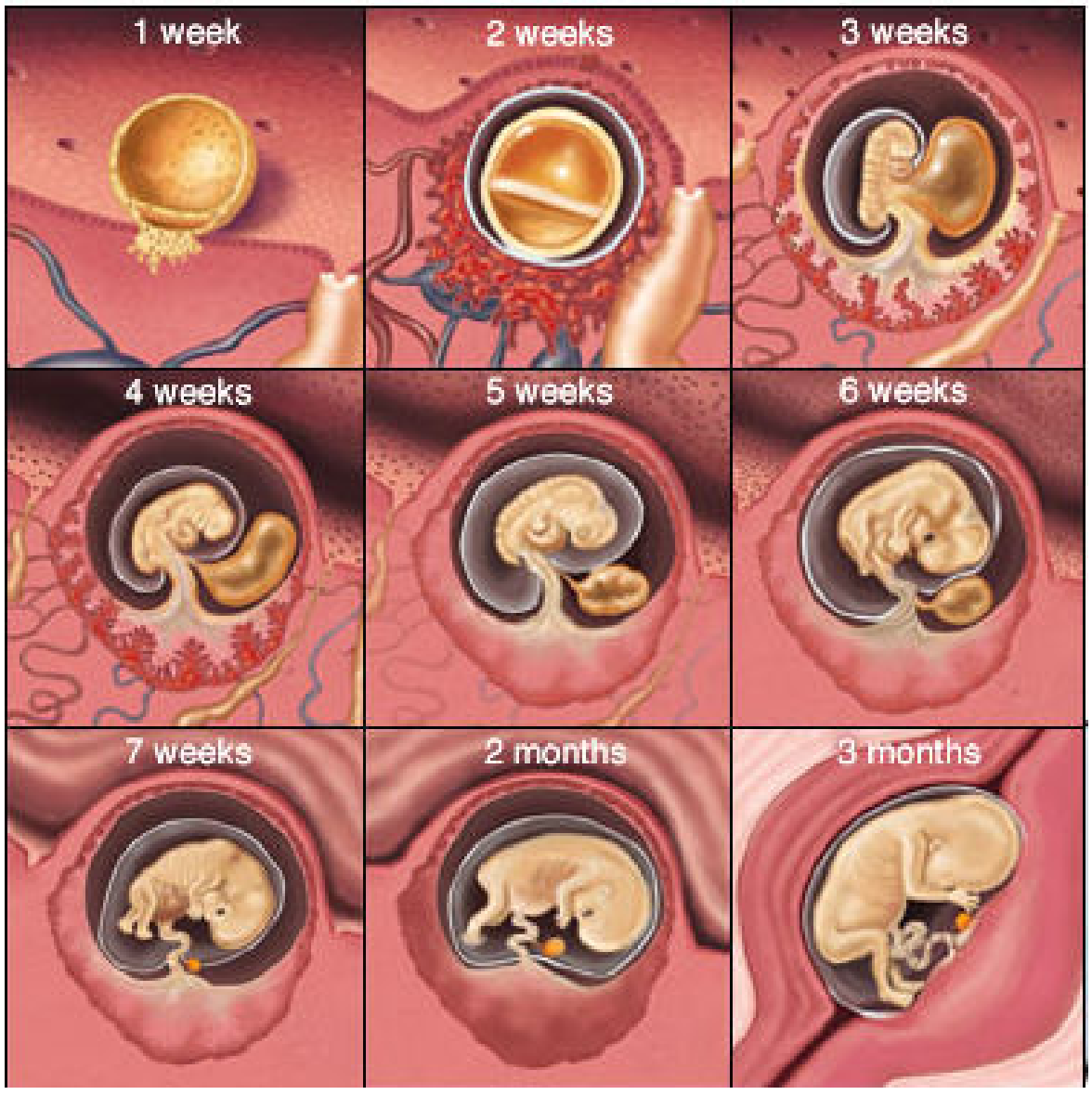
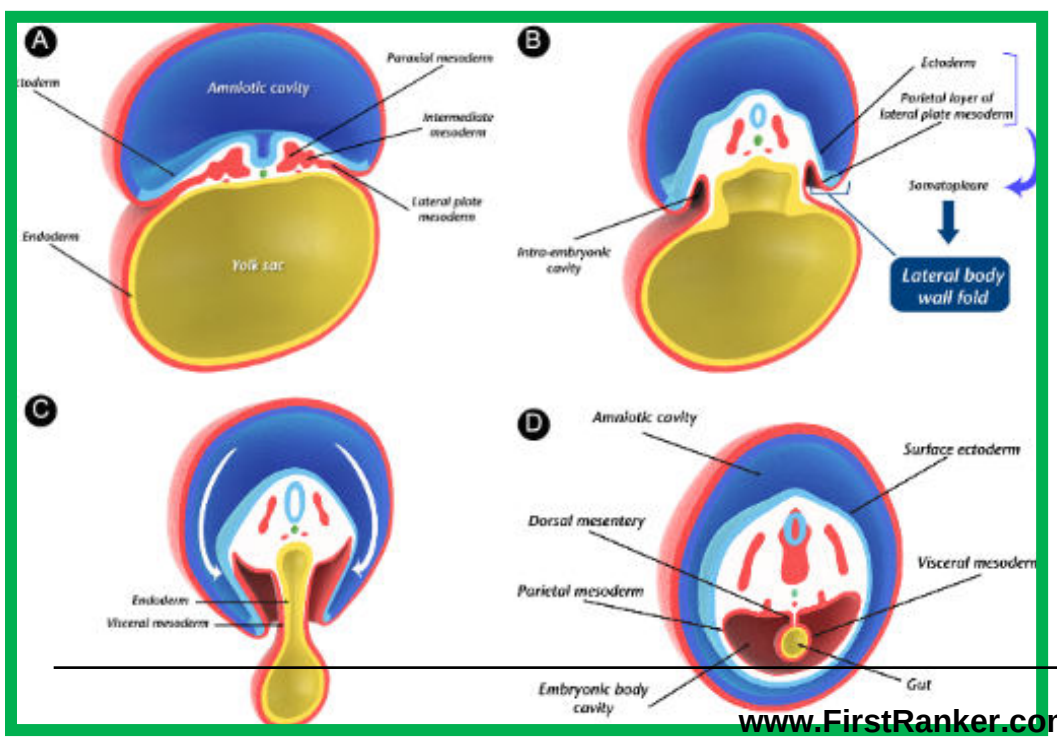
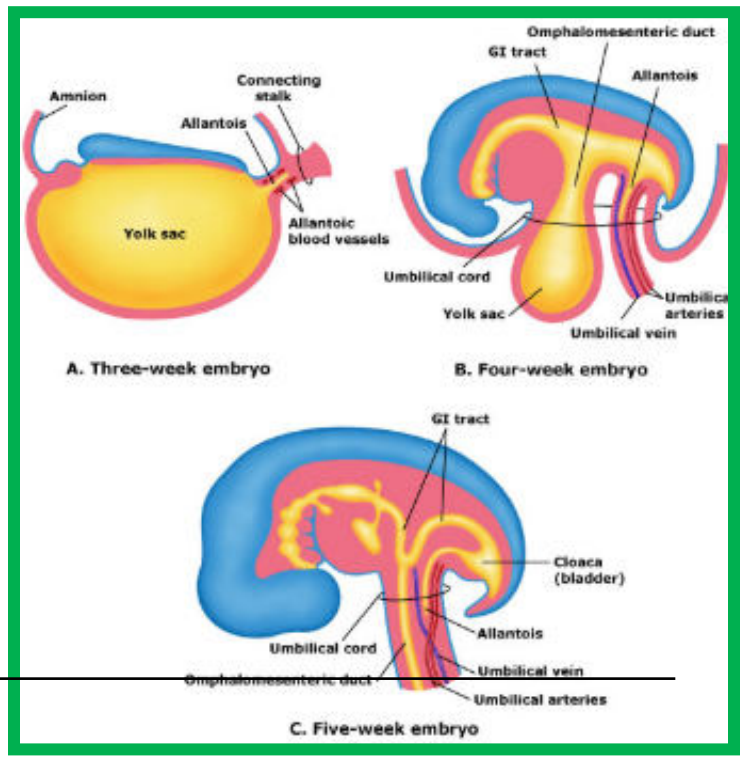


EMBRYOLOGY

- Closure of the body wall begins at **3 weeks' gestation** and results from growth and longitudinal infolding of the embryonic disks.





