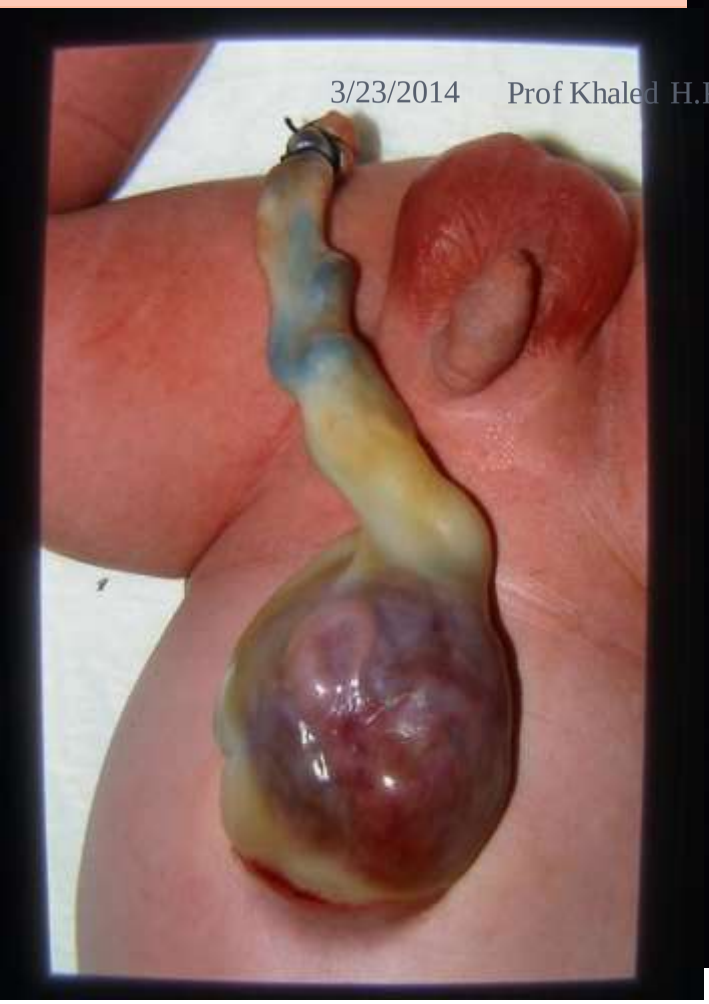
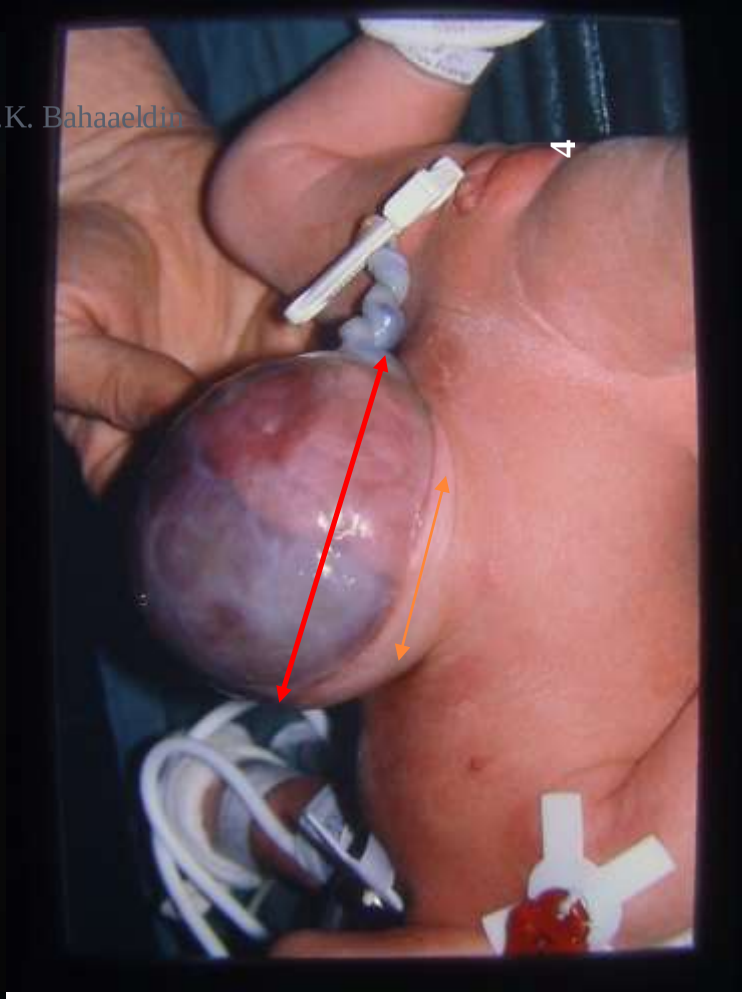
Abdominal Wall Defects & Malrotation

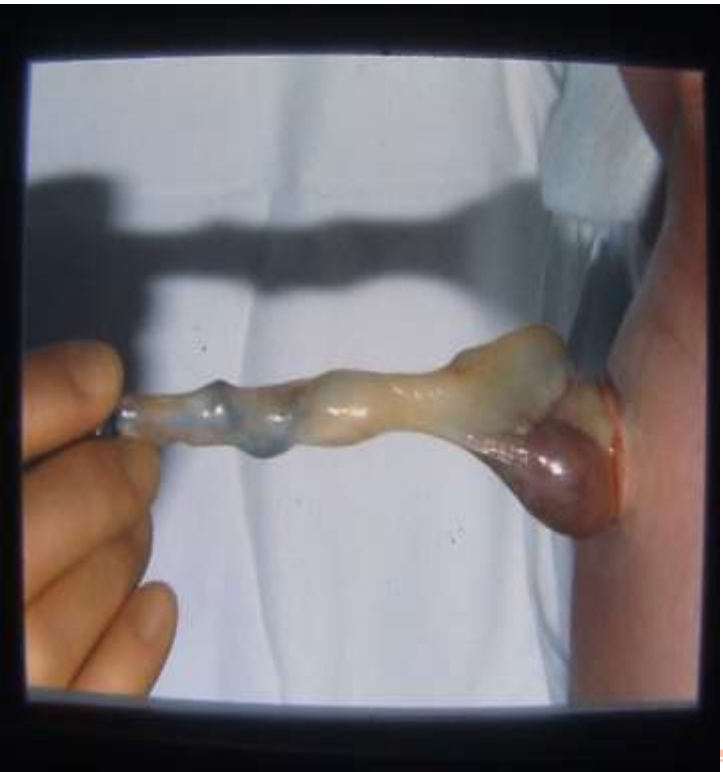
OMPHALOCOELE

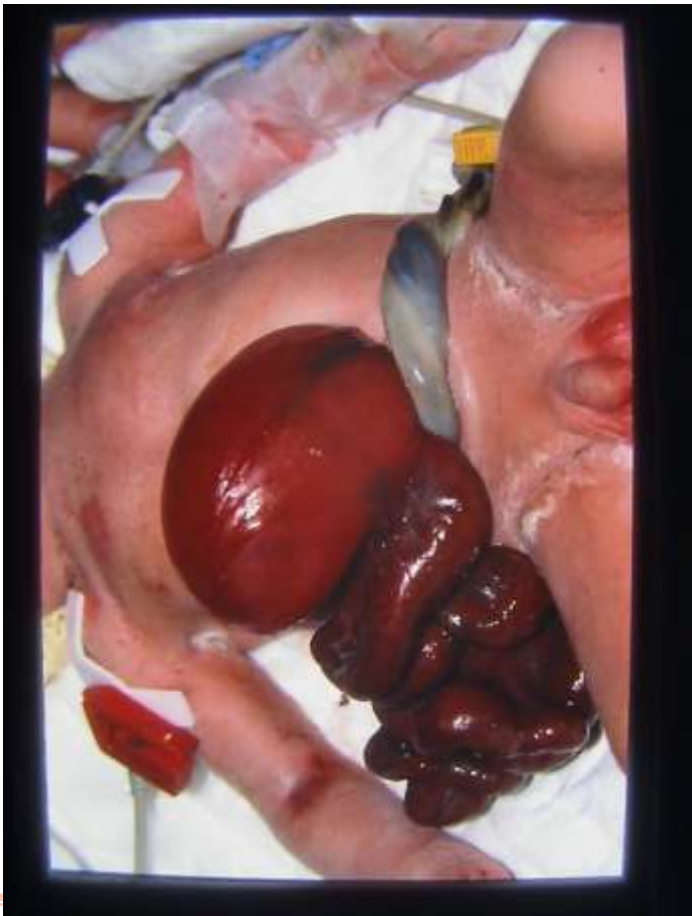
- It is a defect in abdominal wall musculature and skin with protrusion of abdominal viscera contained within a membranous sac.

- **Exomphalos major** (umbilical defect >5cm)
- **Exomphalos minor** (umbilical defect<5cm)



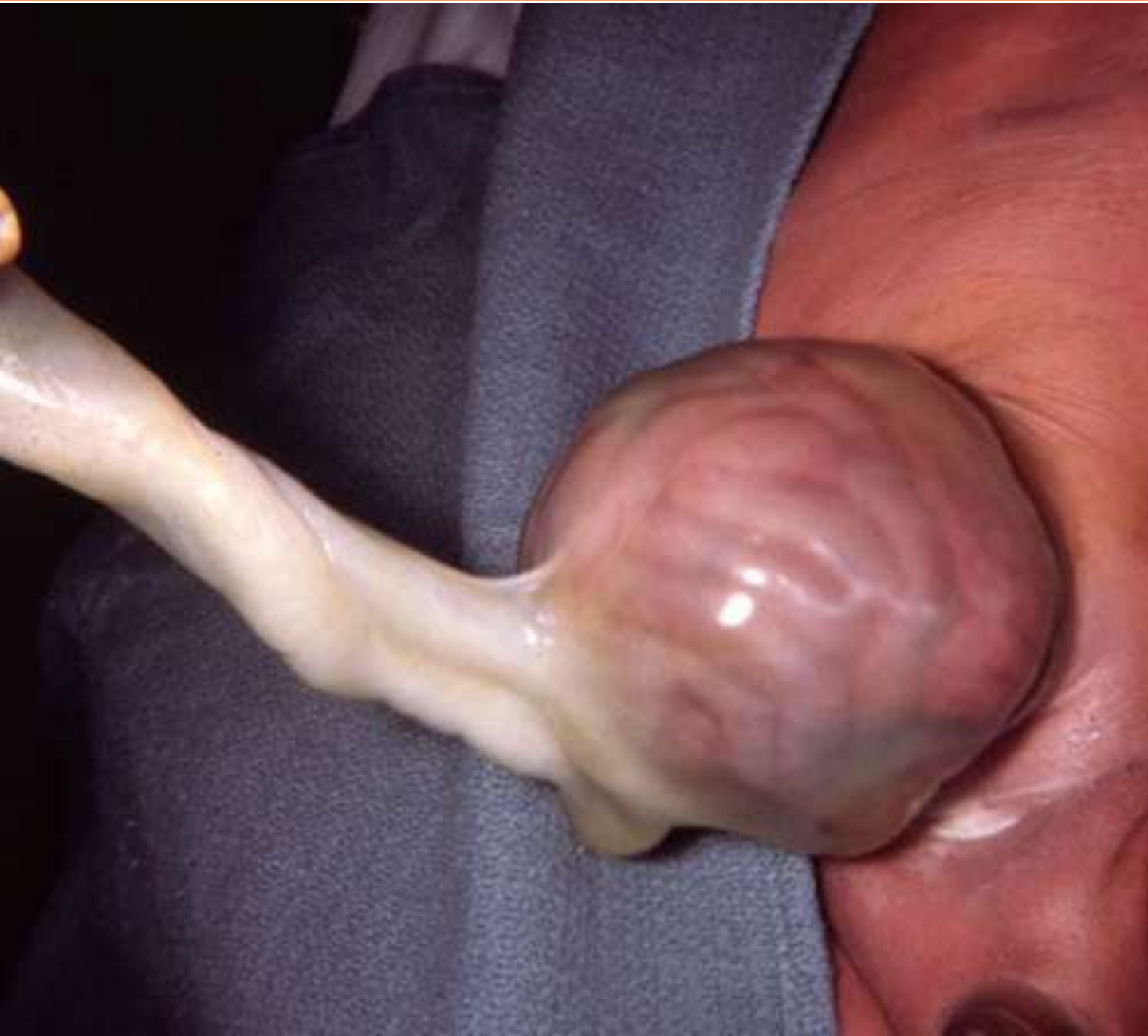






Abdominal wall defect

OMPHALOCELE

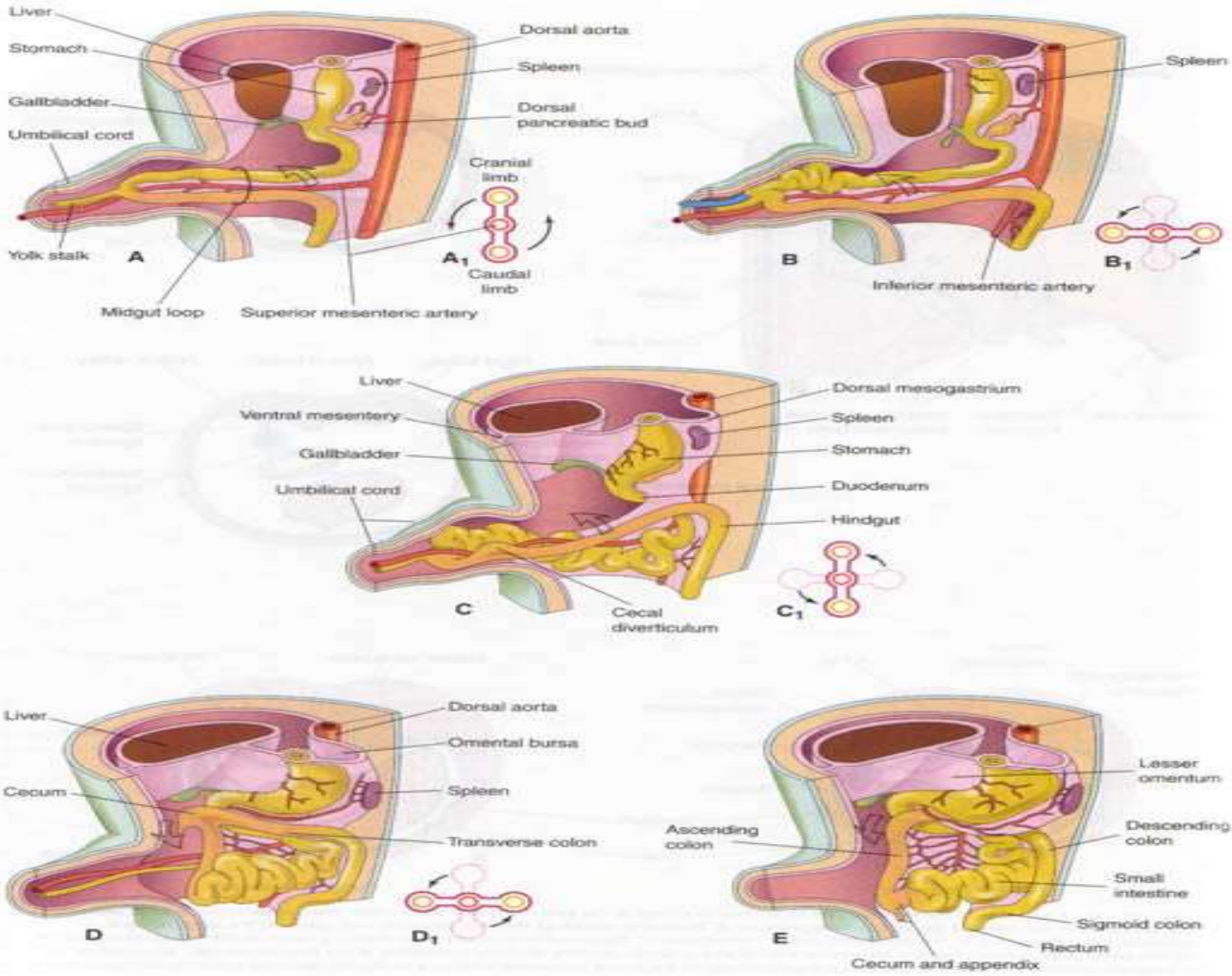


Incidence

- Small omphalocoele 1:5000
- Large omphalocoele 1:10000
- Male to female ratio 1:1
- Pacific Islanders have low risk for omphalocoele