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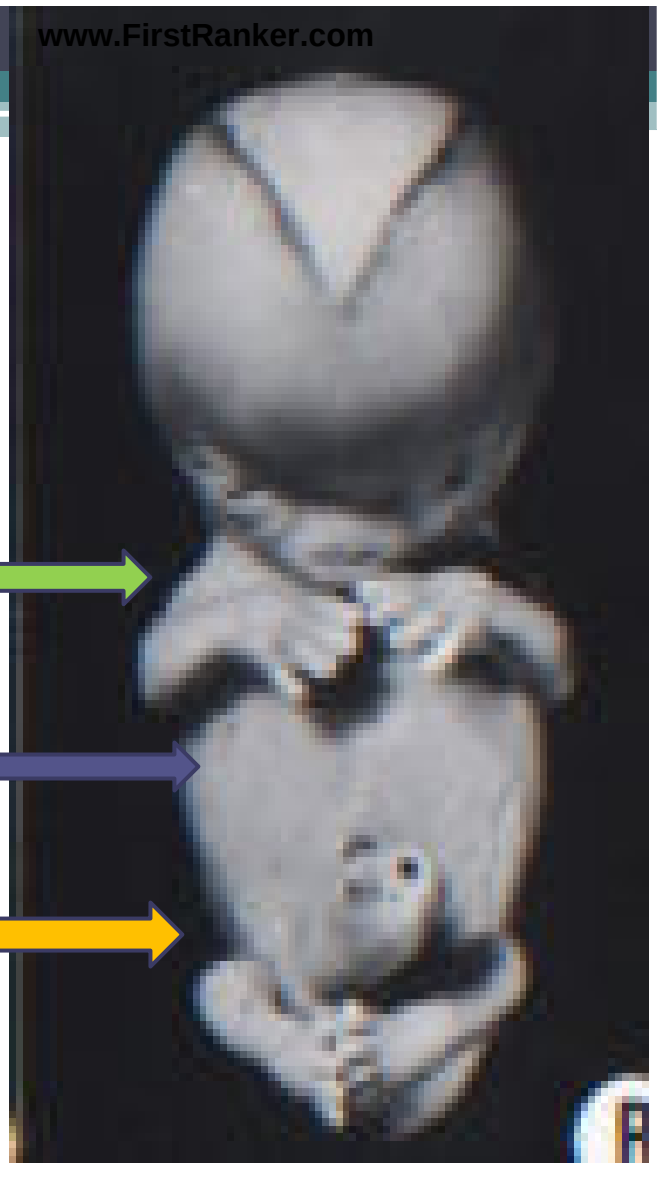
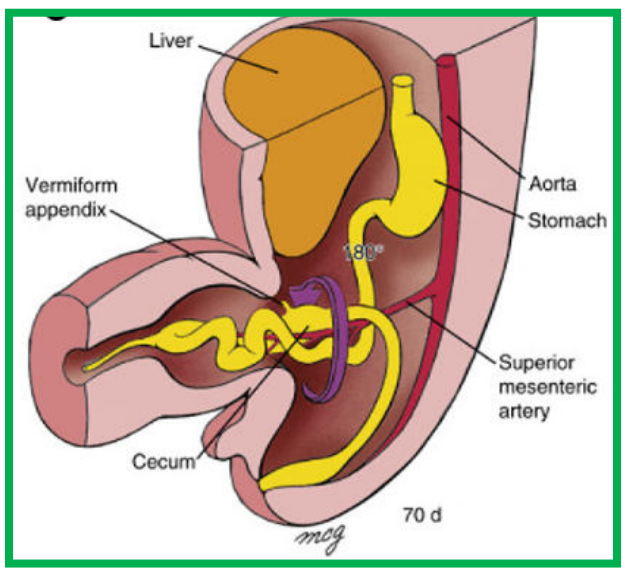
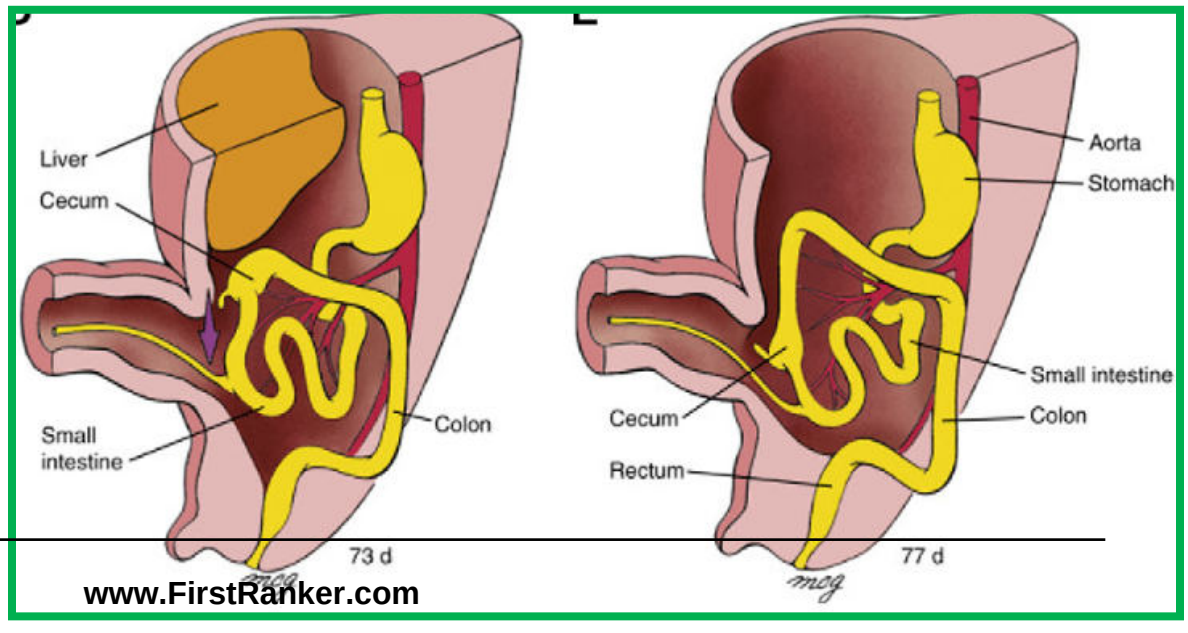


- The **cephalic fold** forms the thoracic and epigastric wall.
- The **lateral folds** form the lateral abdominal walls.
- The **caudal fold** contributes the hindgut, bladder, and hypogastric wall.
- These four folds meet in the midline to form **the umbilical ring.**

- **During 6th week** of gestation, rapid growth of intestines causes **herniation of the midgut** into the umbilical cord.
- **Week 10**, the midgut is **returned to the abdominal cavity** and the small bowel and colon assumes a fixed position.
- Any disruption in process may result in an **abdominal wall defect.**





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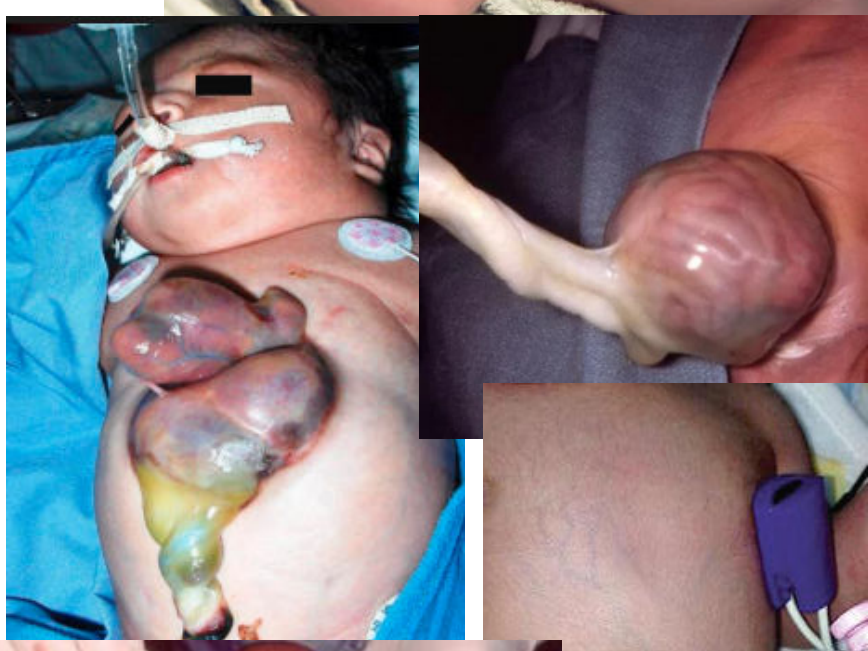
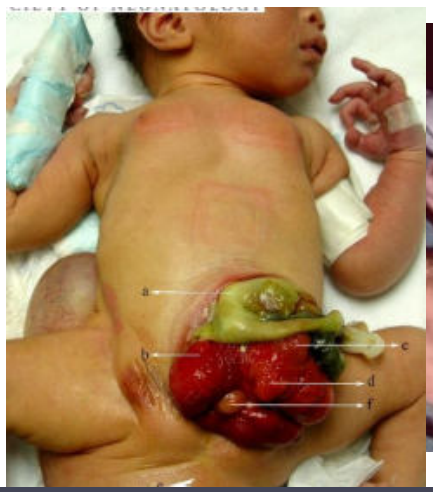
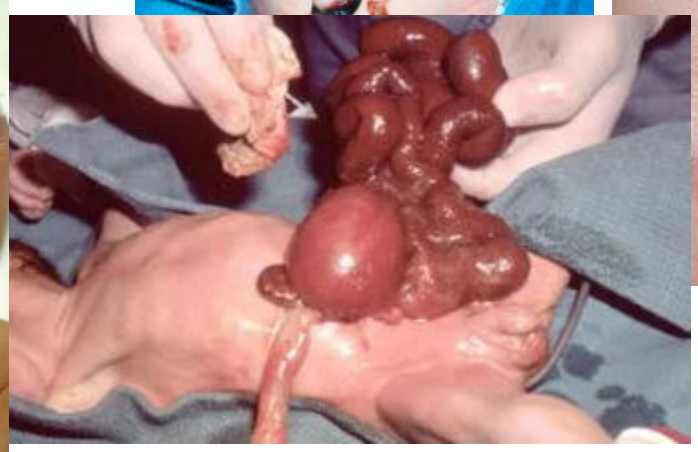
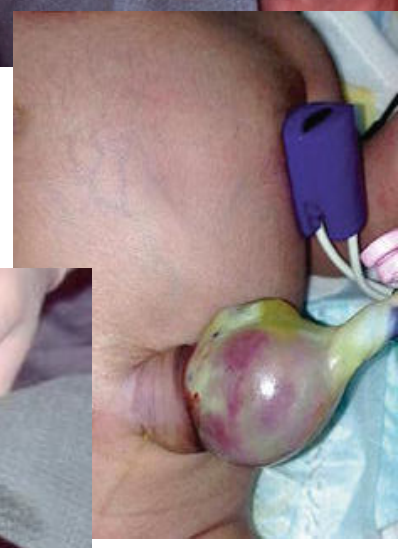
TYPES

1. Ectopia cordis thoracis – cephalic fold defect.
2. Pentalogy of Cantrell- cephalic fold defect.
3. Omphalocele – Failure of folding.
4. Umbilical cord Hernia – Small defect and normal abdominal wall.
5. Gastroschisis –
6. Cloacal exstrophy – caudal fold defect.







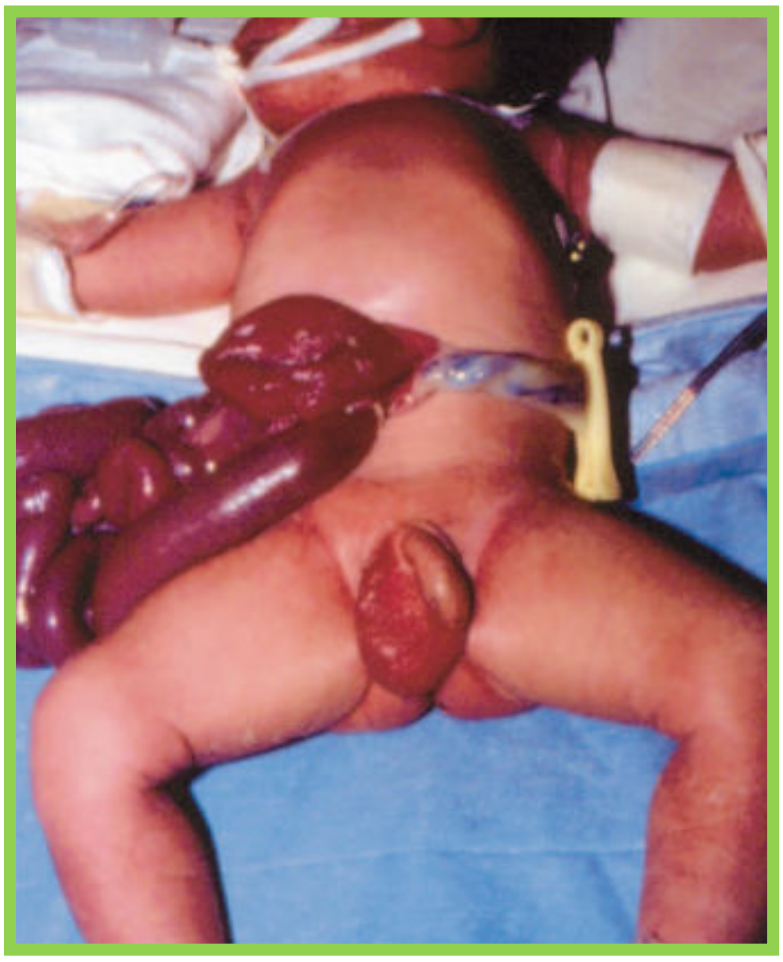


GASTROSCHISIS

OMPHALOCELE

GASTROSCHISIS- Most common

- Incidence : **2 to 4.9 per 10,000** live births.
- **Herniation of intestinal loops** through full-thickness defect in anterior abdominal wall.
- Defect lateral to the umbilicus (**right>left**), usually less than 4cm in size.
- **No sac** covers the extruded viscera (usu. only intestines).
- **Preterm** babies (28%).
- **Young** mothers (<25years).



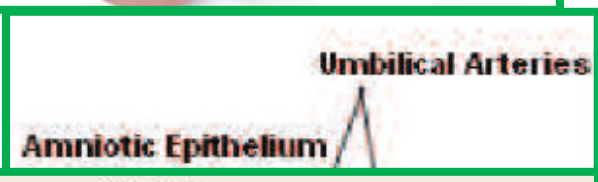
Etiology:

- **In-utero vascular accident.**

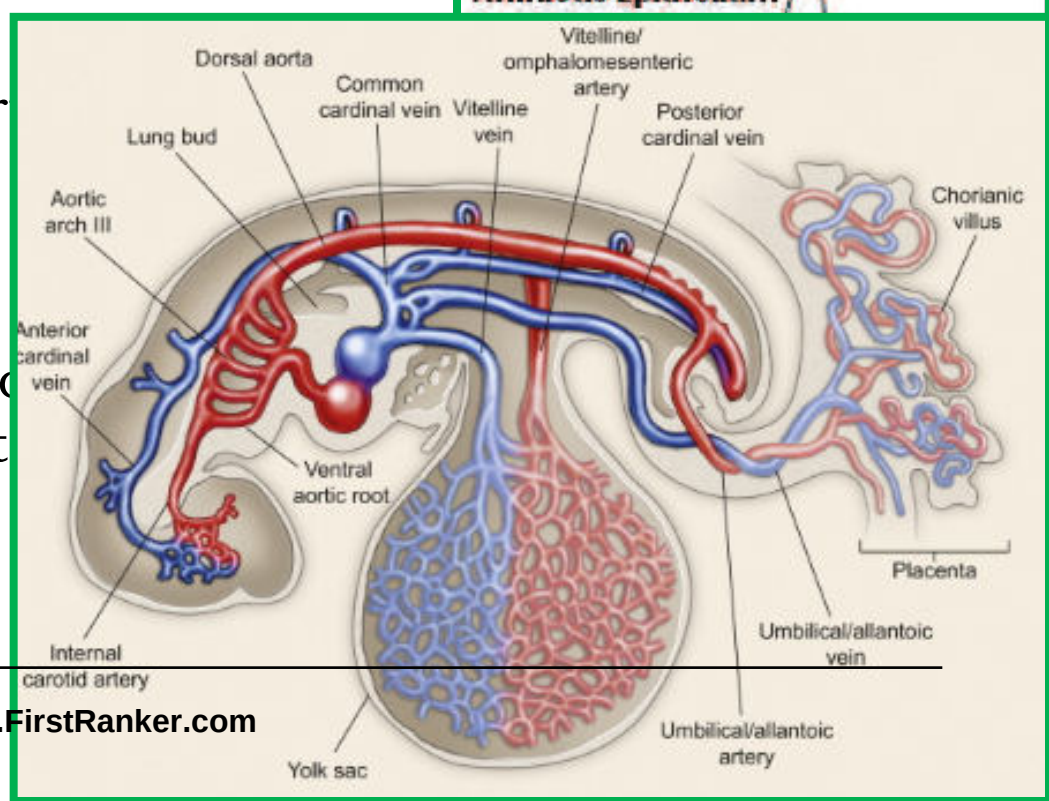
  2 theories

  1. Involution of the right umbilical vein causes necrosis in the abdominal wall leading to a right-sided defect.

  2.Right omphalomesenteric ar involutes

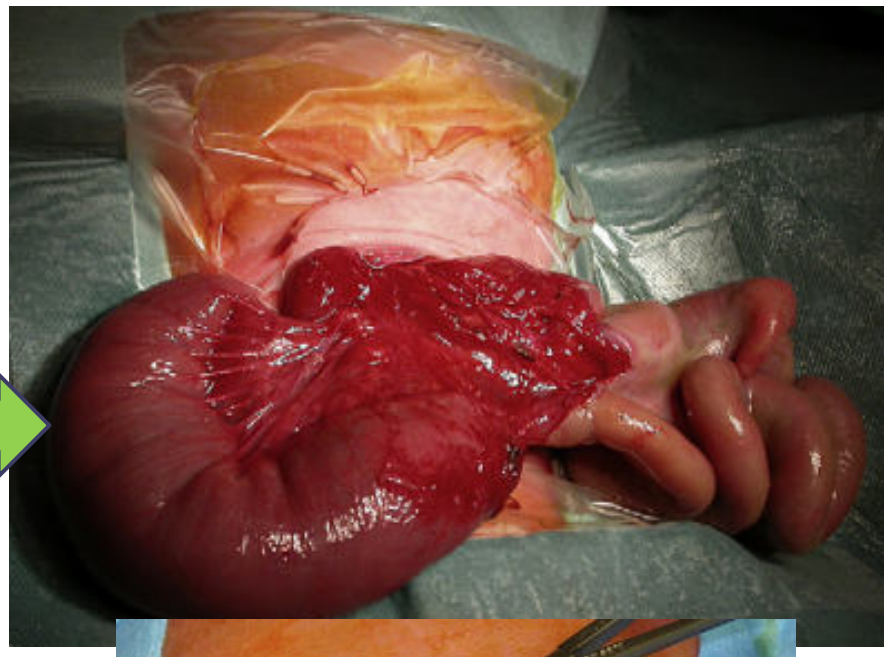


- **Other theories:**
  - In-utero rupture of omphaloc
  - Abnormal midline fusion of t abdominal folds.

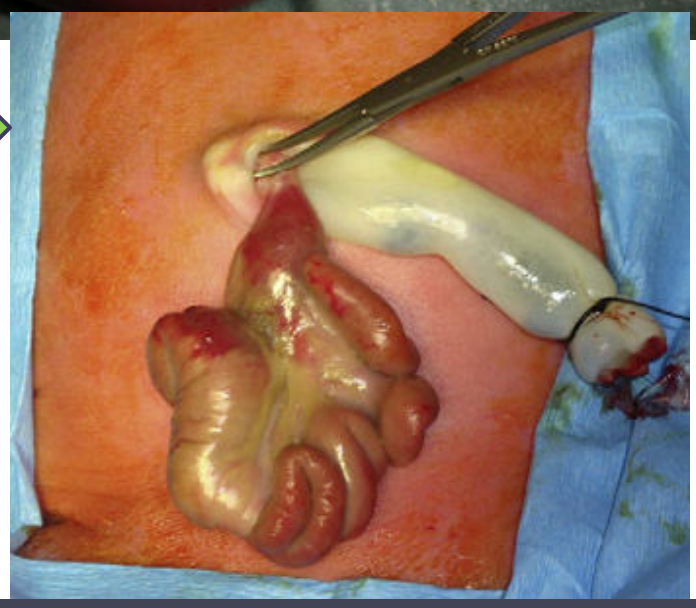


ASSOCIATED ANOMALIES

- 10-20% - intestinal stenosis or atresia that results from vascular insufficiency to the bowel.

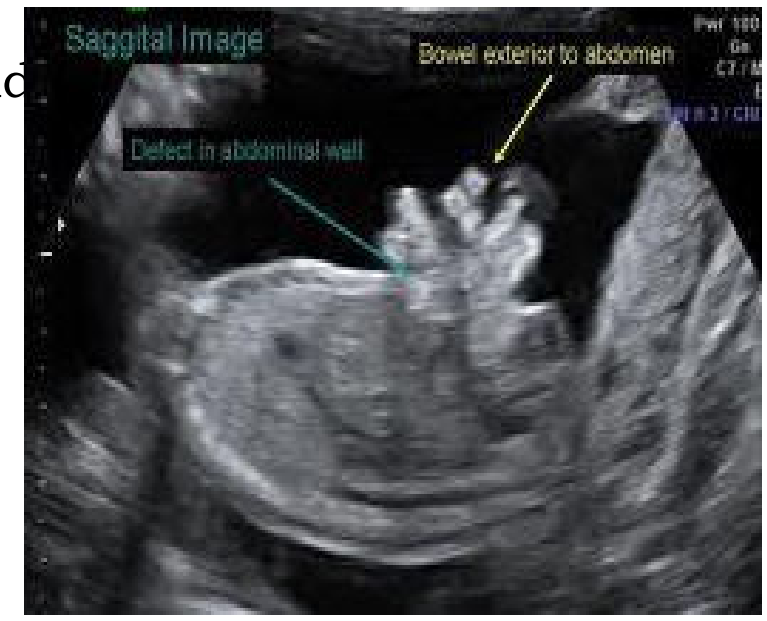


- 'Vanishing bowel'- very small defect strangulates bowel development.



ANTENATAL CONSIDERATIONS

- Diagnosis can often be **made < 20 weeks** of pregnancy by ultrasound.
- Amniotic fluid and serum tests of **AFP** and amniotic fluid **acetylcholinesterase (AChE)-** raised in abdominal wall defects.
- Opportunity to **counsel** the family (Increased risk :
  - Intrauterine growth retardation (IUGR),
  - Fetal death, and
  - Premature delivery).
- Prepare for optimal postnatal care.



Sensitivity 83% (18-100%)

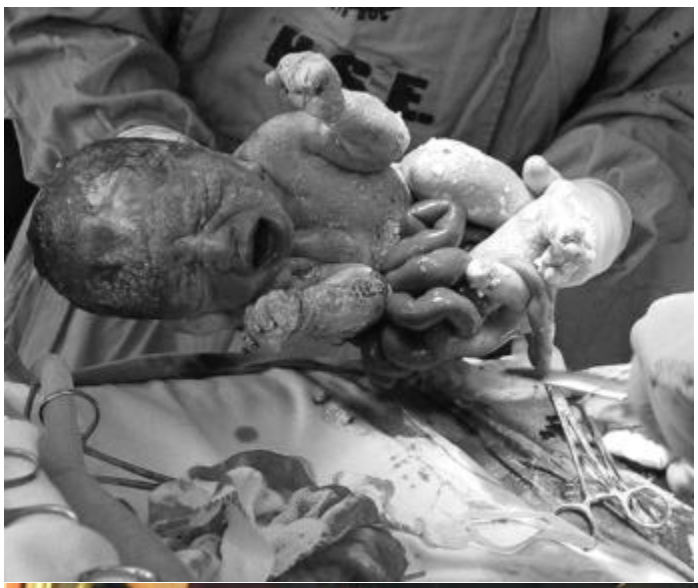
Mode of delivery.

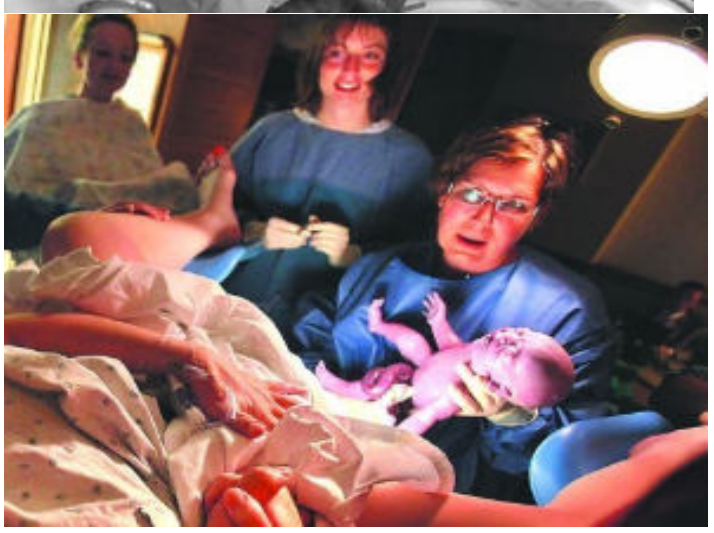
- **Optimal mode-** debated.