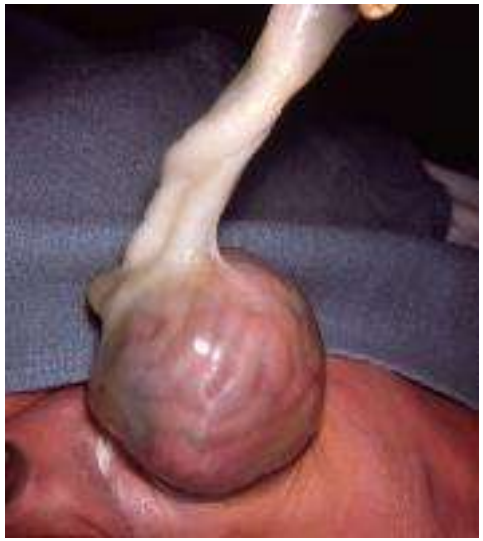
Pathophysiology

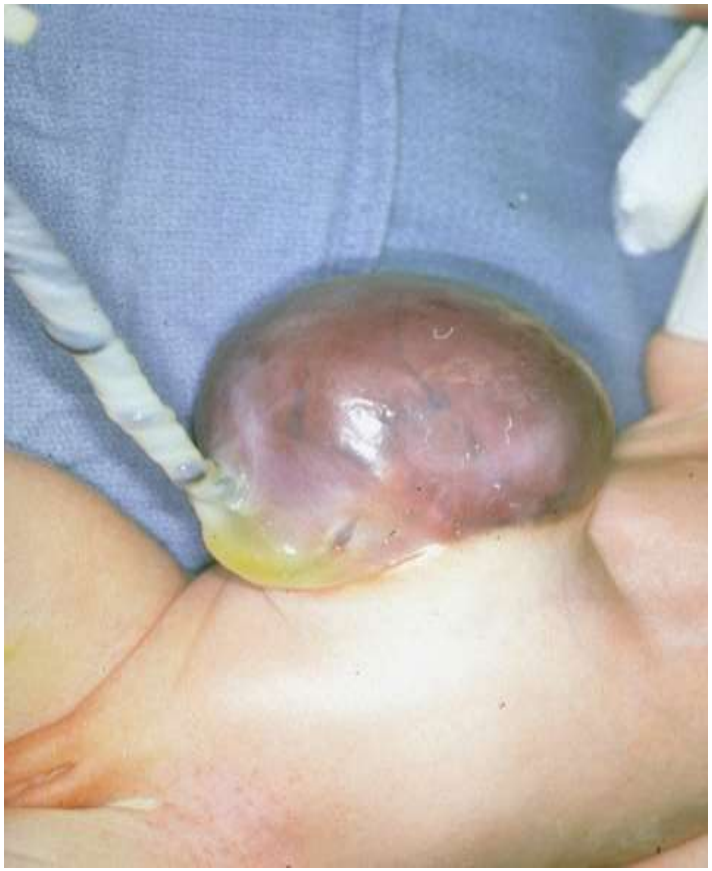
- Failure of the midgut to return to abdomen by the 10^{th} week of gestation


Liver
Stomach
Gallbladder
Umbilical cord
Yolk stalk
A
Dorsal aorta
Spleen
Dorsal pancreatic bud
Cranial limb
A1
Caudal limb
Midgut loop
Superior mesenteric artery
Spleen
B
B1
Inferior mesenteric artery
Liver
Ventral mesentery
Gallbladder
Umbilical cord
C
Cecal diverticulum
Dorsal mesogastrium
Spleen
Stomach
Duodenum
Hindgut
C1
Liver
Cecum
D
Dorsal aorta
Omental bursa
Spleen
Transverse colon
D1
Lesser omentum
Ascending colon
Descending colon
Small intestine
Sigmoid colon
Rectum
E
Cecum and appendix

Clinical Findings

- Covered clinical defect of the umbilical ring
- Defect may vary from 2-10 cm
- Sac is composed of amnion, Wharton's jelly and peritoneum





- 50% have accompanying liver, spleen, testes/ovary
- >50% have associated defects
- Location:
  - Epigastric
  - Central
  - Hypogastric
- Cord attachment is on the sac

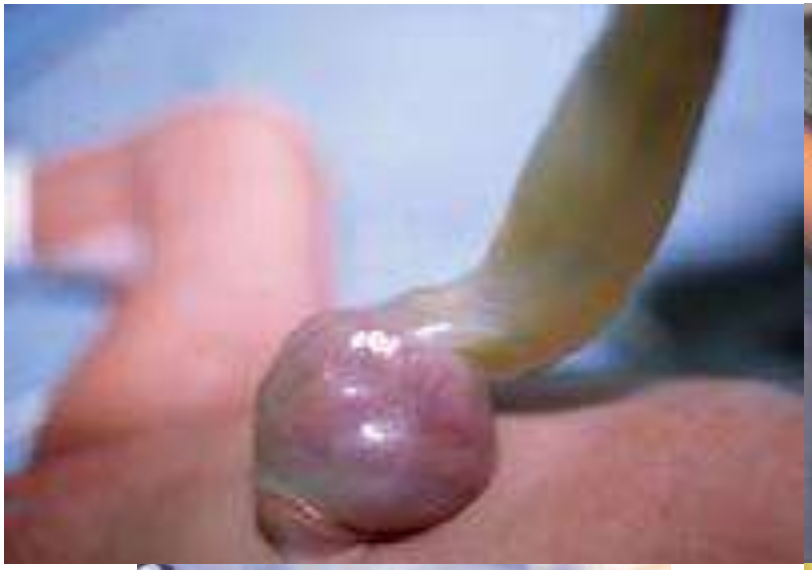
- The sac may rupture in utero in 10-18% or from the delivery process in4%.
- The incidence of associated major congenital anomalies in up to 81%.

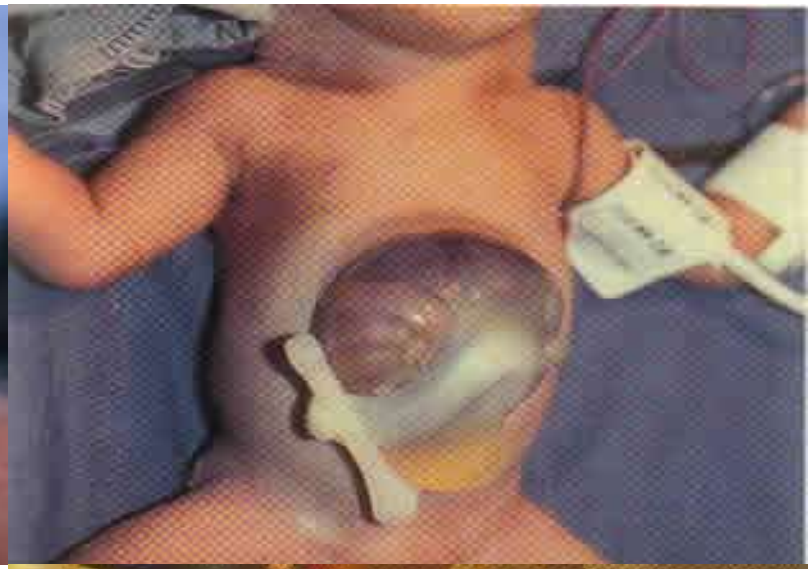
Defects of cranial fold

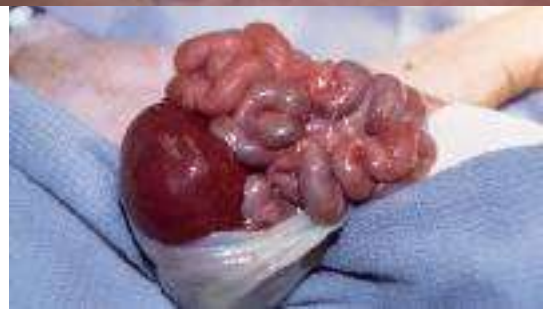
- congenital heart disease
- diaphragmatic hernia
- ectopia cordis
- sternal cleft,

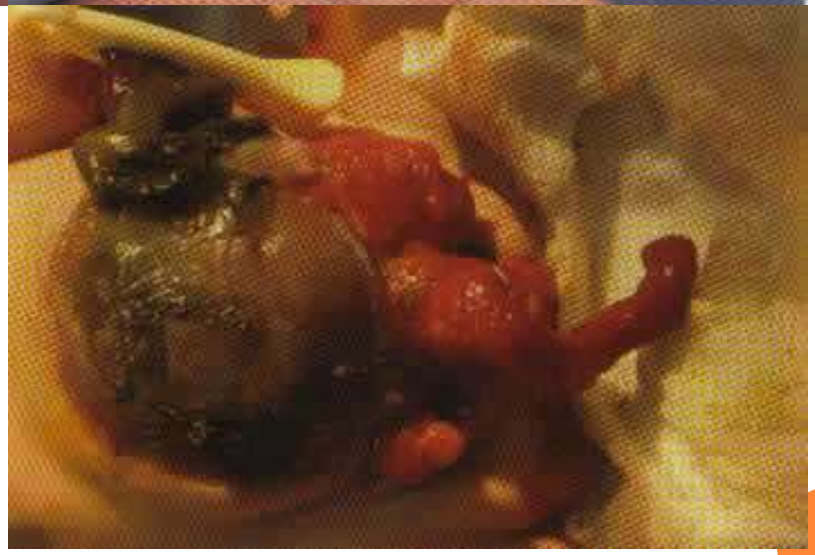
Defects of caudad fold

- Imperforate anus
- Genitourinary malformations
- Bladder or cloacal exstrophy
- Colon atresia
- Sacral and vertebral anomalies, and
- Meningomyelocele.



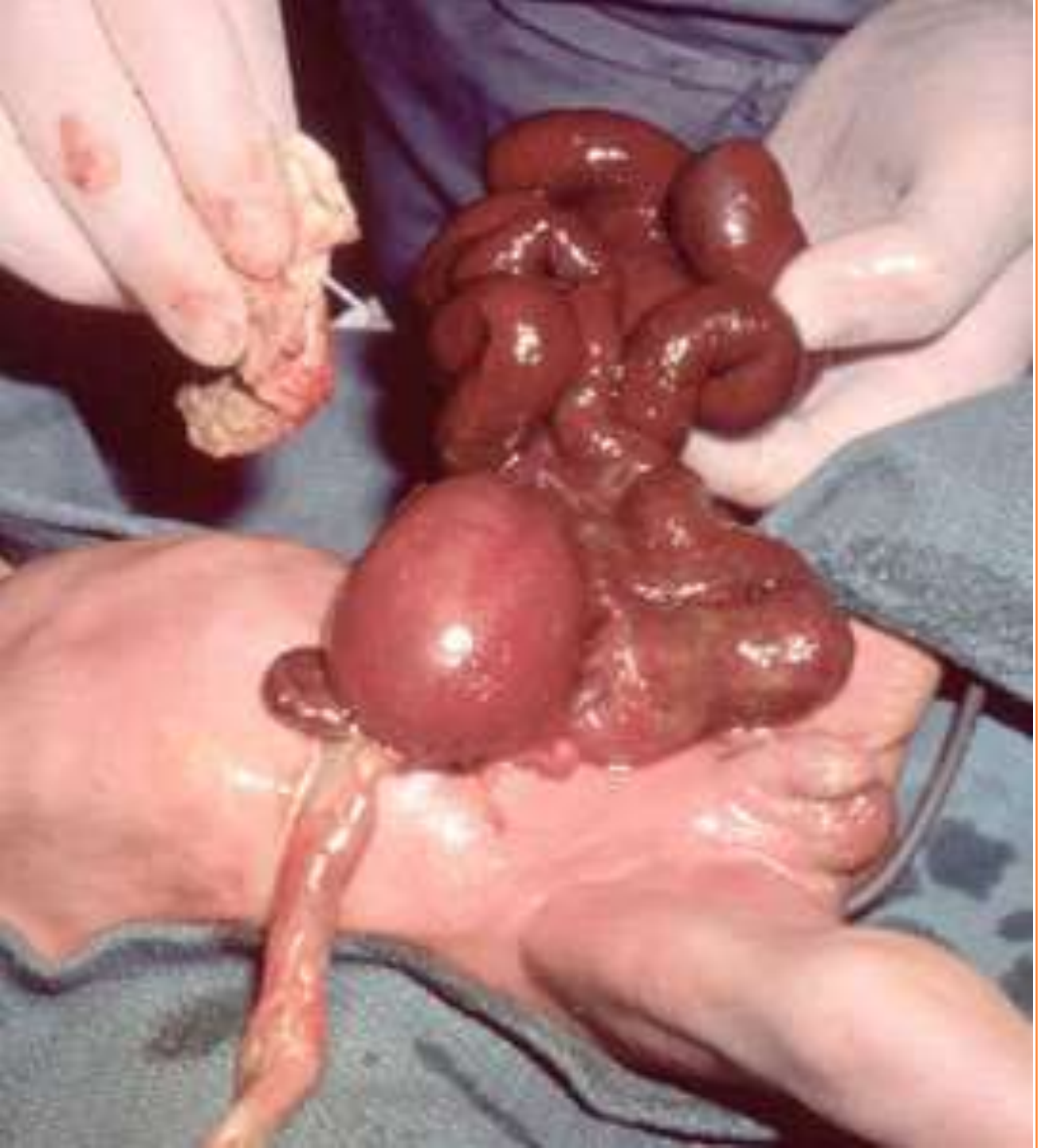






GASTROSCHISIS

- It is the defect in the abdominal wall was displaced to the right of the umbilicus and eviscerated bowel was not covered by a membrane.



Gastroschisis

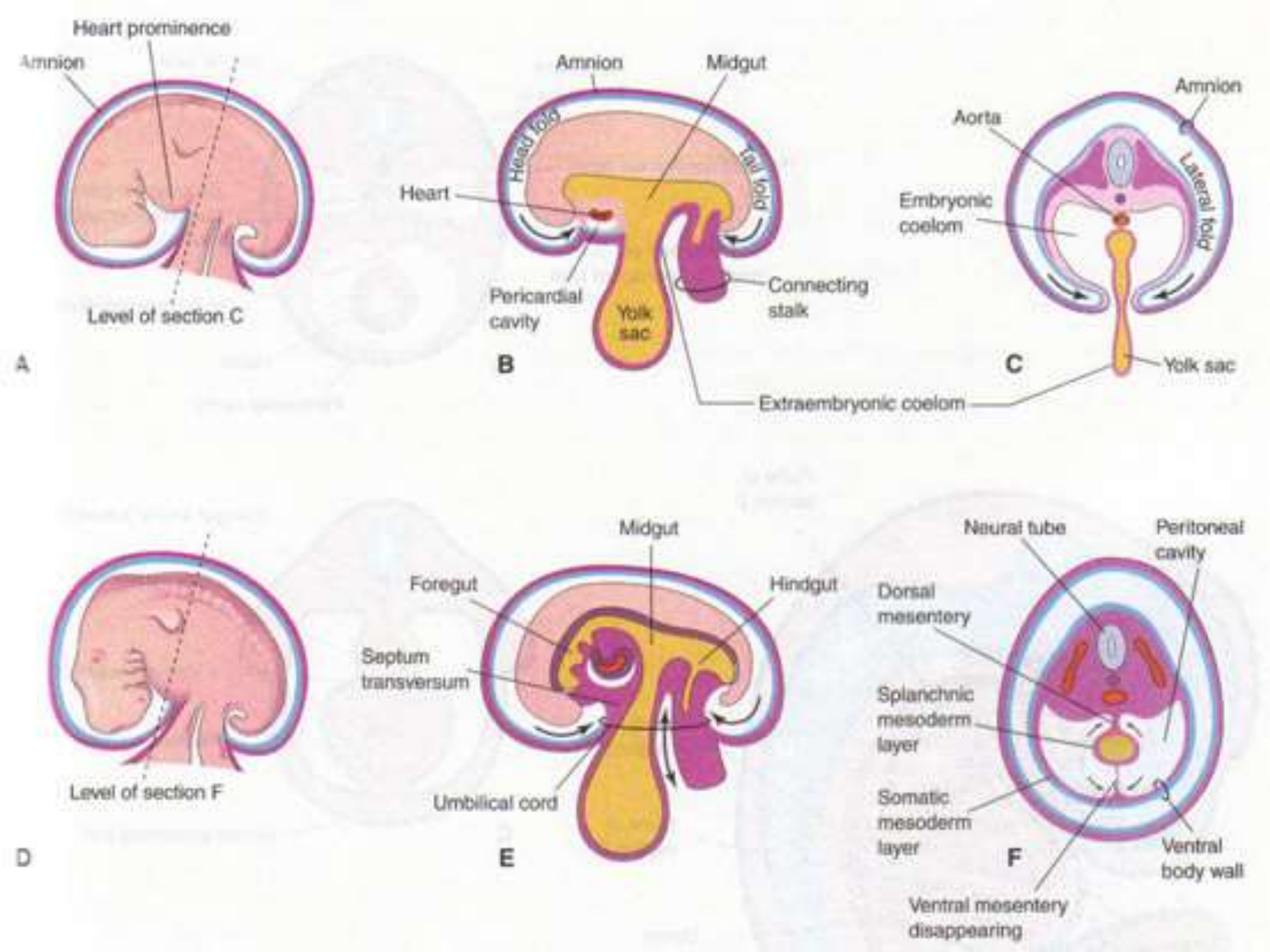
Abdominal wall defect

Incidence

- 1:20,000-30,000
- Sex ratio 1:1
- 10-15% have associated anomalies
- 40% are premature/SGA

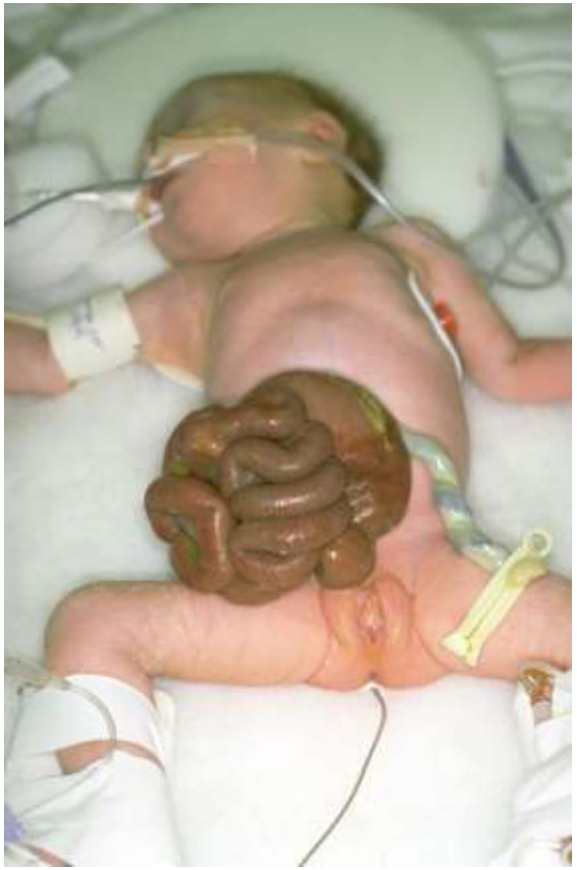
Pathophysiology

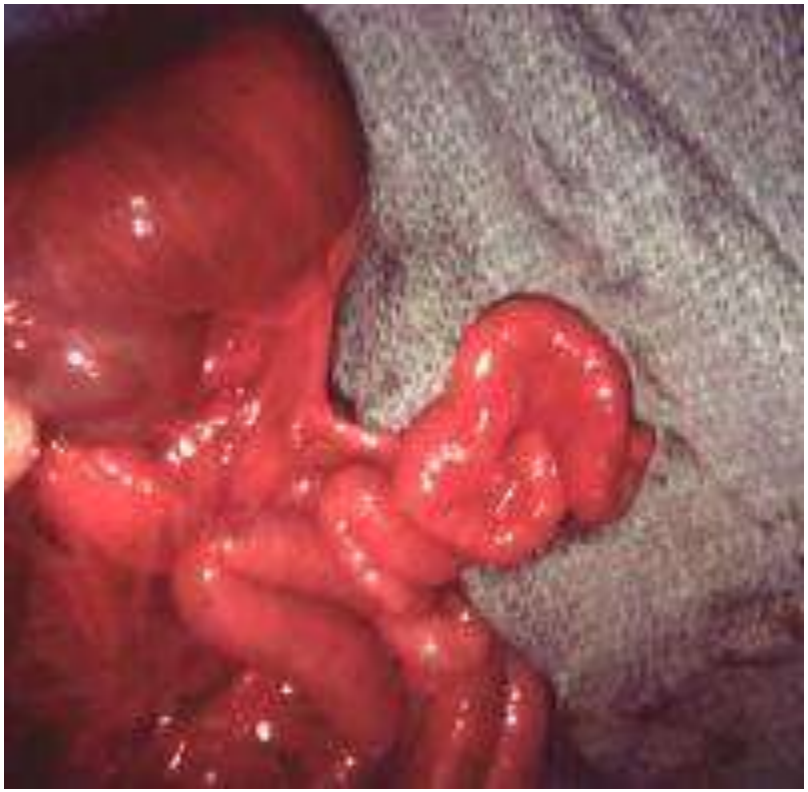
- Abnormal involution of right umbilical vein
- Rupture of a small omphalocoele
- Failure of migration and fusion of the lateral folds of the embryonic disc on the 3^{rd}-4^{th} week of gestation


Heart prominence
Amnion
Level of section C
A
Amnion
Midgut
Head fold
Tail fold
Heart
Pericardial cavity
Yolk sac
Connecting stalk
B
Amnion
Aorta
Lateral fold
Embryonic coelom
Yolk sac
C
Extraembryonic coelom
Level of section F
D
Midgut
Foregut
Hindgut
Septum transversum
Umbilical cord
E
Neural tube
Peritoneal cavity
Dorsal mesentery
Splanchnic mesoderm layer
Somatic mesoderm layer
Ventral body wall
F
Ventral mesentery disappearing

Clinical Findings

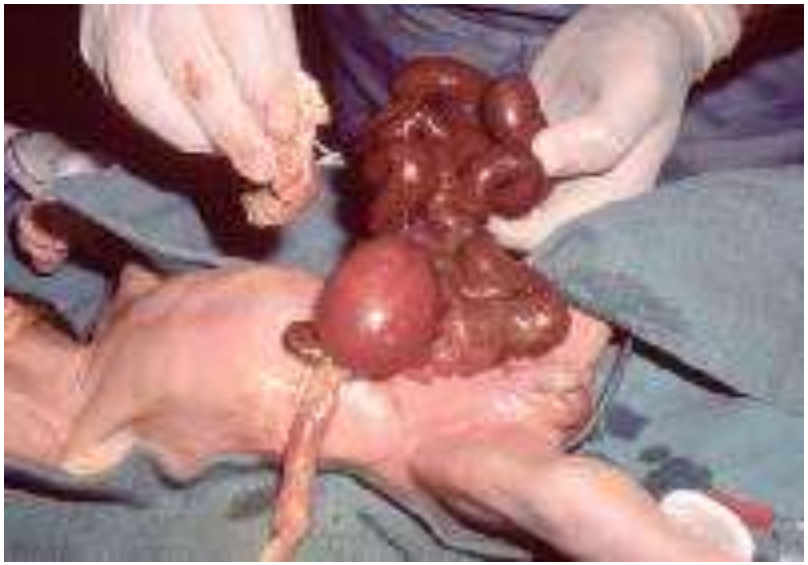
- Defect to the right of an intact umbilical cord allowing extrusion of abdominal content
- Opening ≅ 5 cm
- No covering sac

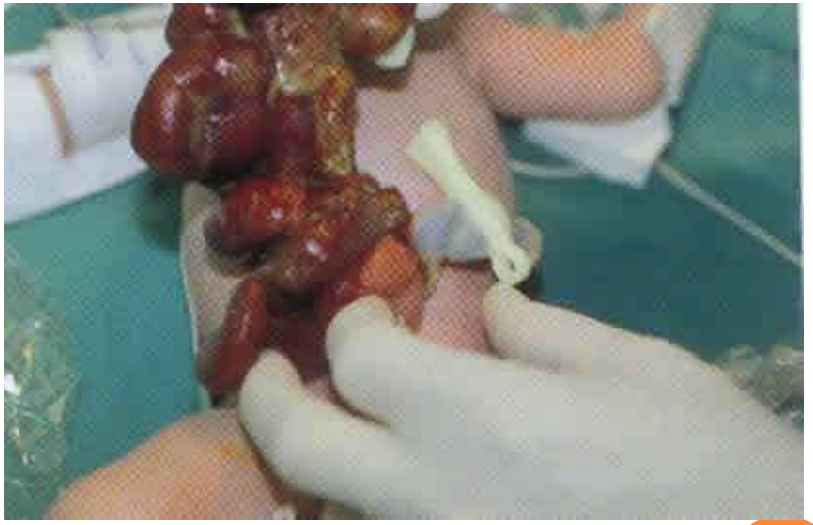




- Bowels often thickened, matted and edematous
- 10-15% with intestinal atresia

- Evisceration of the bowel leads to malrotation.
- Constriction of the base may cause intestinal stenosis, atresia, and volvulus
- Undescended testicles
- preterm or small for gestational age (SGA)





Causes

- Folic acid deficiency
- hypoxia
- salicylates

Diagnosis

- History : Prenatal U/S
- Polyhydramnios
- MSAFP
- Amniocentesis

MANAGEMENT

- ABC
- Heat Management
  - Sterile wrap or sterile bowel bag
  - Radiant warmer
- Fluid Management
  - IV bolus 20 ml/kg LR/NS
  - D10¼NS 2-3 maintenance rate

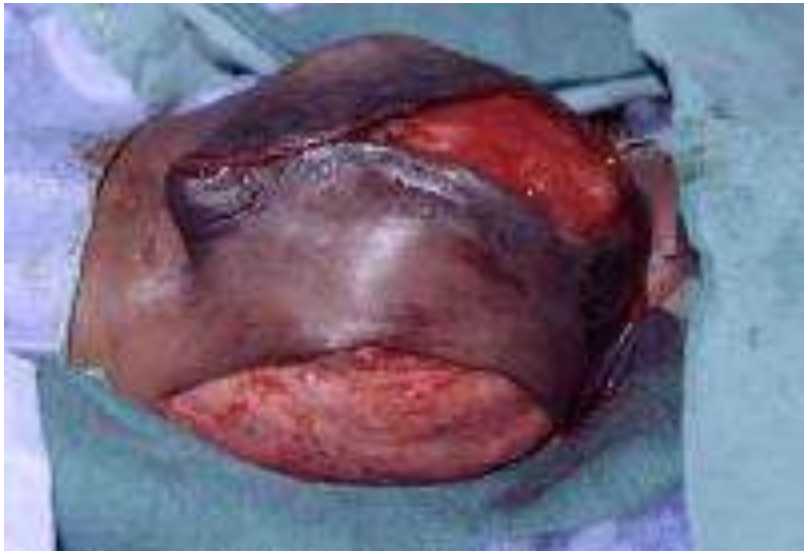
- Nutrition
  - NPO and TPN (central venous line)
- Gastric Distention
  - OG/NG tube
  - urinary catheter
- Infection Control

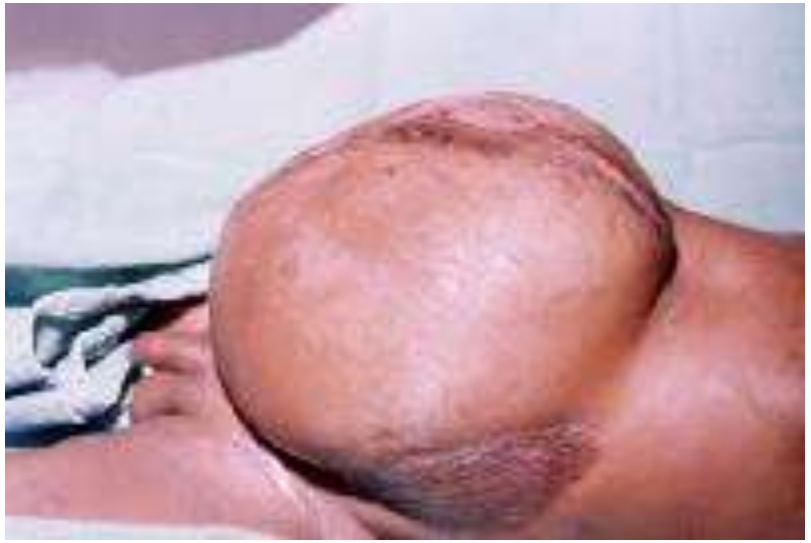
  Broad-spectrum antibiotics
- Associated Defects

- Conservative treatment
  - Reduction by squeezing the sac
  - Painting sac with escharotic agent
    - 0.25% Silver nitrate
    - 0.25% Merbromin (Mercurochrome)

- Surgical Management
  - Skin Flaps
  - Primary Closure
  - Staged Closure
    - Staged repair using silo pouch