- Proponents of LSCS: Vaginal delivery may damage bowel.

- Studies have failed to show difference in outcome between Caesarean and vaginal delivery.

- The delivery method should be at the discretion of the obstetrician and the mother





Timing of delivery

- Considerations :

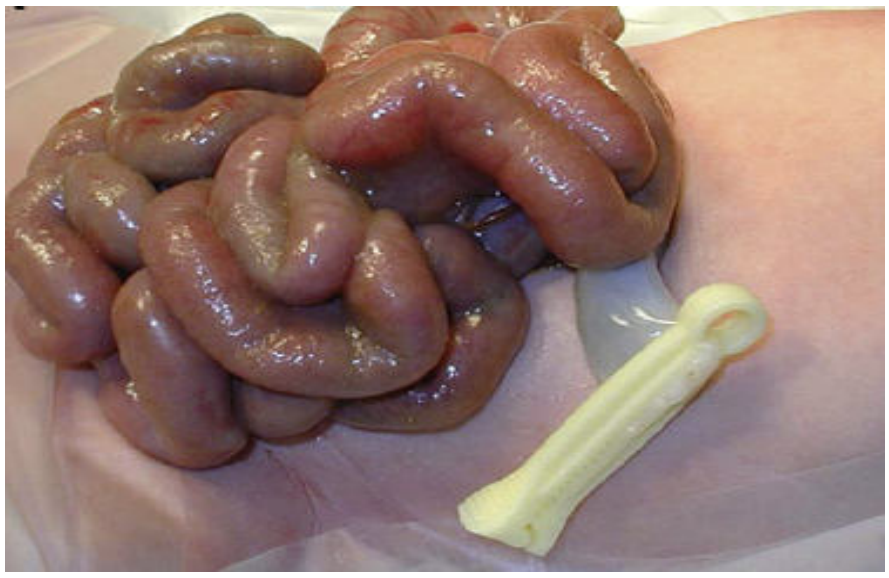
1. **Because bowel edema and peel formation** increase as pregnancy progresses.

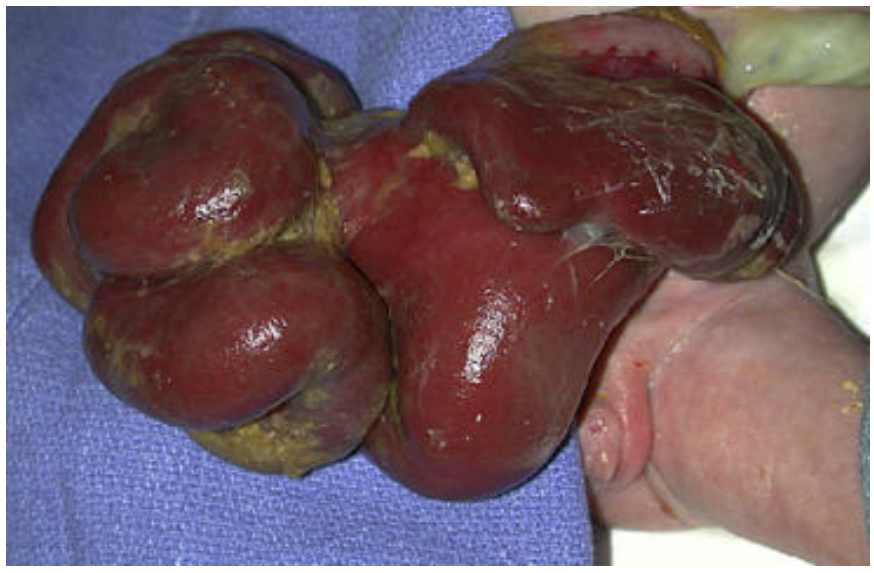
2. **LBW and preterm negatively influences outcome**, with neonates weighing <2 kg having
   - increased time to full enteral feeding,
   - ventilated days, and
   - duration of parenteral nutrition.

- The presumption is that **earlier delivery based on serial measurements of the bowel** may decrease the incidence of intestinal complications.

PERINATAL CARE

- **Outcome depends** on - amount of intestinal damage that occurs during fetal life.
- Combination of **exposure to amniotic fluid** and **constriction of the bowel** at the abdominal wall defect.
- Intestinal damage → impaired motility and mucosal absorptive function → prolonged need for total parenteral nutrition and severe irreversible intestinal failure.



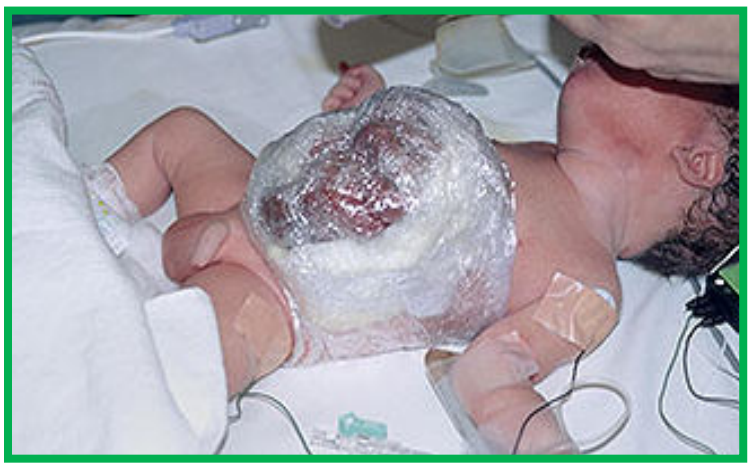


- Prenatal diagnosis provides a potential opportunity to modulate mode, location, and timing of delivery in order to minimize these complications.

Neonatal resuscitation and management

- Gastroschisis causes **significant evaporative water losses** from the exposed bowel.

1. Warm saline-soaked gauze, placed in a central position on the abdominal wall and wrapped with plastic wrap.
2. IV Fluid resuscitation.
3. Gastric decompression.
4. Baby right side down- prevent mesenteric pedicle kinking.
5. IV antibiotics.



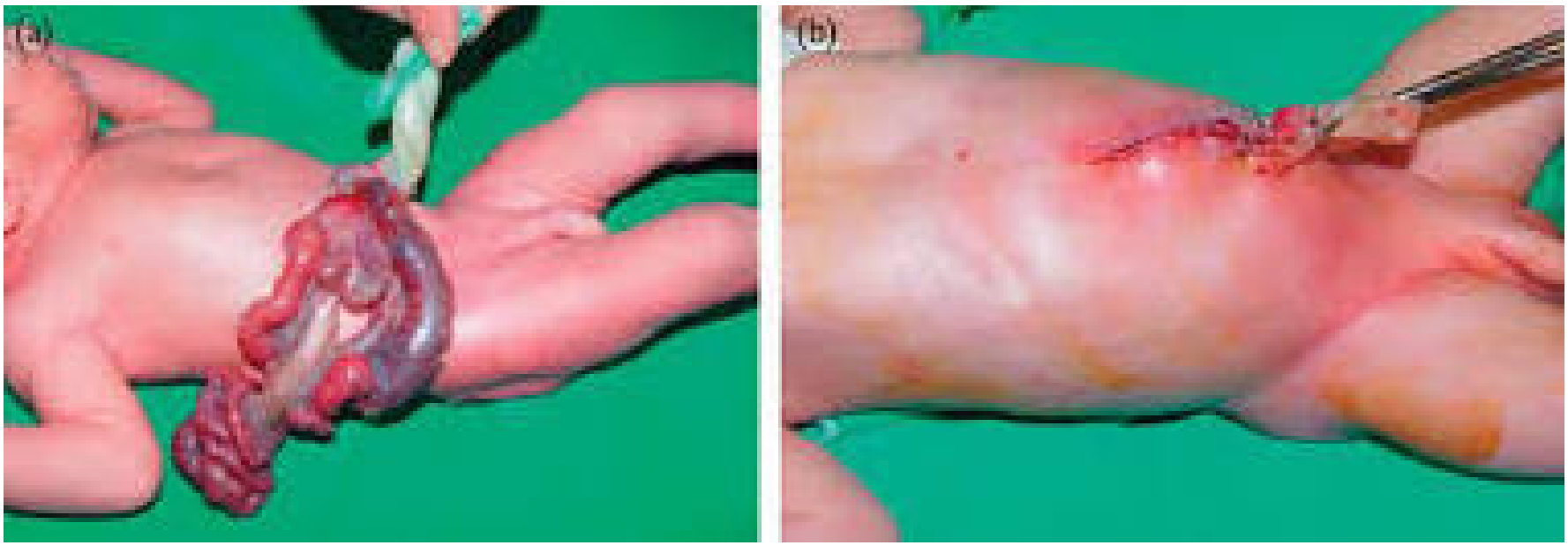
SURGICAL MANAGEMENT

- The primary goal of every surgical repair is to **return the viscera to the abdominal cavity while minimizing the risk of damage to the viscera.**

- Options include:
  (i) Primary reduction with operative closure of the fascia;
  (ii) silo placement, serial reductions, and delayed fascial closure;

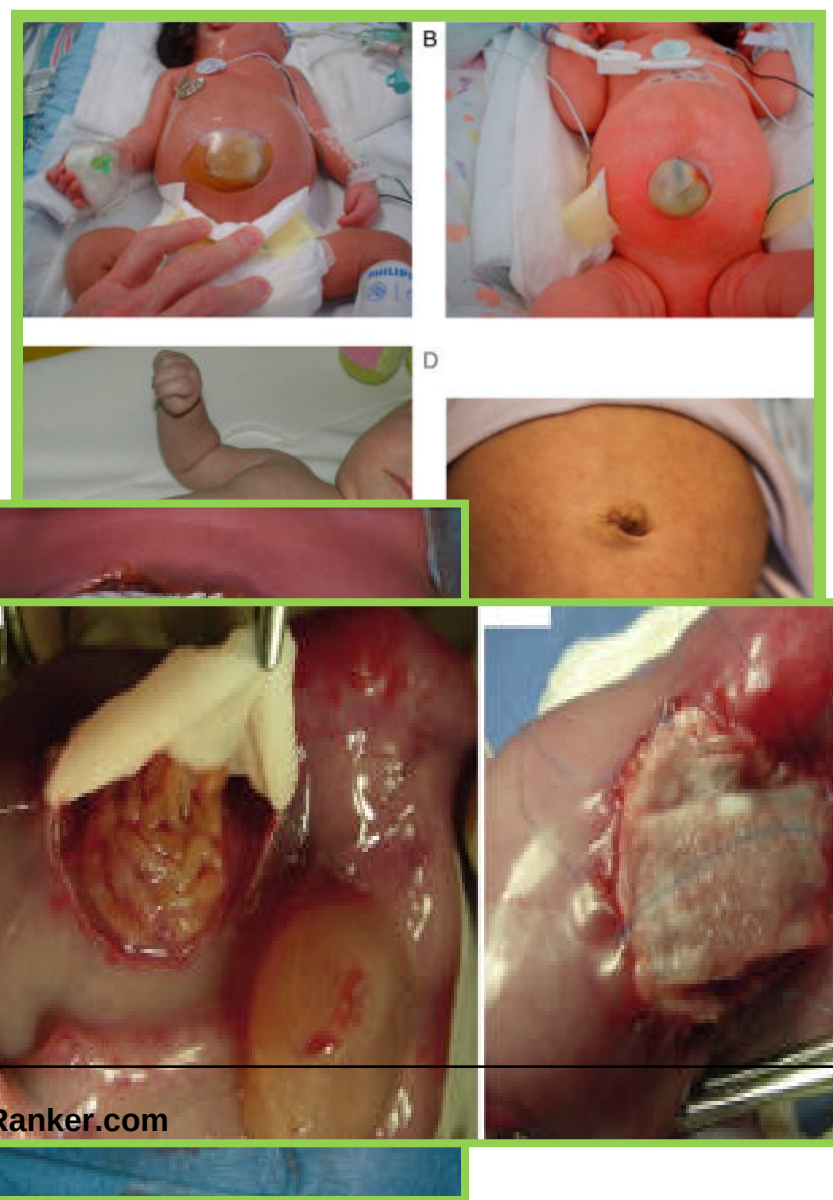
Primary closure – with fascial closure

- In neonates considered to possess sufficient intraabdominal domain to permit full reduction of the herniated viscera.
- Warm bowel and clean the peel; check quickly for intestinal anomalies.



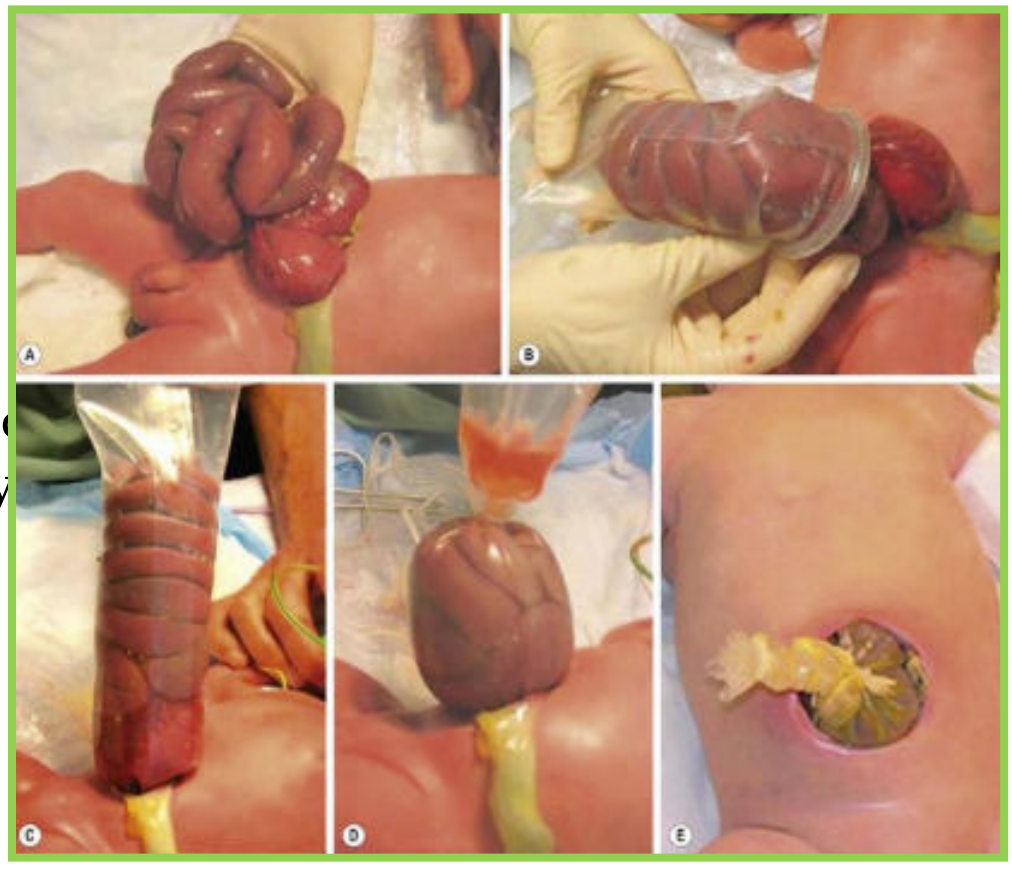
Primary closure- without fascial closure

- Umbilicus as an allograft.
- Prosthetic non absorbable mesh.
- Prosthetic biosynthetic absorbable options – dura or porcine small intestinal submucosa.



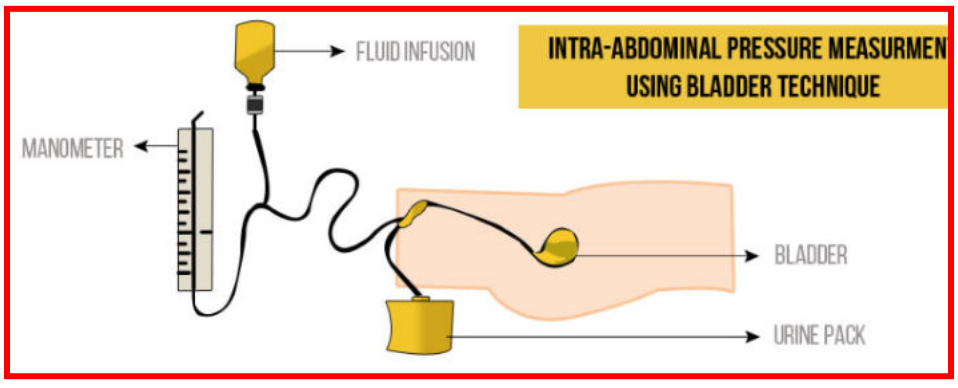
Staged closure

- Bowel placed into
  – Spring loaded silo
  - Silastic sheet silo
- Delivery room or OT.
- Bowel is reduced once or twice daily into the abdominal cavity as the silo is shortened by sequential ligation.
- Once contents entirely reduced, definitive closure.
- Usually takes 1-14 days.

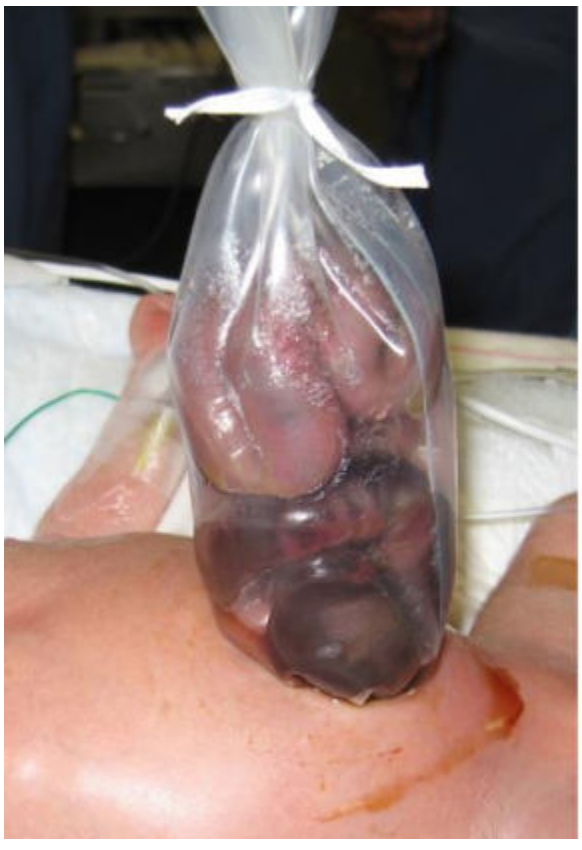


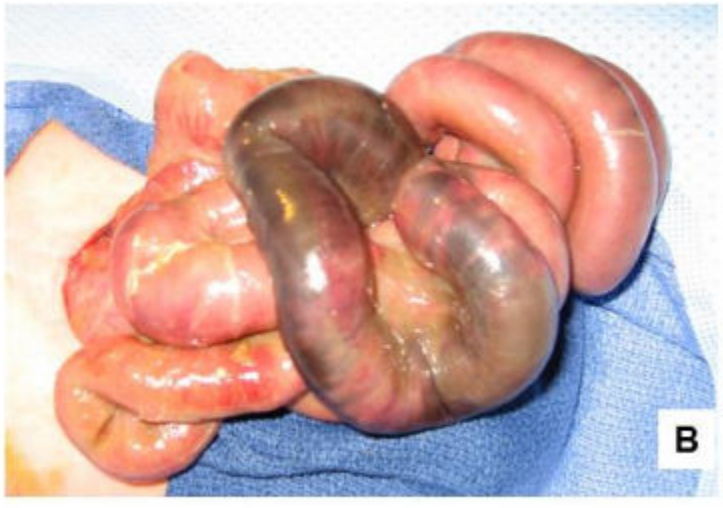
Intra-abdominal pressure

- Either as intravesical or intragastric pressure, can be used to guide the surgeon during reduction.
- Pressures >20 mmHg are correlated with decreased perfusion to the kidneys and bowel.
- Following reduction, monitor:
  - Physical examination,
  - Urine output, and
  - lower limb perfusion



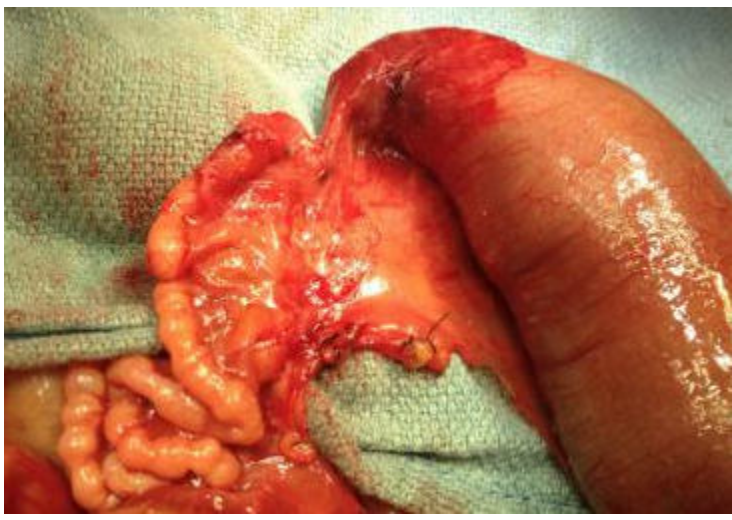
With a low threshold to reopen a closed abdomen for signs of abdominal compartment syndrome



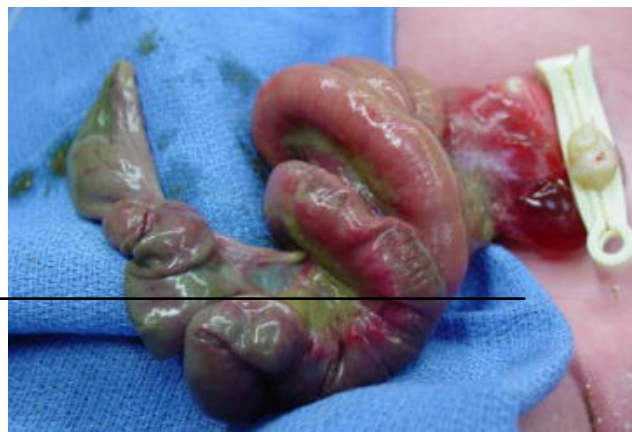


- Gangrenous intestinal loop within the silo.