Skin Flaps

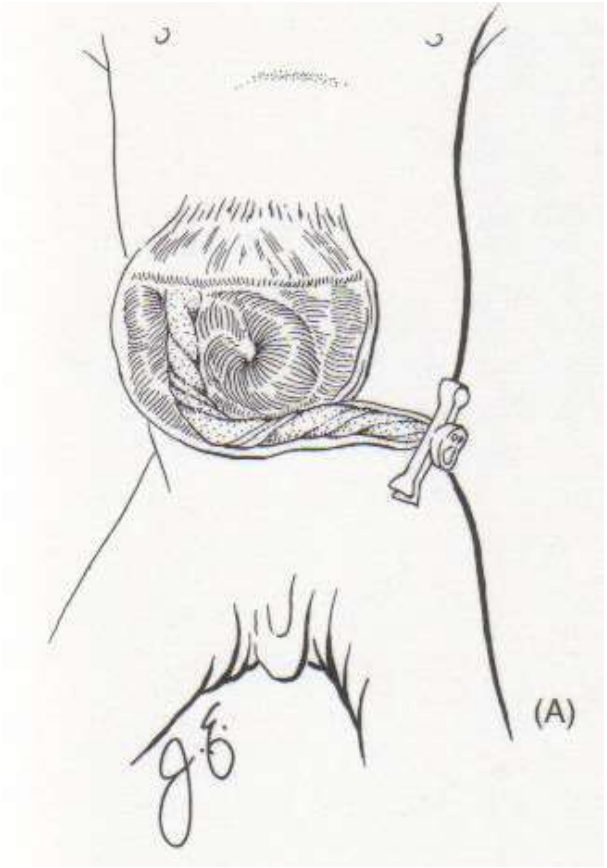


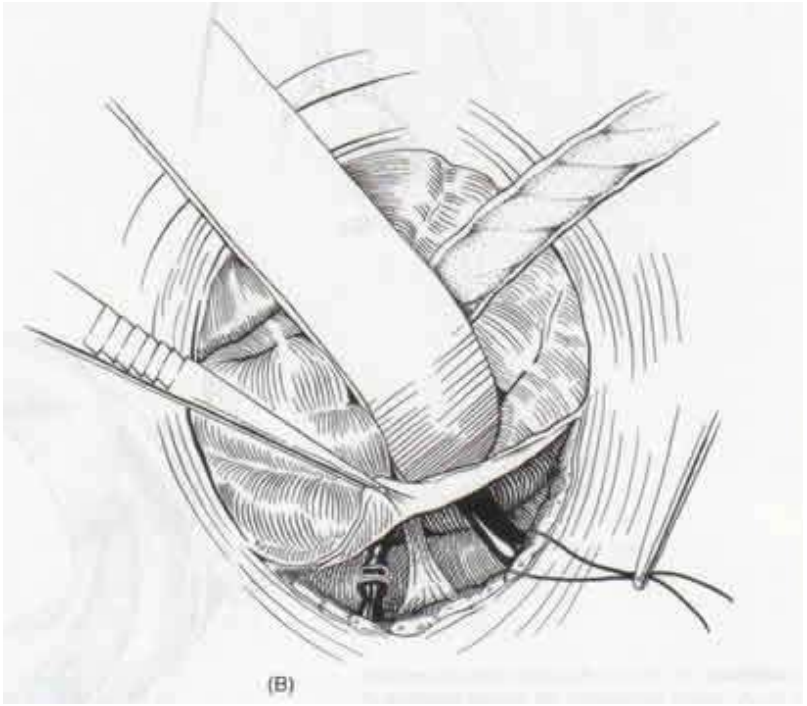


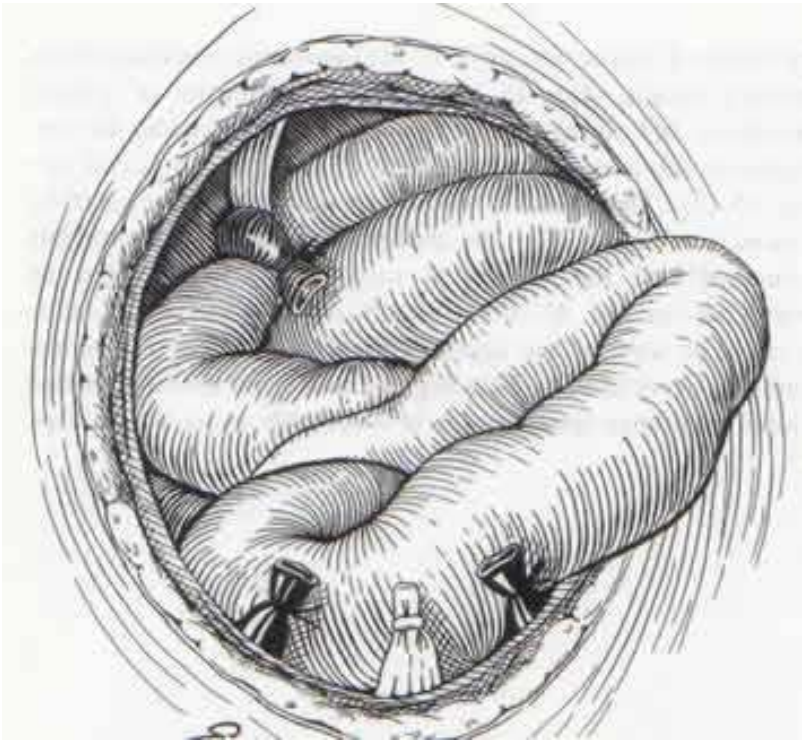
Primary Closure

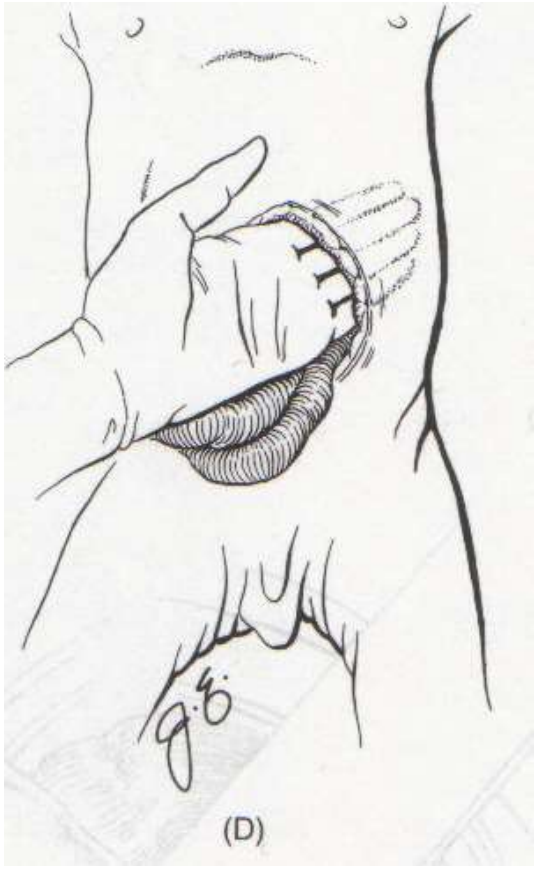
- In 1967, Schuster technique
- A circumferential incision along the skin-omphalocele junction; the omphalocele membrane is left intact
- Teflon sheets
- DualMesh patch (Gore-Tex)
- AlloDerm patch (acellular human dermis)

Primary Closure





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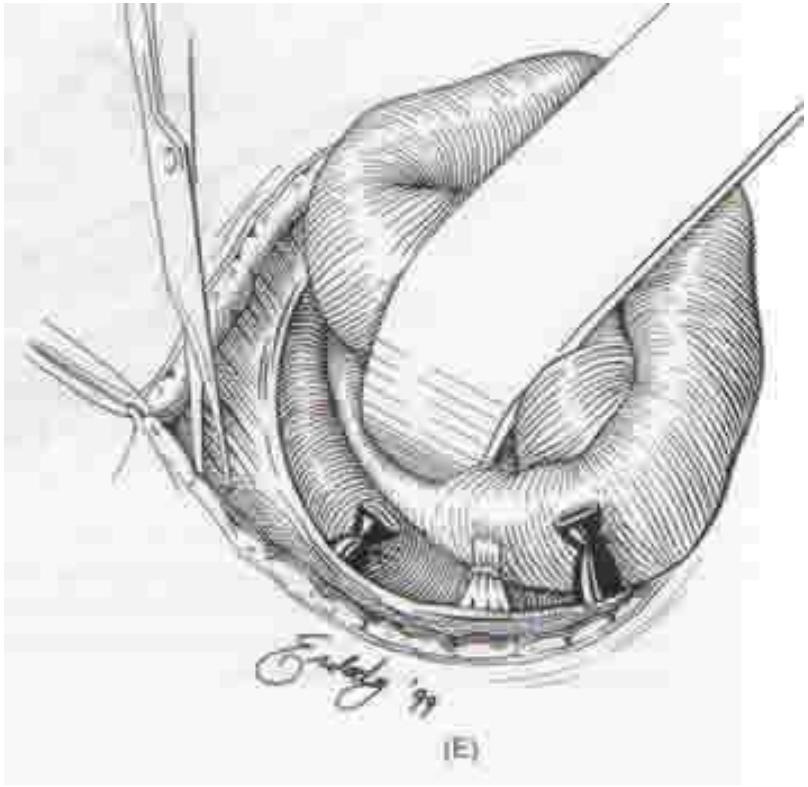
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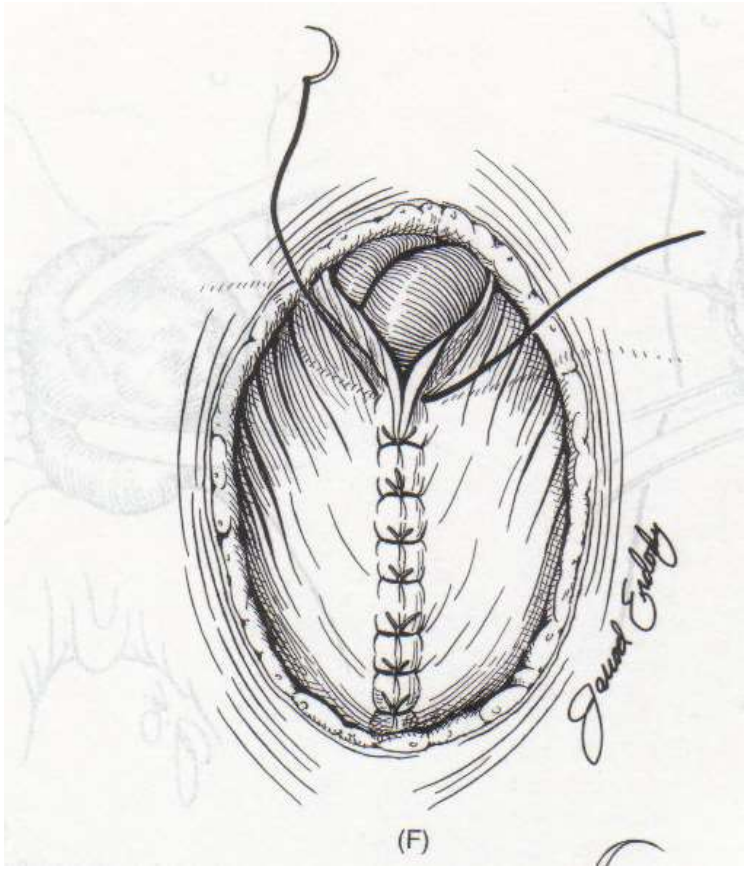
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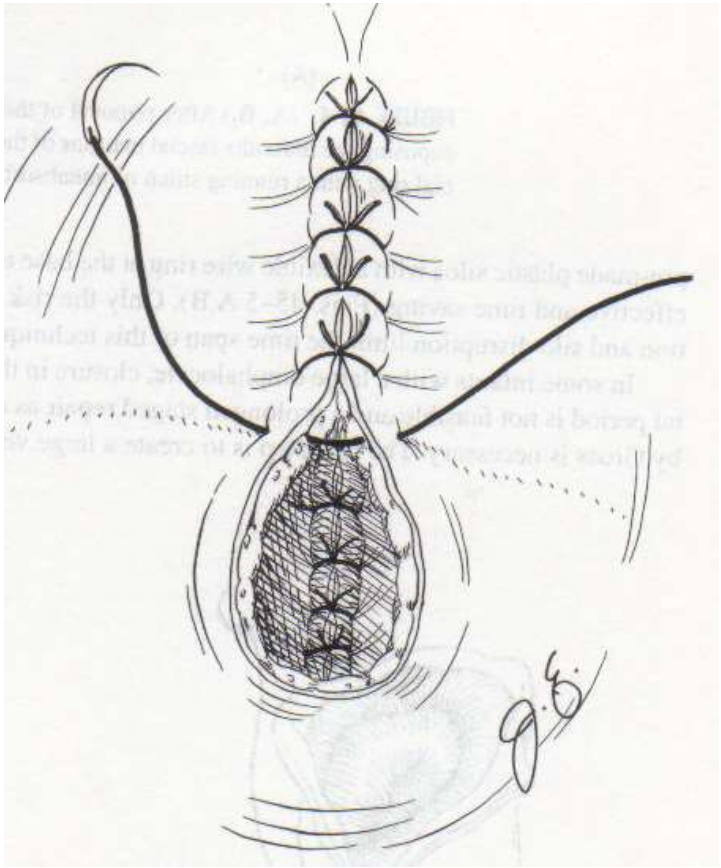
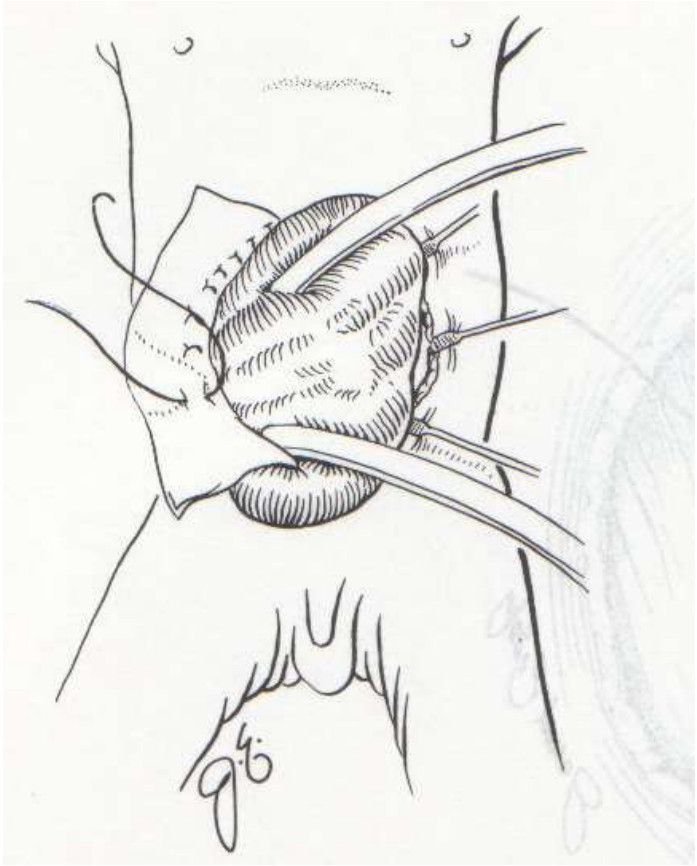


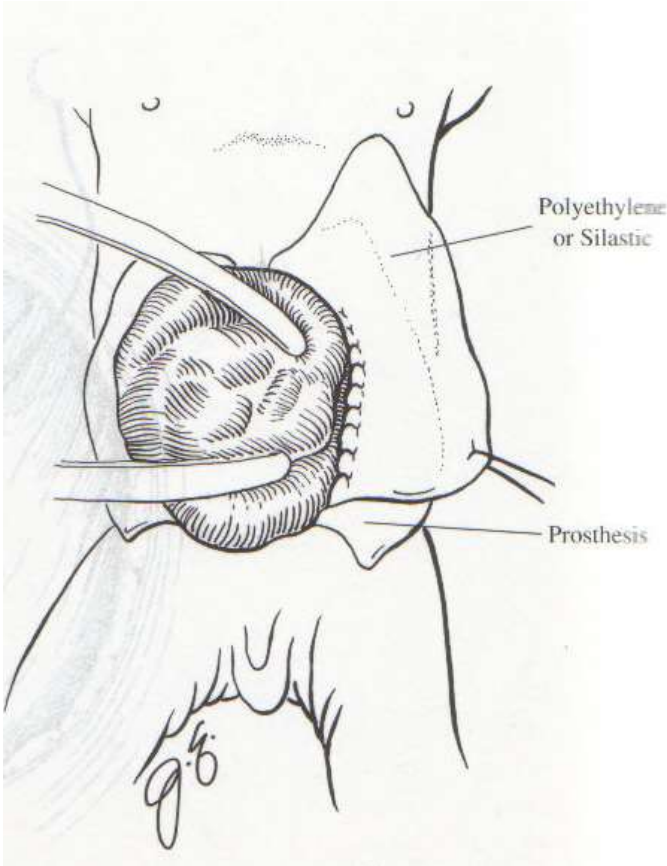
Staged Closure

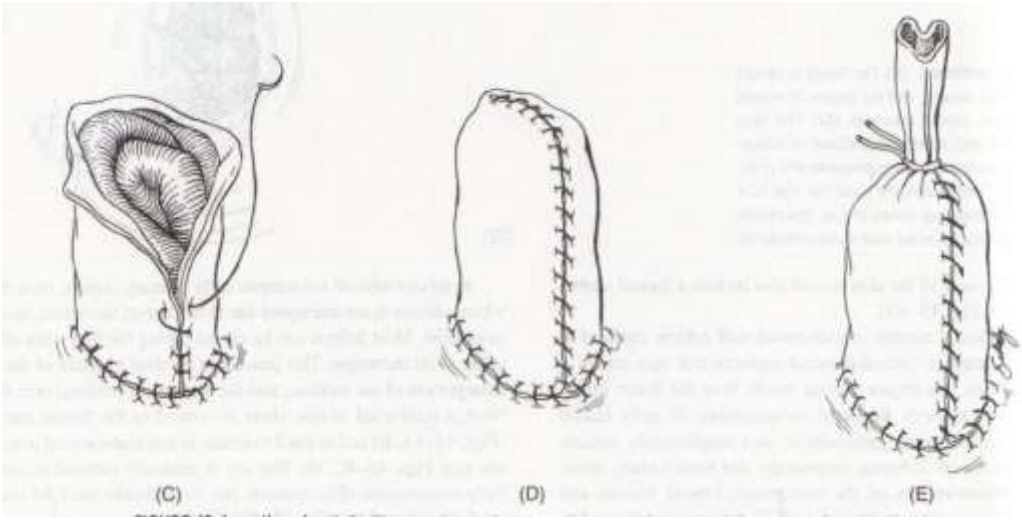
In 1969, Allen and Wrenn adapted Schuster's technique to treat gastroschisis

Silo Procedure.

Staged Closure



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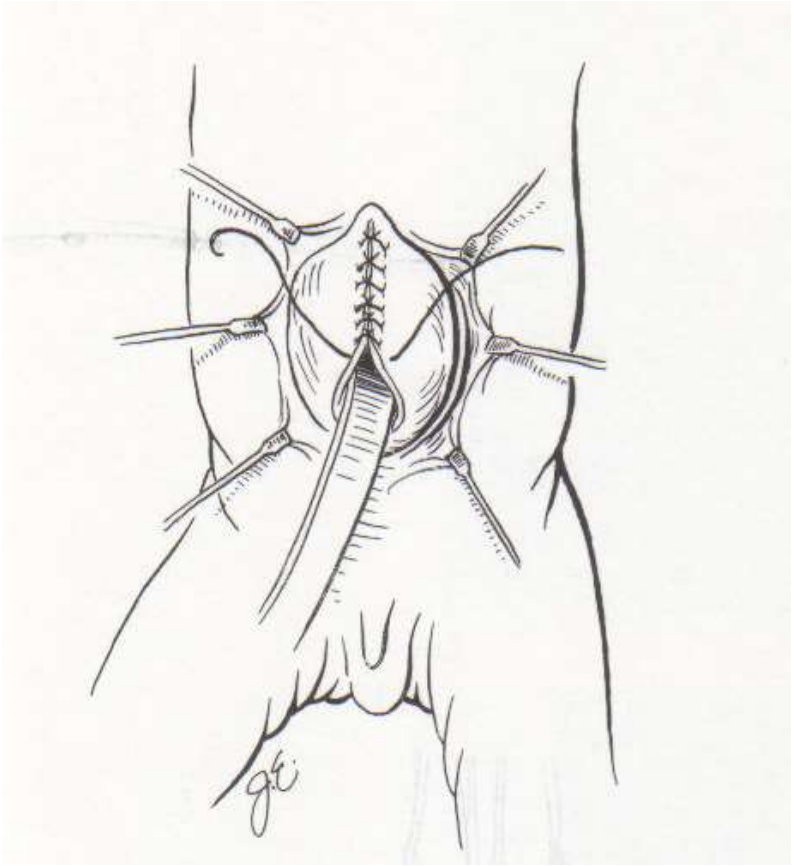

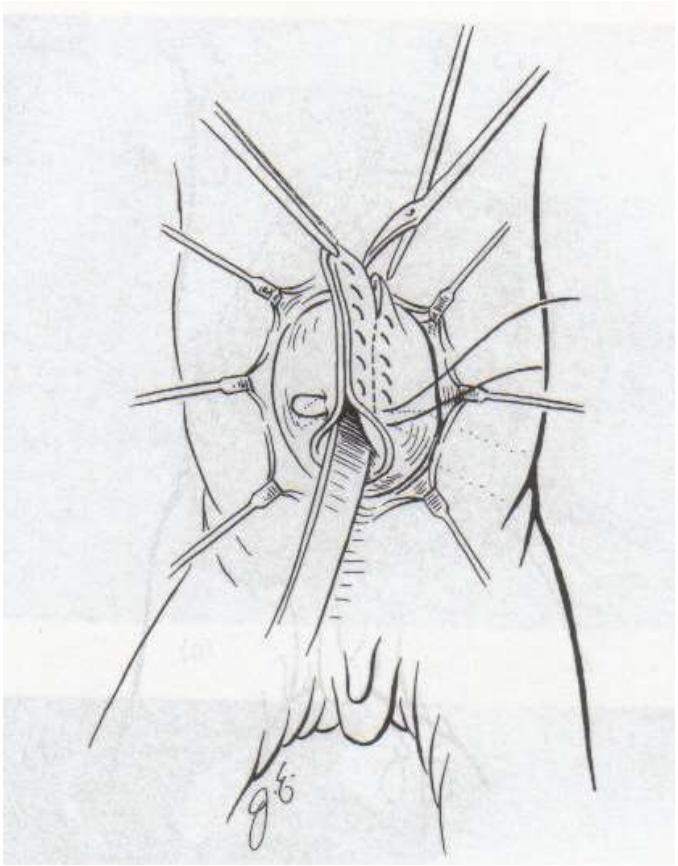

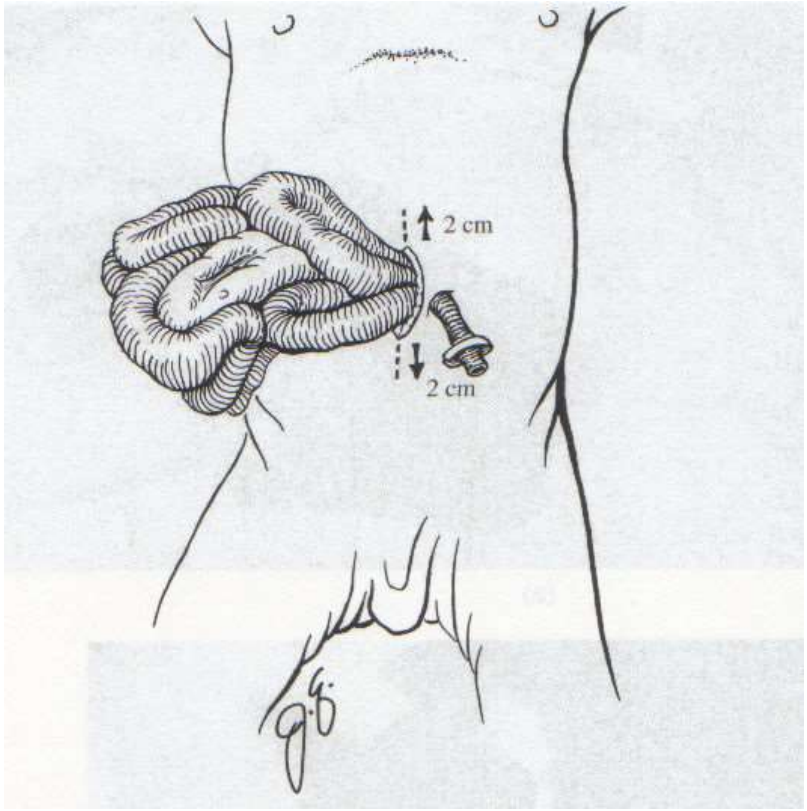
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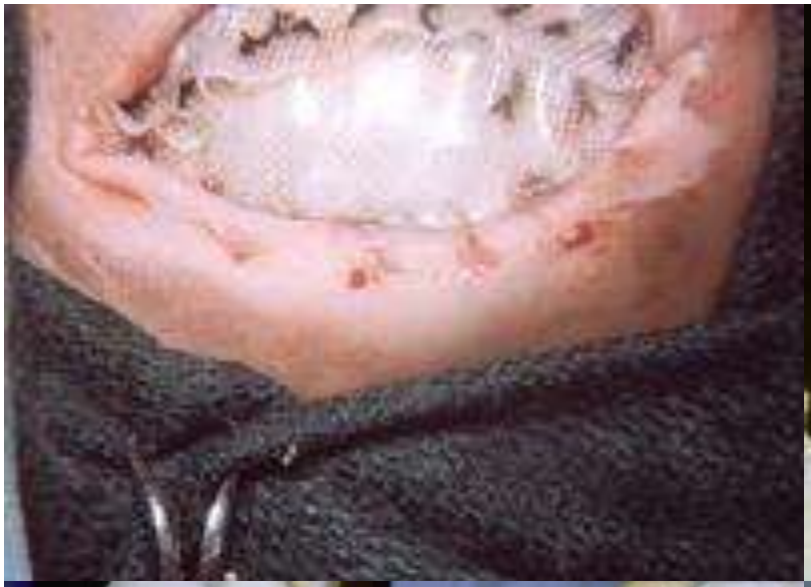
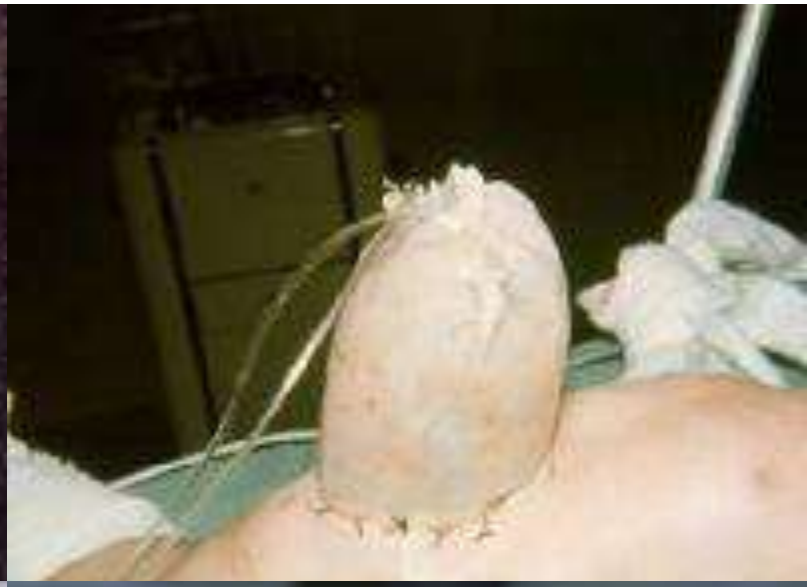
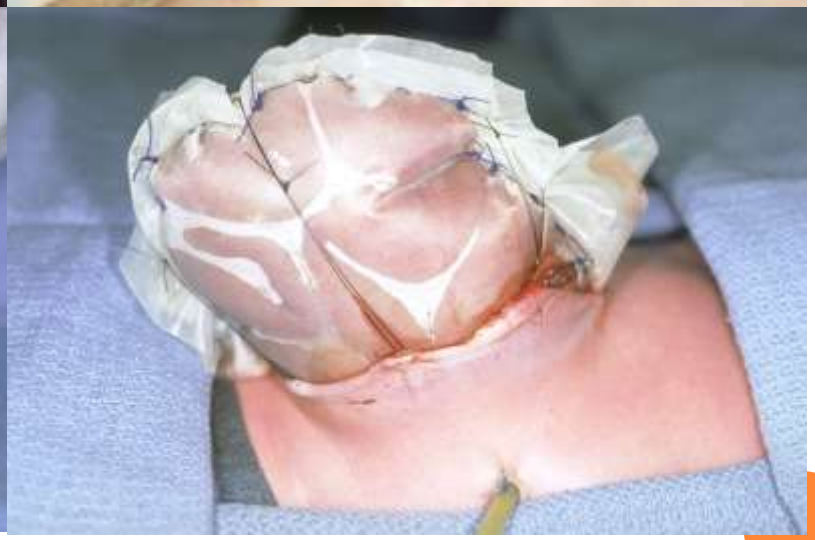
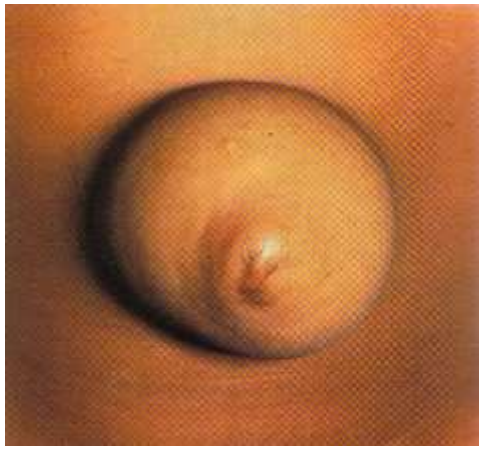





- Technically Gastroschisis and Omphaloceles are all malrotated.
- Are they at increased risk of volvulus after closure of the defect?