Management of associated intestinal atresia or perforation



- Upto 10 % cases associated.
- Usually jejunal and ileal.
- Options
  - Resection and end to end anastomosis
  - Stoma
  - Initial gastroschisis repair and 4-5 weeks later, atresia surgery.



Postoperative Course

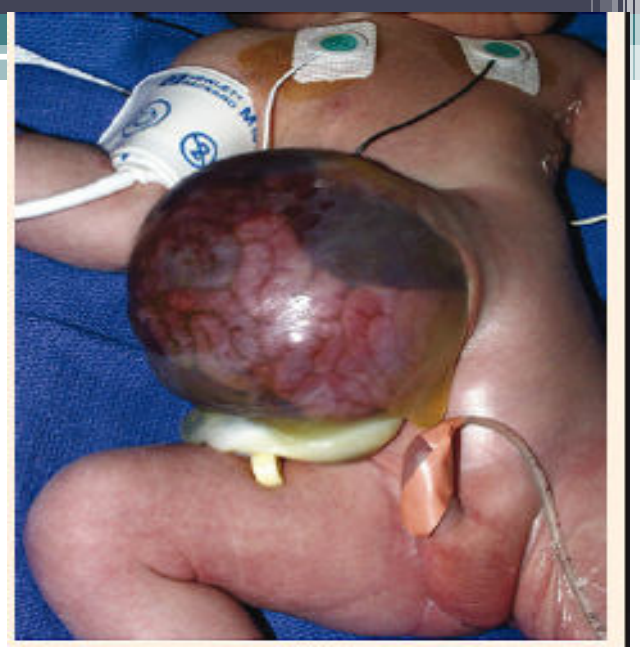
- Abnormal intestinal motility.
- Abnormal nutrient absorption.

- Delayed enteral feeding.
- Prokinetics.
- Parenteral nutrition.

OMPHALOCELE- 2nd Most common

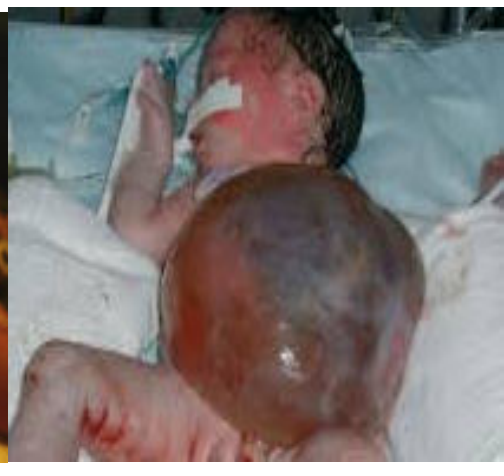
- Incidence is 1.5 to 3 per 10,000 live births.
- Omphalocele represents a failure of the body folds to complete their journey.
- Herniated viscera covered by a membrane consisting of peritoneum on the inner surface, amnion on the outer surface, and Wharton's jelly between the layers.

OMPHALOCELE (EXOMPHALOS)



- The umbilical vessels insert into the membrane and not the body wall.
- The hernia contents include a variable amount of intestine, often parts of the liver, and occasionally other organs.

	E. Minor	E. Major
Defect	< 5 cm	> 5 cm
Content	Intestine	Viscera + liver



OMPHALOCELE (EXOMPHALOS)

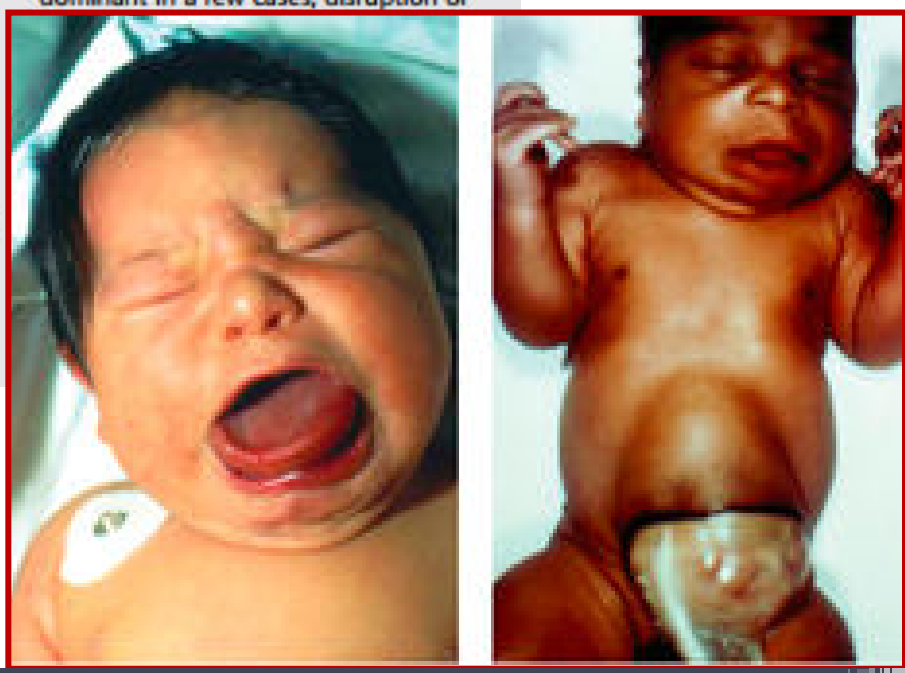
- Whatever the insult may be that causes it, this aberration occurs **early in embryogenesis-** more associated anomalies.

Associated Conditions

System	*Gastroschisis (%)*	*Omphalocele (%)*
Cardiac	2-12	7-47
Respiratory	<1	1-4
Central nervous system	2-10	4-30
Musculoskeletal	<1-10	4-25
Gastrointestinal	5-40	3-20
Genitourinary	3-10	6-20
Facial	1-3	1-14
Chromosomal	<1-3	3-20

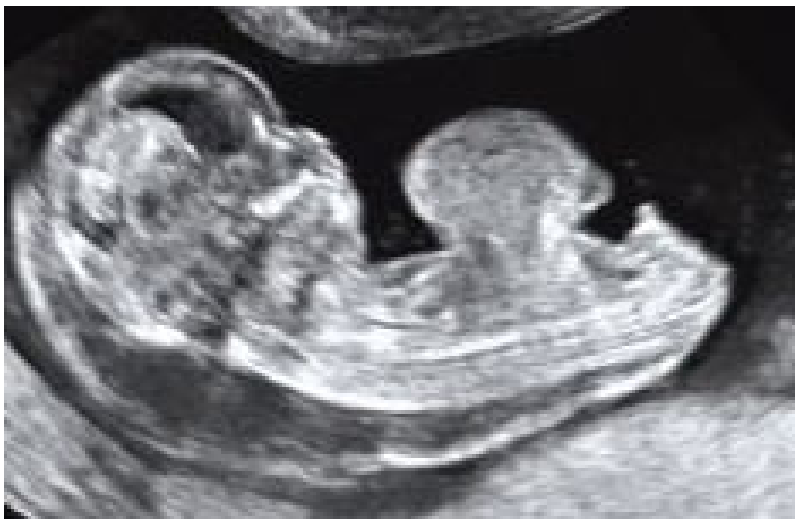
Omphalocele Syndromes

Name	Description	Inheritance
Shprintzen omphalocele syndrome	Malformation syndrome that includes mildly dysmorphic facies, omphalocele, scoliosis, learning disabilities, and pharyngeal and laryngeal hypoplasia	Autosomal dominant
Omphalocele–cleft palate syndrome	Lethal syndrome associated with uterus bicornis in one case, uvula duplex and hydrocephalus internus in another, omphalocele, and cleft palate	
Beckwith-Wiedemann syndrome (also known as exomphalos-macroglossia-gigantism syndrome [EMG syndrome] and Wiedemann-Beckwith syndrome [WBS])	Pediatric overgrowth disorder involving a predisposition to tumor development. The clinical presentation is highly variable; some cases lack the hallmark features of exomphalos, macroglossia, and gigantism. Abdominal wall defects common, as well as visceromegaly including liver, spleen, pancreas, kidneys, and adrenals	Inheritance of BWS is complex. Possible patterns include autosomal dominant inheritance with variable expressivity, contiguous gene duplication at 11p15, and genomic imprinting resulting from a defective or absent copy of the maternally derived gene
Gershoni-Baruch syndrome	Large/giant omphalocele containing liver and intestines. Also associated with diaphragmatic hernia and radial ray defects	Autosomal recessive hypothesis in one case
C syndrome (Opitz trigonocephaly syndrome, Trigonocephaly syndrome)	Unusual facies, polydactyly, cardiac abnormality, large omphalocele in a few cases	Autosomal recessive mostly, autosomal dominant in a few cases; disruption of
Donnai-Barrow syndrome (Facio oculoacousticorenal syndrome)	Facial anomalies, ocular anomalies, sensorineural hearing loss, and proteinuria. Some cases include omphalocele as an associated anomaly	
Thoracoabdominal syndrome (THAS)	Diaphragmatic and ventral hernias, hypoplastic lung, cardiac anomalies, cleft palate, omphalocele, sporadic pentalogy of Cantrell	
Manitoba oculotrichoanal syndrome (MOTA) (Marles syndrome)	Hypertelorism, unilateral eye malformations, aberrant anterolateral scalp hairline, nasal and anal anomalies. Omphalocele noted	



ANTENATAL CONSIDERATIONS

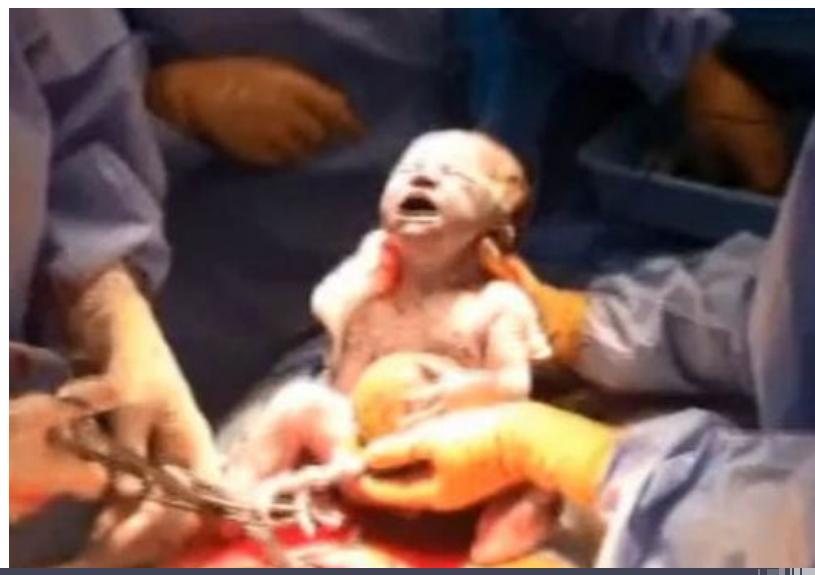
- Distinguished by presence of sac and presence of liver.
- Other associated anomalies- ultrasound especially for cardiac and chromosomal studies.
- Increased levels of AFP and AChE
- Risks of :
  - IUGR (5-35%)
  - Fetal death
  - Premature labour (5-60%)



Sensitivity 75%(25-100%)

PERINATAL CARE

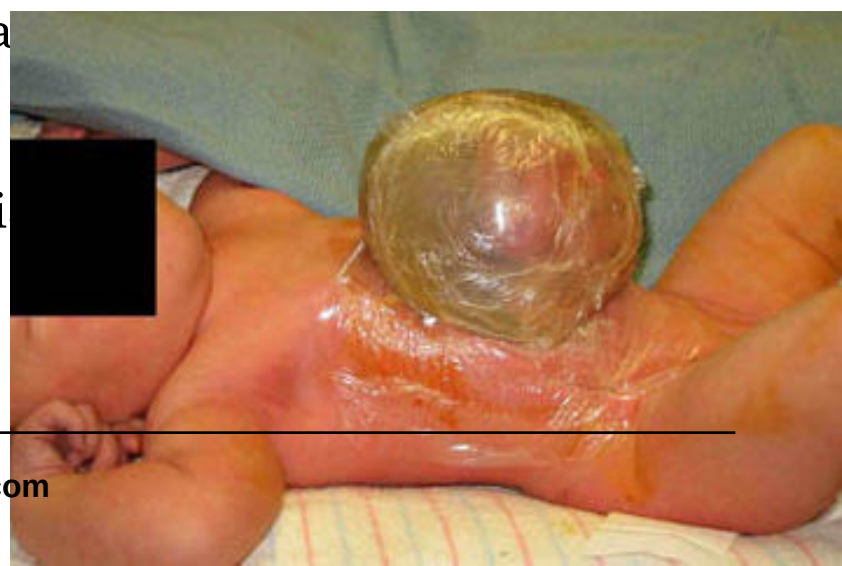
- Neither caesarean nor vaginal delivery superior.
- Most practitioners choose to deliver neonates with large omphaloceles by cesarean section because of the fear of liver injury or sac rupture during vaginal delivery.
- Delivery at tertiary perinatal centre- immediate access to expert care.
- No advantage of preterm delivery.



NEONATAL RESUSCITATION AND MANAGEMENT

- Careful attention to **cardiopulmonary status-** unsuspected pulmonary hypoplasia- requires immediate intubation and ventilation.

- Directed **cardiac evaluation**:
  - auscultation,
  - four-limb blood pressures, and
  - peripheral pulse examination.
- **Dressed** with saline soaked gauze and a
  minimize fluid and temperature losses.
- If sac ruptured, then treat as gastroschi
- **IV fluids and nasogastric tube**.



SURGICAL MANAGEMENT

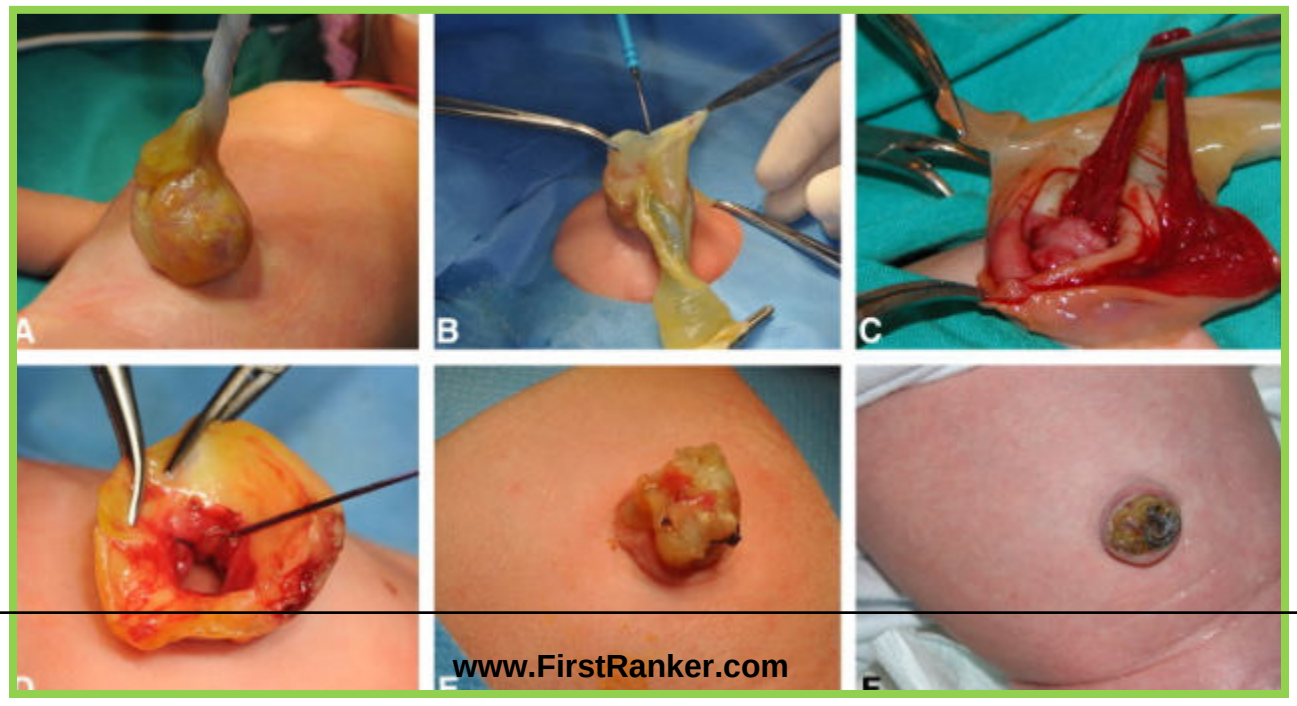
- Treatment options in infants with omphalocele depend on:
- The size of the defect,
- gestational age, and
- the presence of associated anomalies.

- Options:

1. Primary closure
2. Staged closure

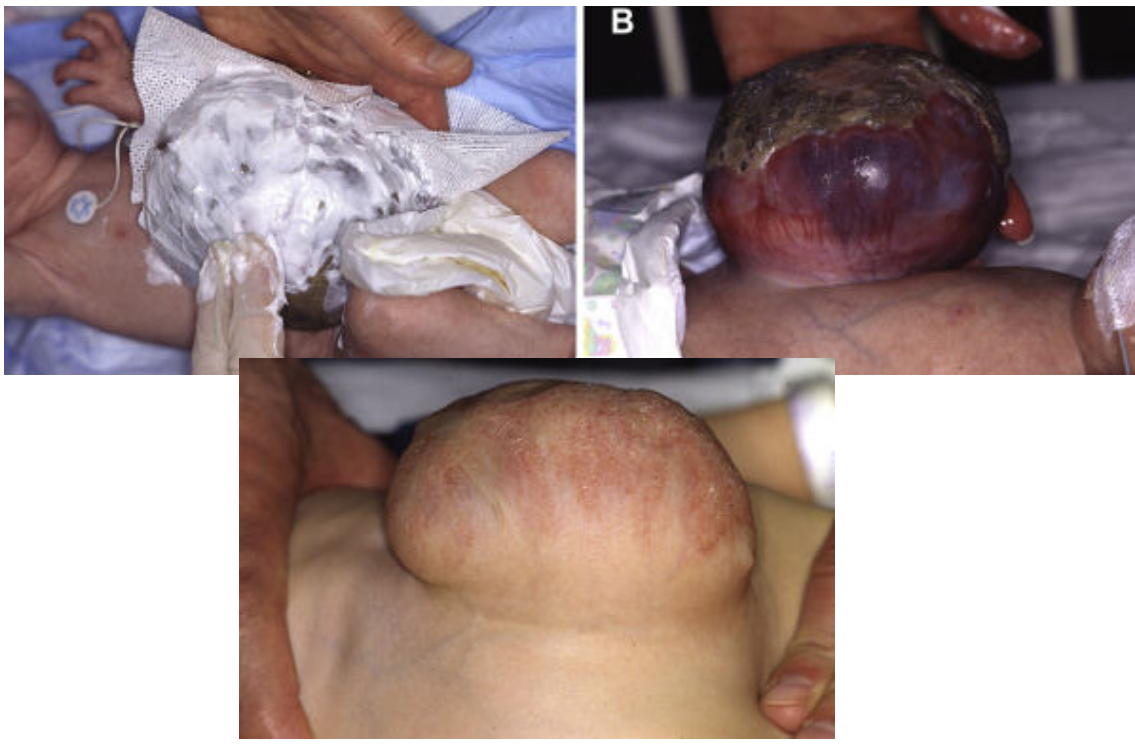
PRIMARY CLOSURE

- Only when the **baby is stable and defect is small.**
- Steps:
- Excising the omphalocele membrane,
- reducing the herniated viscera, and
- closing the fascia and skin.

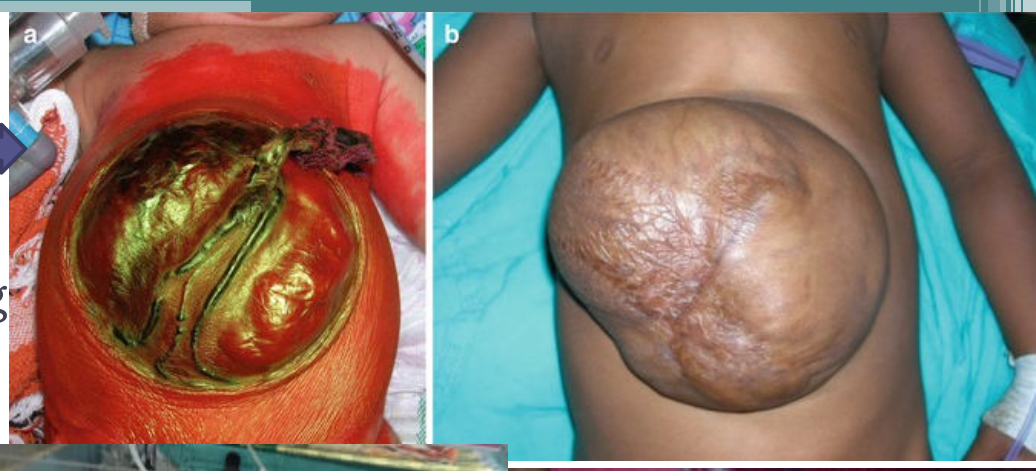


STAGED CLOSURE

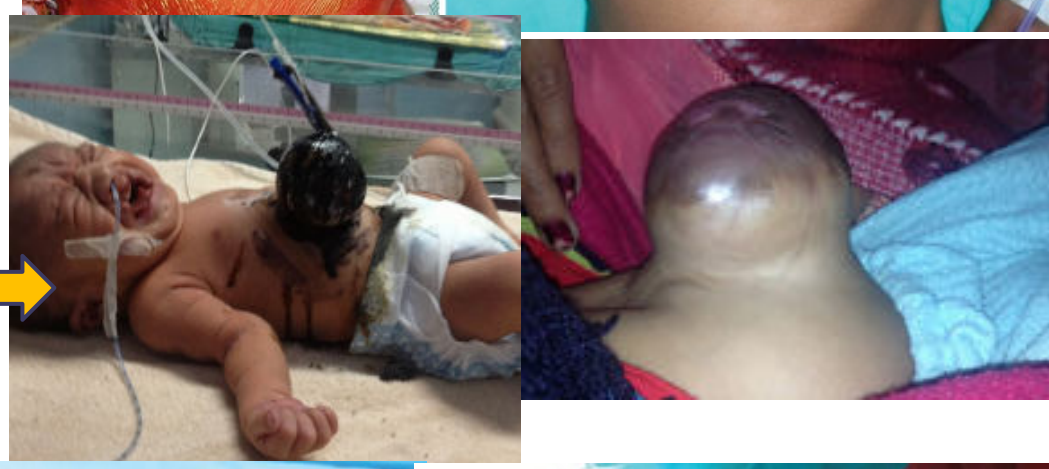
- If the covering sac is intact, then there **is no urgency to perform operative closure.**
- **‘Escharotic therapy’,** which results in gradual epithelialization of the omphalocele sac.
- Usually takes many months for the sac to granulate and epithelialize.
- Options:
  1. Silver sulfadiazine
  2. Mercurochrome
  3. Povidone iodine
  4. Gentian violet