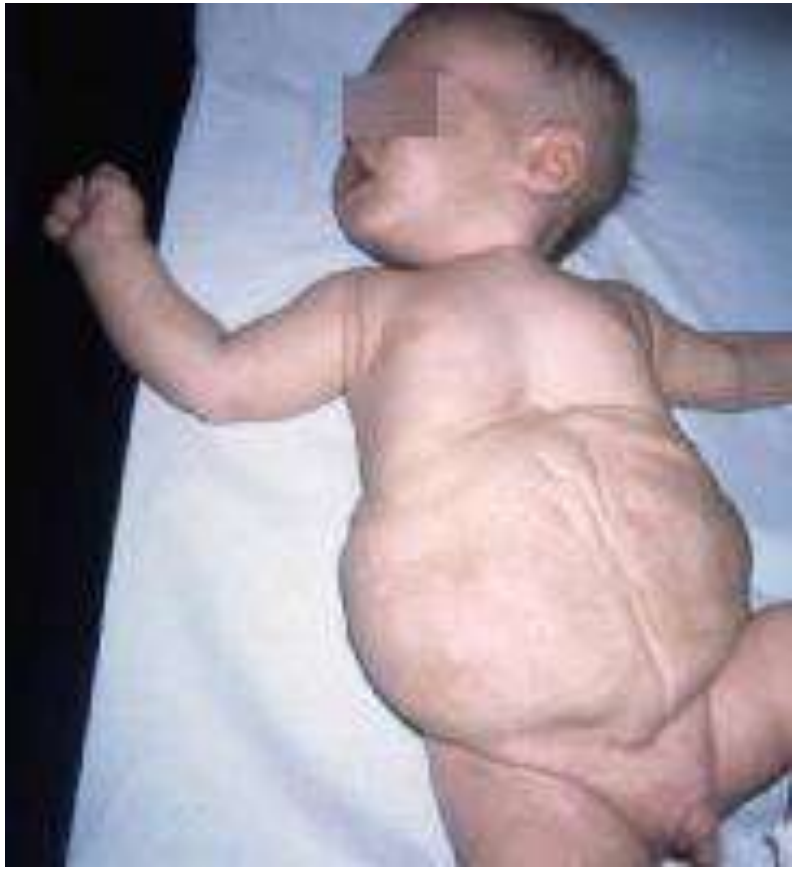
UMBILICAL HERNIA

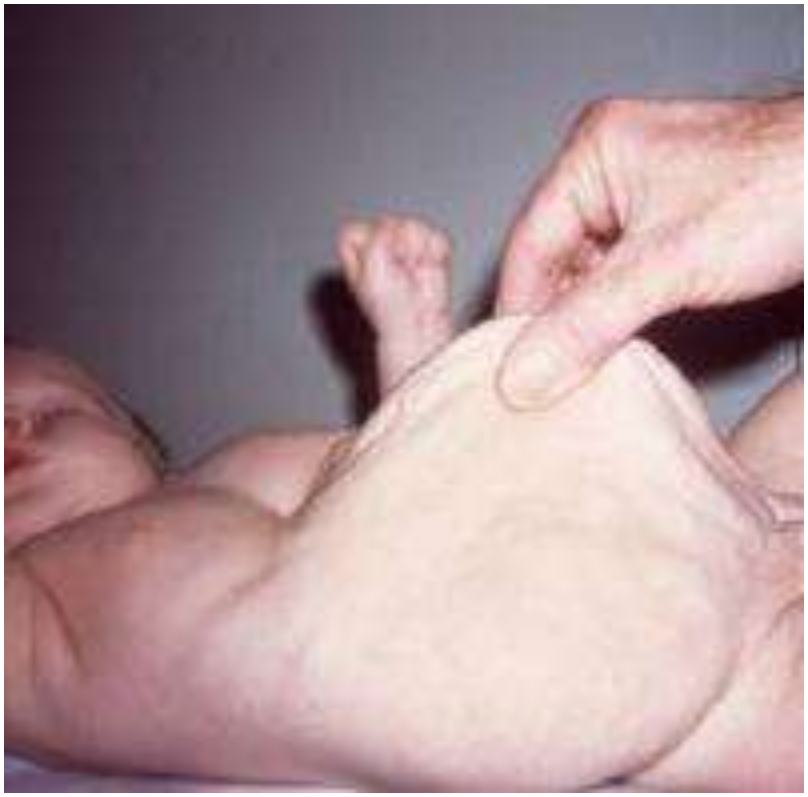
- Defect in linea alba, subcutaneous tissue and skin covering the protruding bowel
- Frequent in premature infants



PRUNE BELLY SYNDROME

- 1:30,000-50,000
- 95% are male
- A partial or complete lack of abdominal wall muscles. There are wrinkly folds of skin covering the abdomen.
- Undescended testicles
- Urinary tract abnormality such as unusually large ureters, distended bladder, vesicoureteral reflux
- Frequent urinary tract infections
- VSD
- Malrotation of the gut
- Club foot

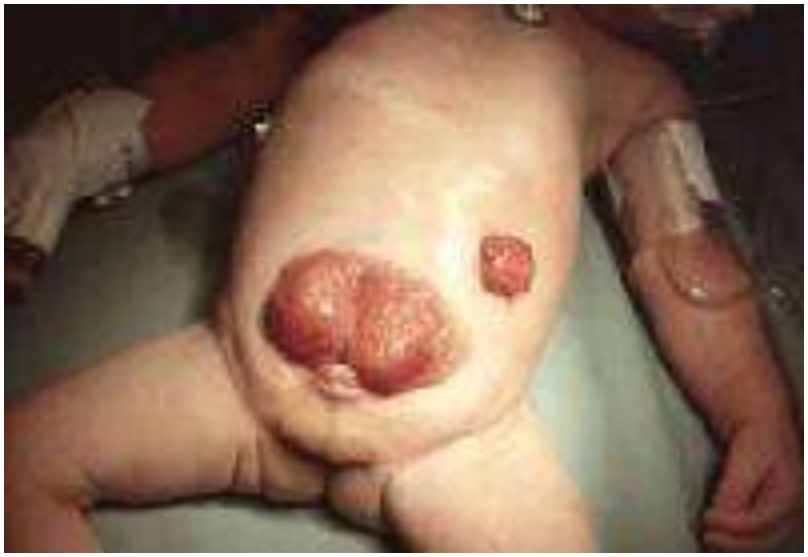




- Tx:
  - Treatment of the associated anomalies
  - Usually end up with a cystostomy for urinary drainage

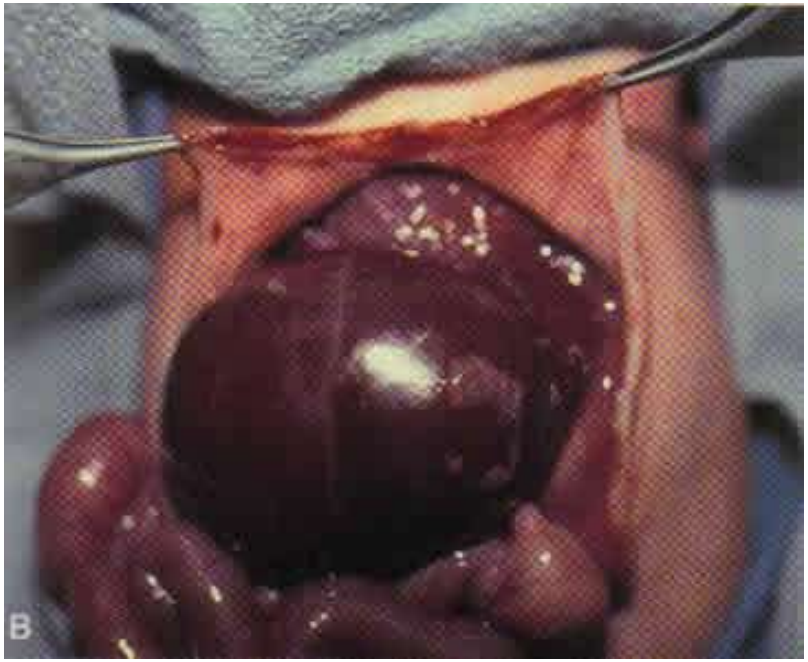
BLADDER EXTROPHY (ECTOPIA VESICAE)

- A defect in the abdominal wall occupied by both the exstrophied bladder as well as a portion of the urethra
- 3.3 in 100,000 births
- Separation of the pubic symphysis
- Shortening of the pubic rami
- External rotation of the pelvis.
- Associated with prolapsed vagina or rectum, epispadias, bifid clitoris or penis
- Tx: Reconstruction



PENTALOGY OF CANTRELL

- Omphalocoele
- Anterior diaphragmatic hernia
- Sternal cleft
- Ectopia Cordis
- Intracardiac defect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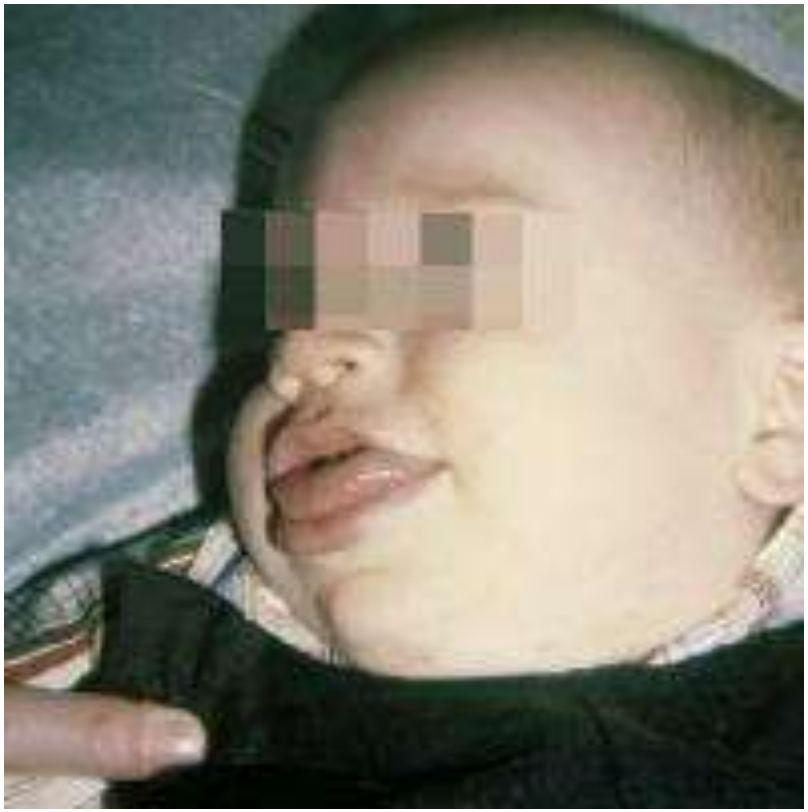
BECKWITH-WIEDEMANN SYNDROME



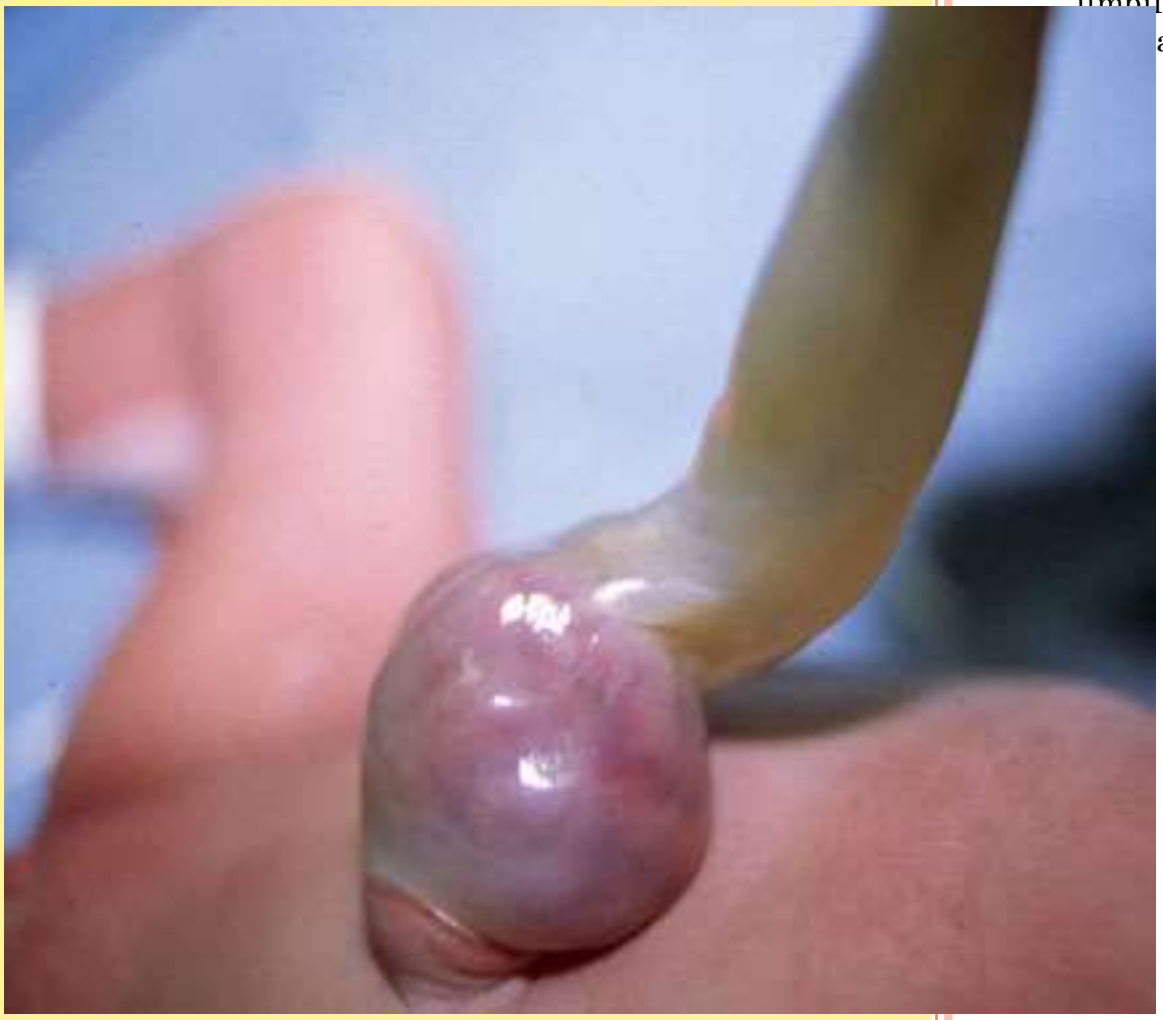
- Macrosomia
- Macroglossia
- Organomegaly
- Abdominal wall defects
- Embryonal tumors

- Have coarse, rounded facial features
- hyperplasia of the pancreatic islet cells with hypoglycemia; visceromegaly
- genitourinary abnormalities

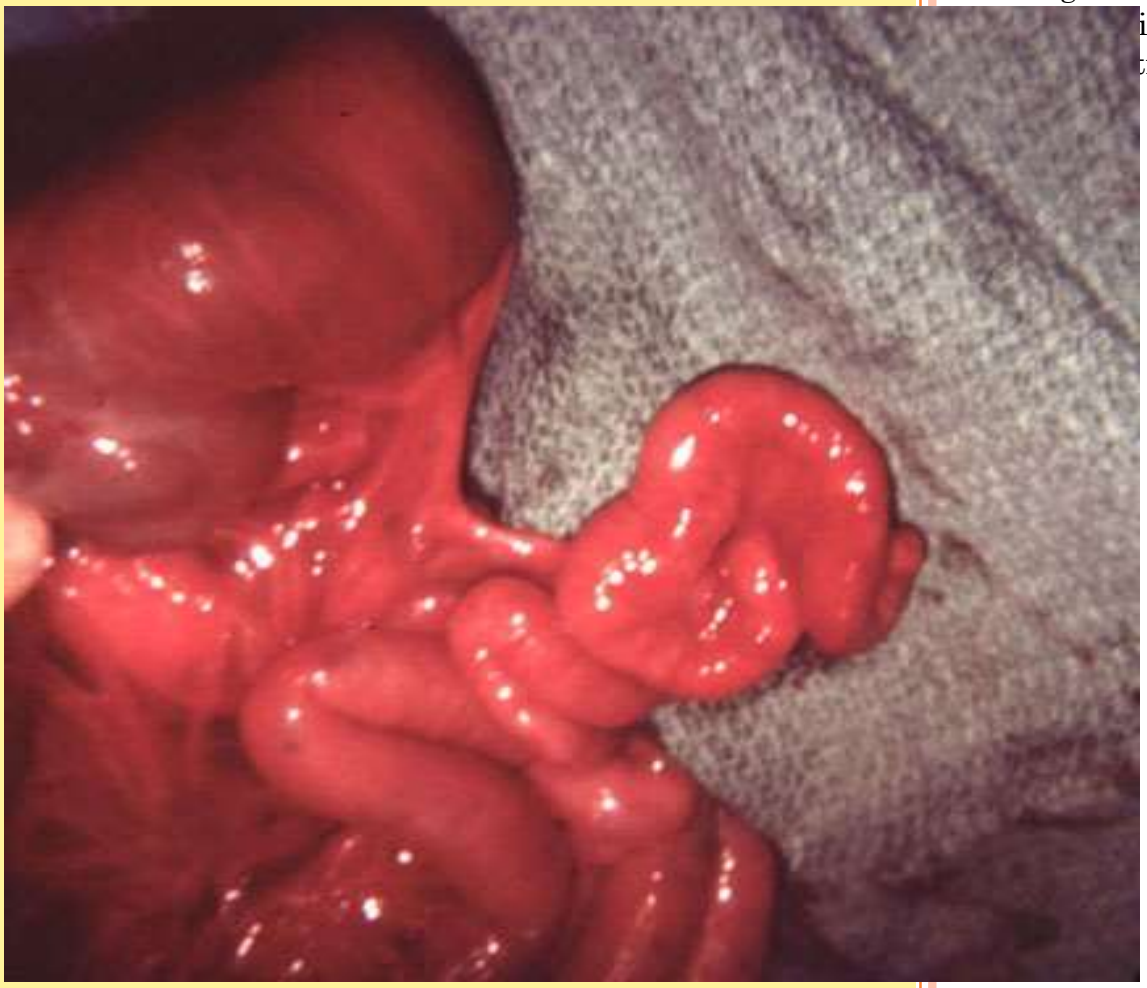
	Omphalocoele	Gastroschisis
Incidence	1:6,000-10,000	1:20,000-30,000
Delivery	Vaginal or CS	CS
Covering Sac	Present	Absent
Size of Defect	Small or large	Small
Cord Location	Onto the sac	On abdominal wall
Bowel	Normal	Edematous, matted

	Omphalocoele	Gastroschisis
Other Organs	Liver often out	Rare
Prematurity	10-20%	50-60%
IUGR	Less common	Common
NEC	If sac is ruptured	18%
Associated Anomalies	>50%	10-15%
Treatment	Often primary	Often staged
Prognosis	20%-70%	70-90%

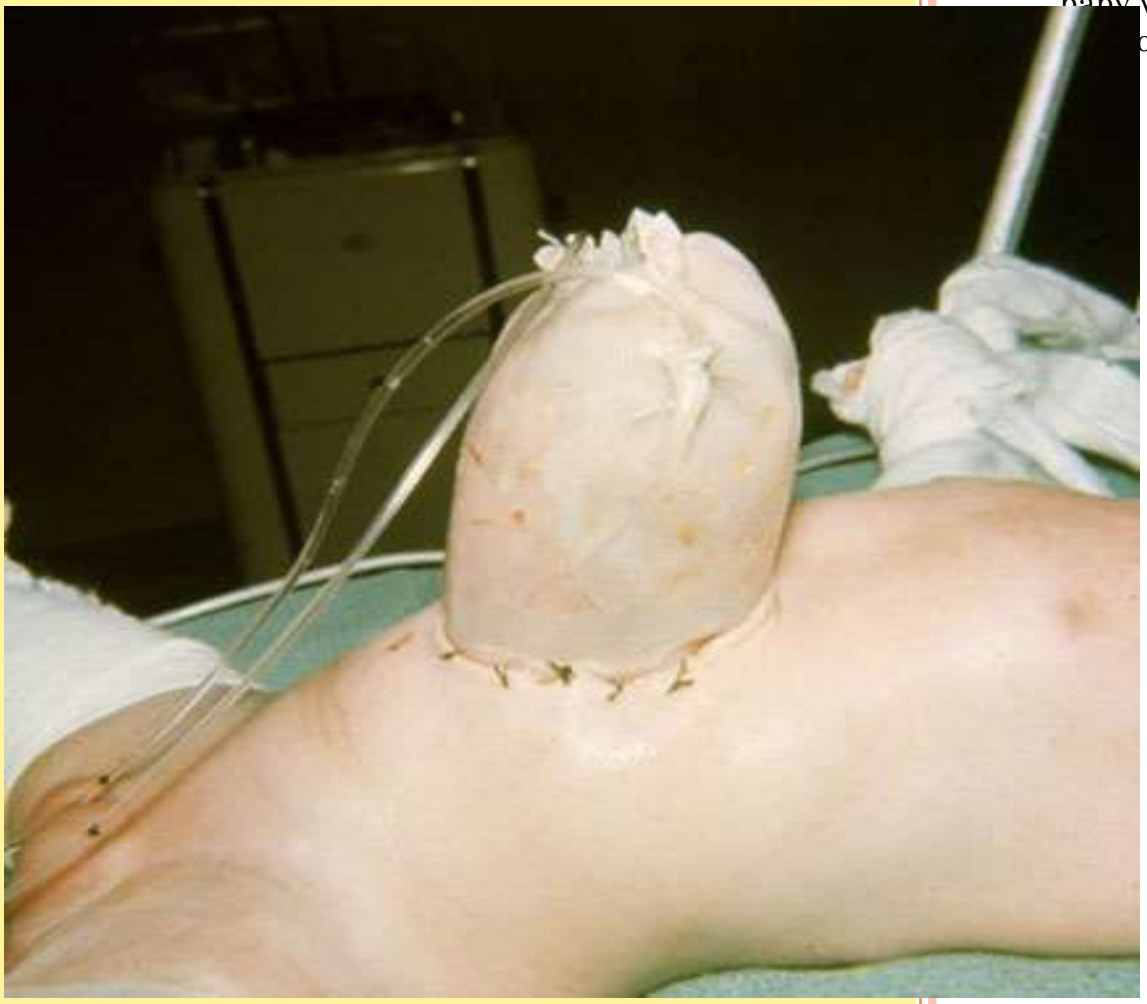
Baby with an umbilical cord a.

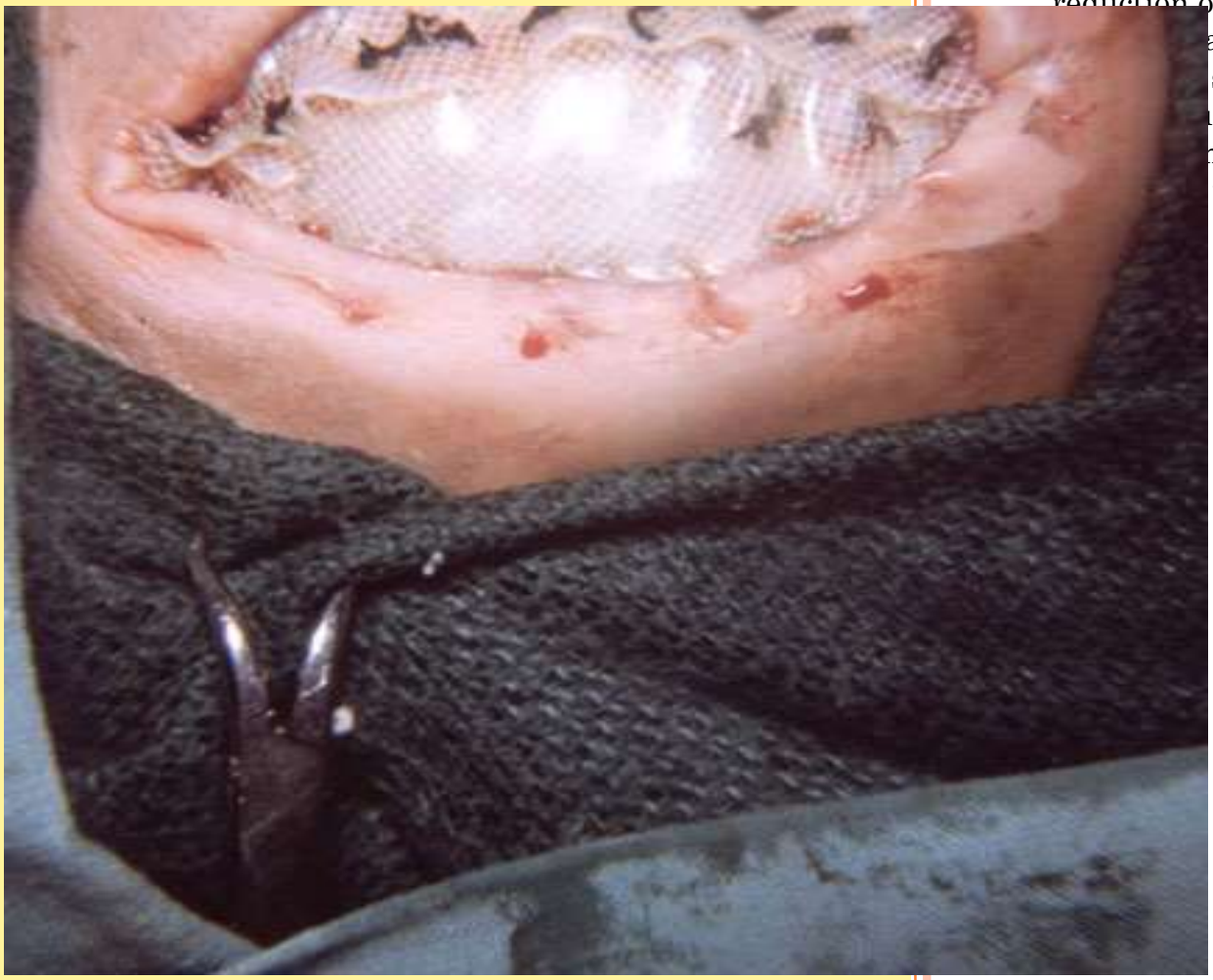


Baby with gastroschisis and iated tinal atresia



Silo closure of a baby with gastroschisis.





Completed
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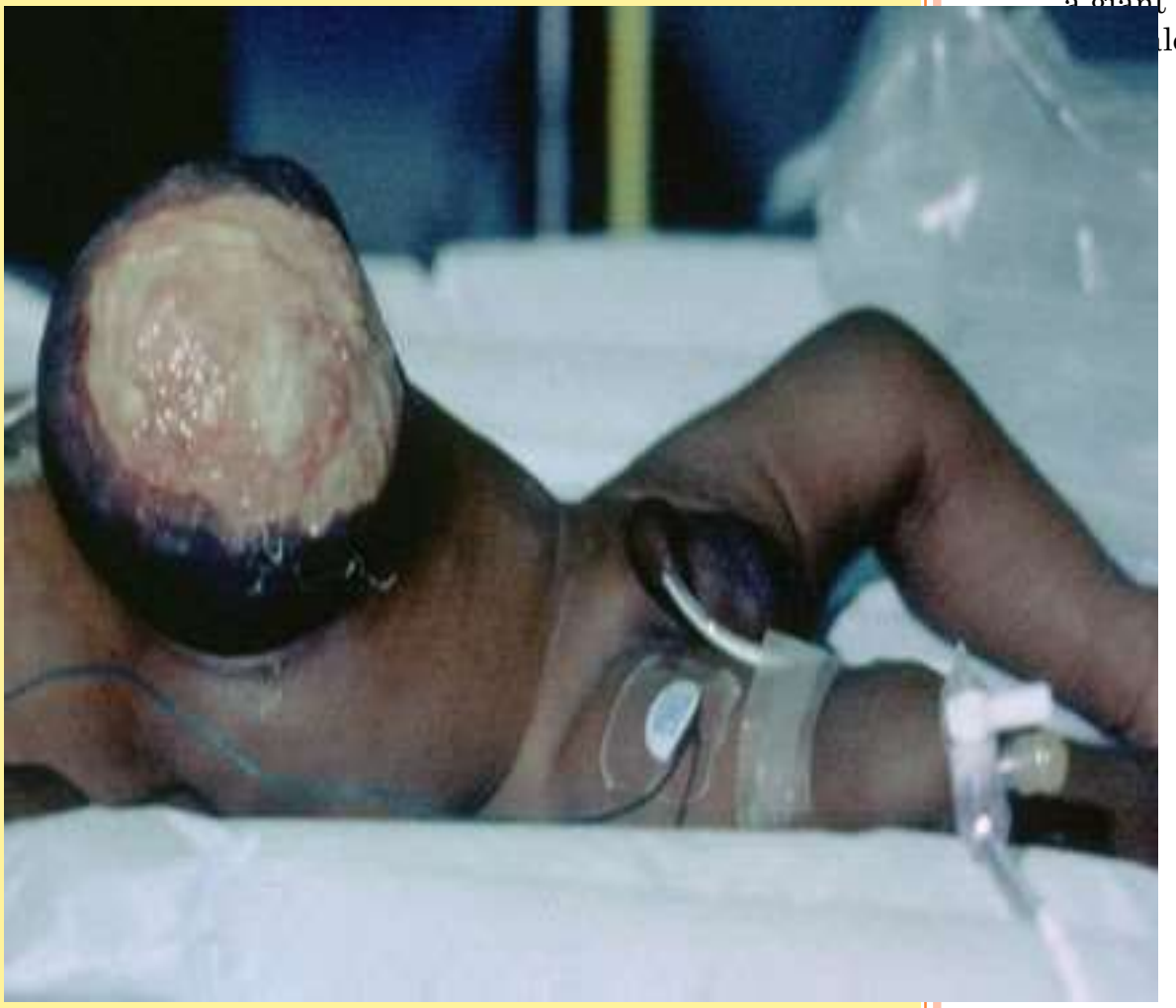
Case A. Baby with a giant
locele.



Case A. Closure of the giant
locele using
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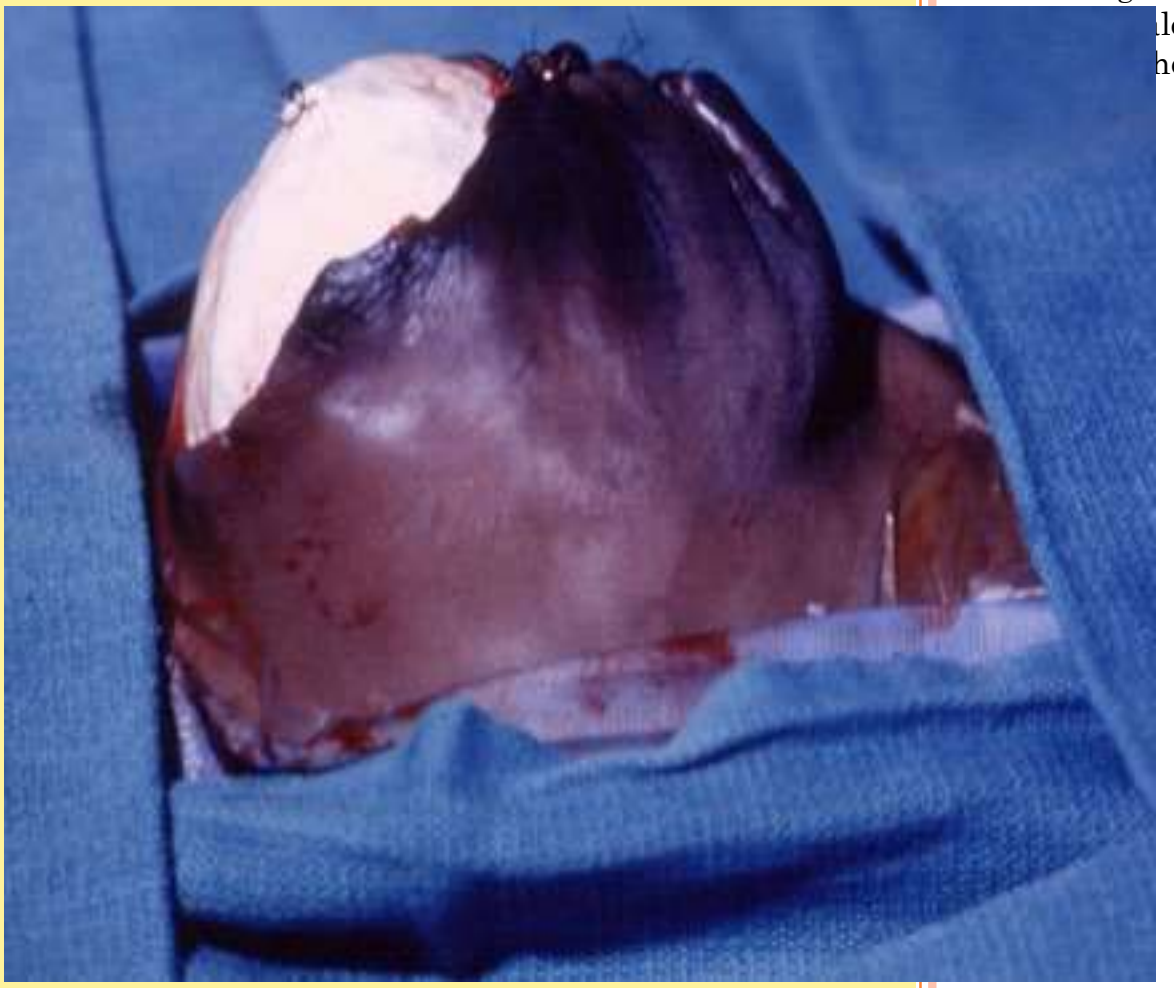

Case A.