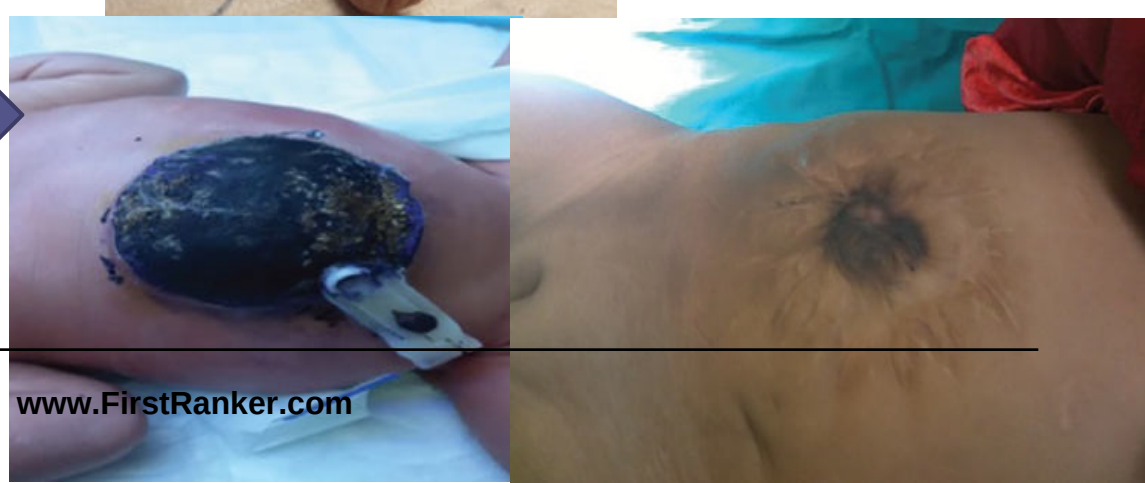
- Mercurochrome
  - scarificant and disinfectant.
  - reports of mercury poisoning



- Povidone iodine - systemic absorption of the iodine- transient hypothyroidism.



- Gentian violet – Antibacterial and antifungal.



STAGED CLOSURE

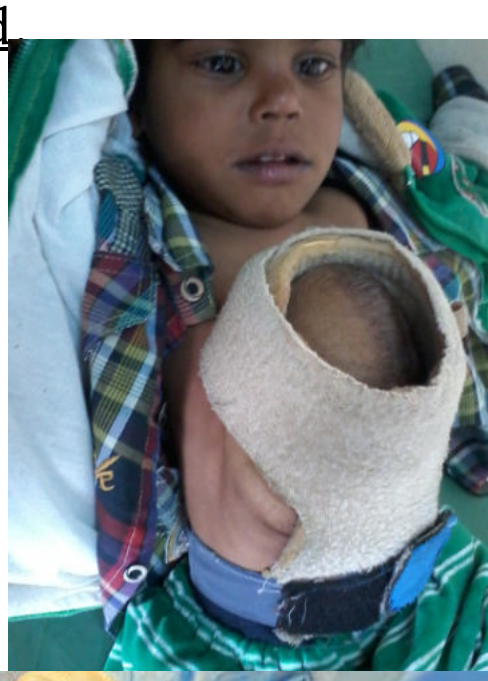
- Sac is epithelialized or sturdy enough to withstand external pressure

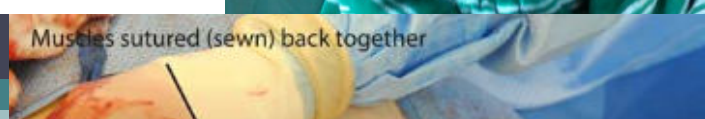
↓

Compression is done with elastic bandages and serially increased until the abdominal contents are reduced

↓

VENTRAL HERNIA REPAIR

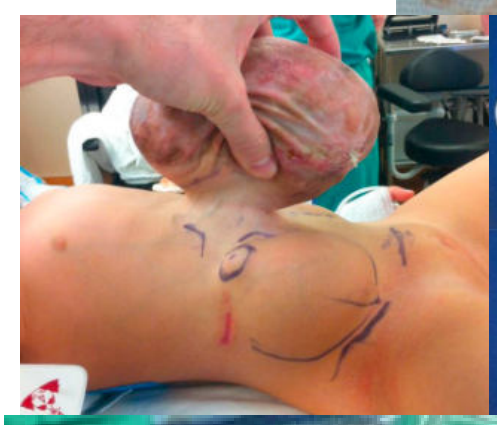


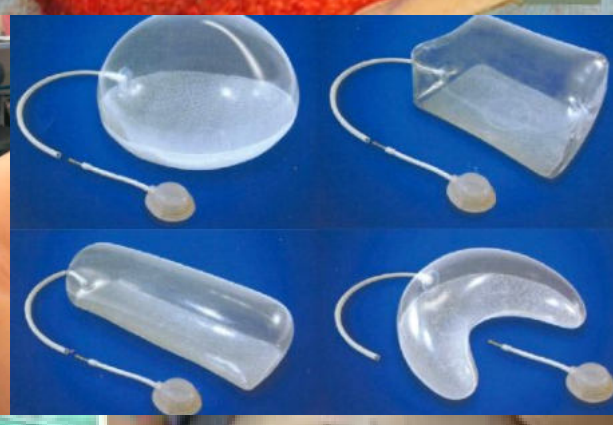


VENTRAL HERNIA REPAIR

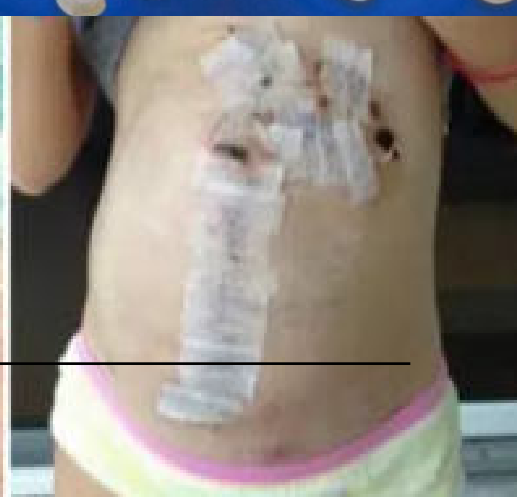
1. **Flaps** that mobilize the muscle, fascia, and skin of the abdominal wall toward the midline and allow midline fascial closure.

2. Tissue expanders-to create an abdominal cavity big enough to house the viscera.



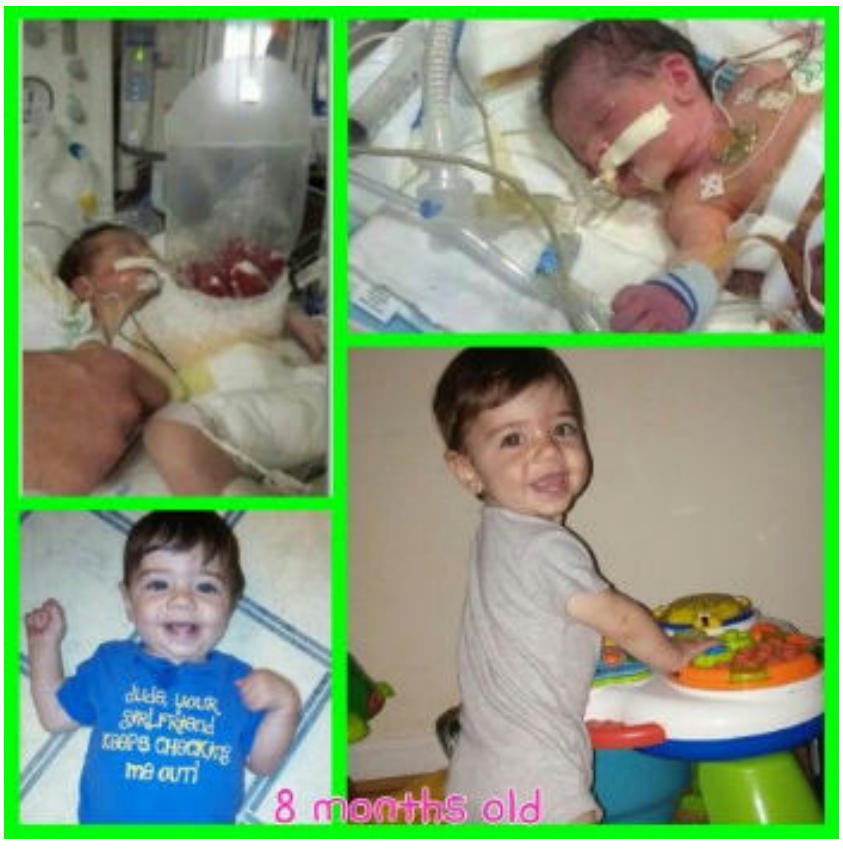


3. Prosthetic patches in abdominal wall.



Long-term outcomes

GASTROSCHISIS



- Generally excellent.
- Many patients with atresia do very well as long as the bowel is not irreversibly damaged during fetal life.
- Majority - will achieve normal growth and development after an initial catch-up period in early childhood.

Long-term outcomes

OMPHALOCELE

- Most infants recover well with no long term issues, provided that there are no significant structural or chromosomal abnormalities.
- Long term medical problems occur in patients with large omphaloceles:
  - gastroesophageal reflux,
  - pulmonary insufficiency,
  - recurrent lung infections or asthma, and
  - feeding difficulty with failure to thrive,

  reported in up to 60% of infants with a giant omphalocele.

	OMPHALOCELE	GASTROSCHISIS
INCIDENCE	1.5-3: 10,000	2 -4.9: 10,000
SAC	Present	Absent
ASSOCIATED ANOMALIES	Common	Uncommon
DEFECT	At umbilicus; 1-15 cm	Right of umbilicus; <4cm
MATERNAL AGE	Average	Younger
SURGICAL MANAGEMENT	Non urgent	Urgent
PROGNOSTIC FACTORS	Associated anomalies	Bowel condition
MORTALITY	<5%