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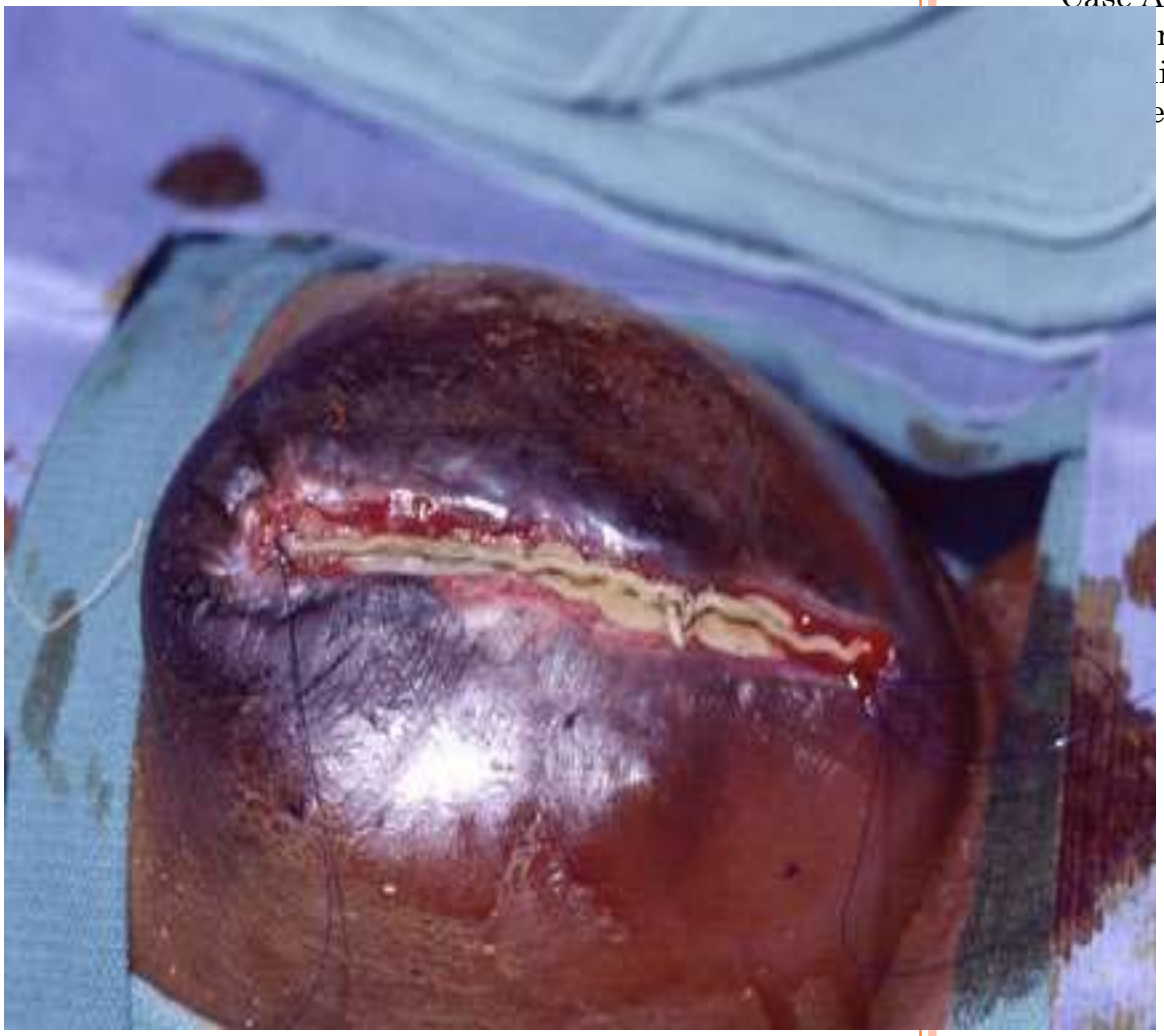
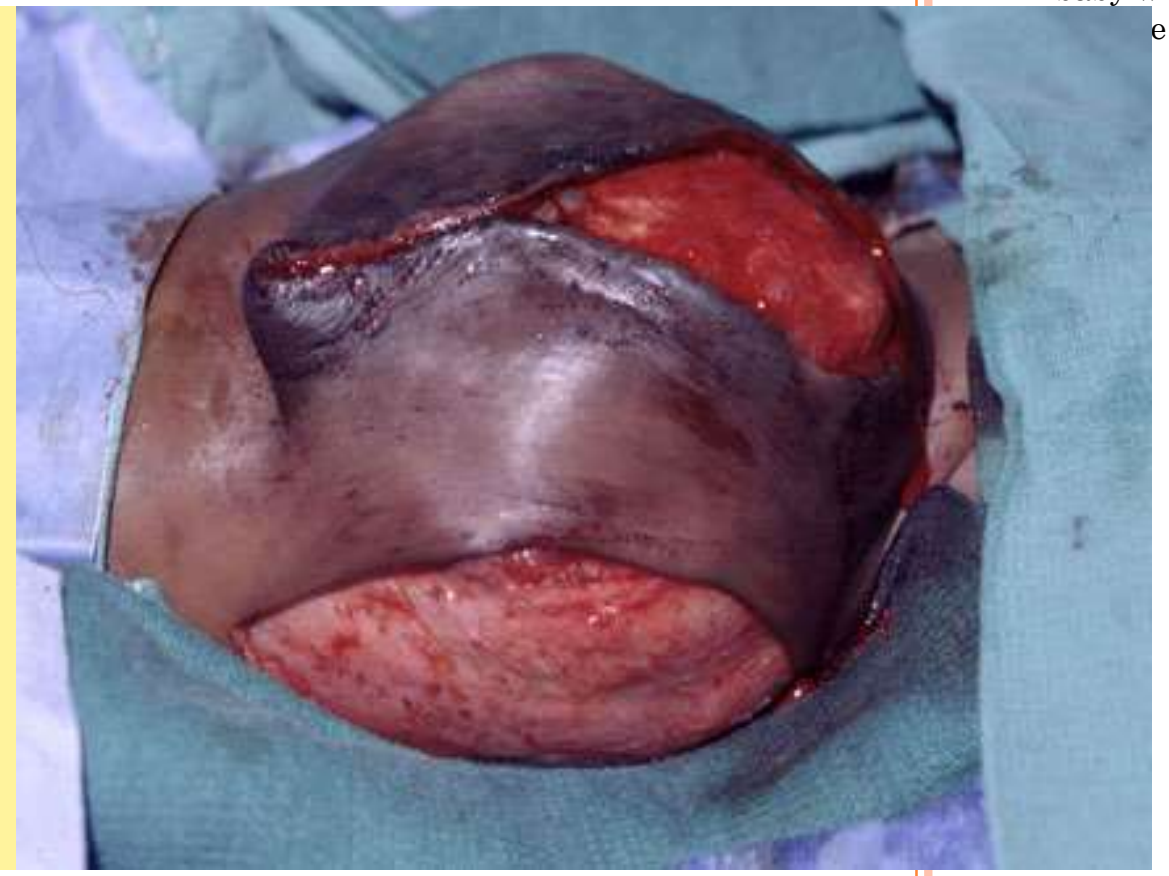
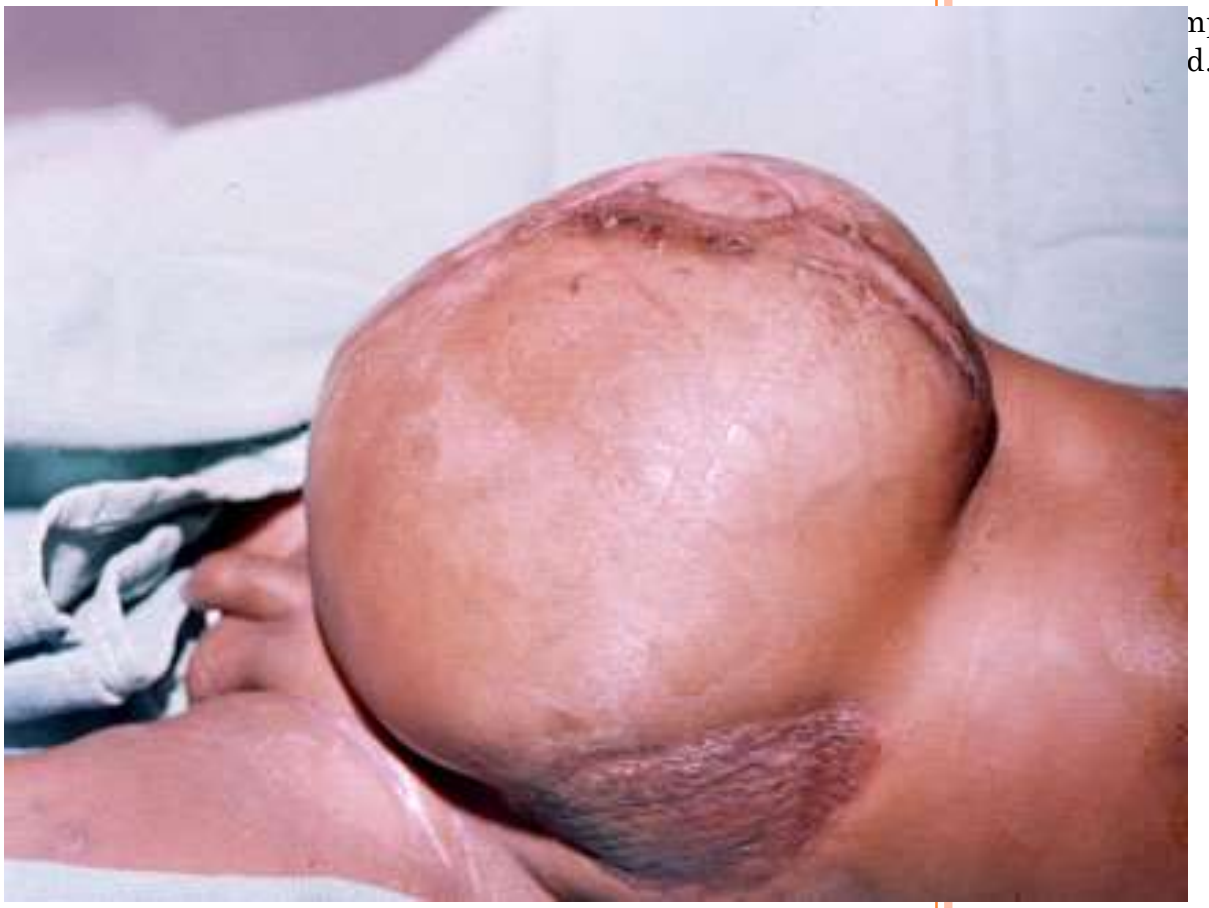


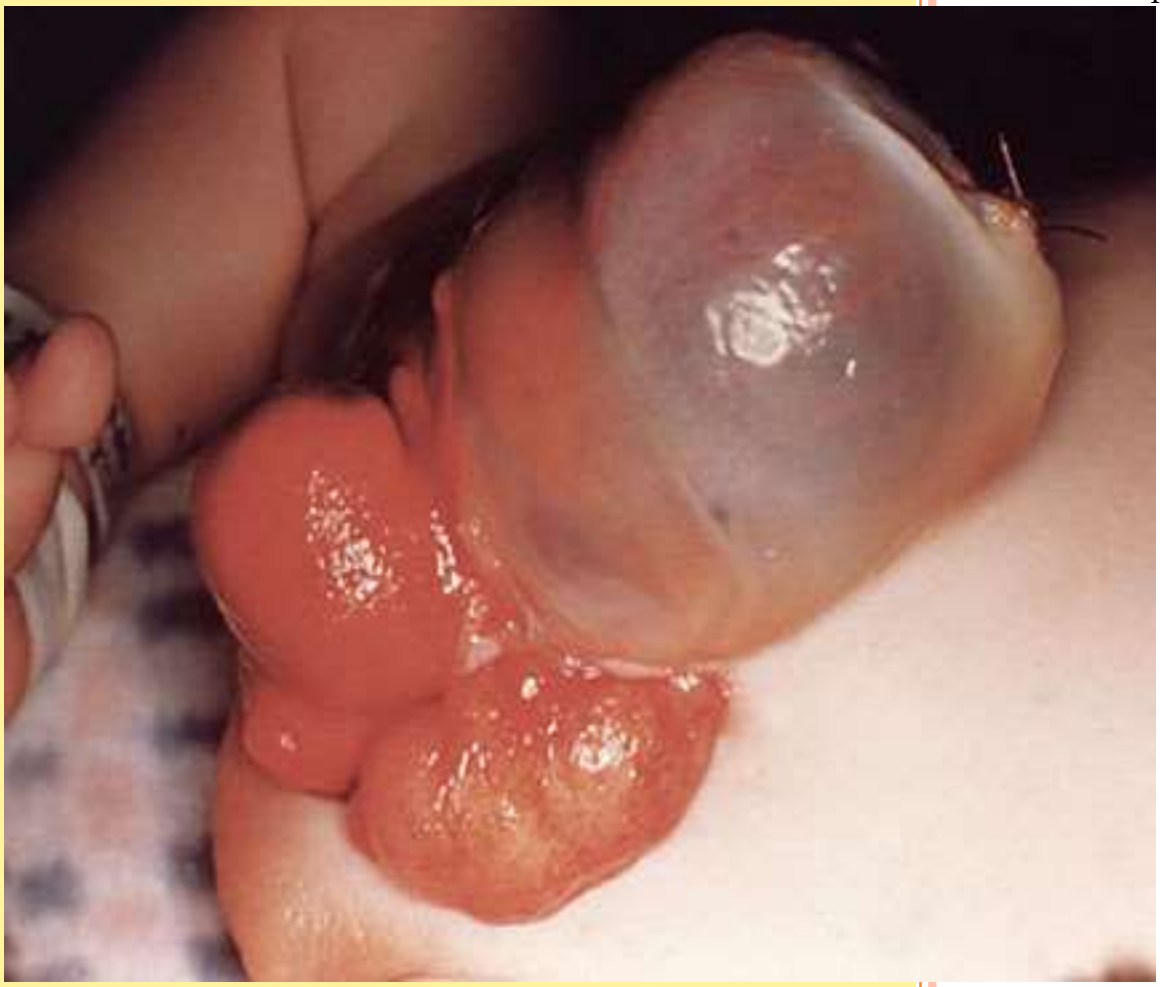
Case A. Flank flaps were used to close the giant omphalocele in the baby whose patch e infected.



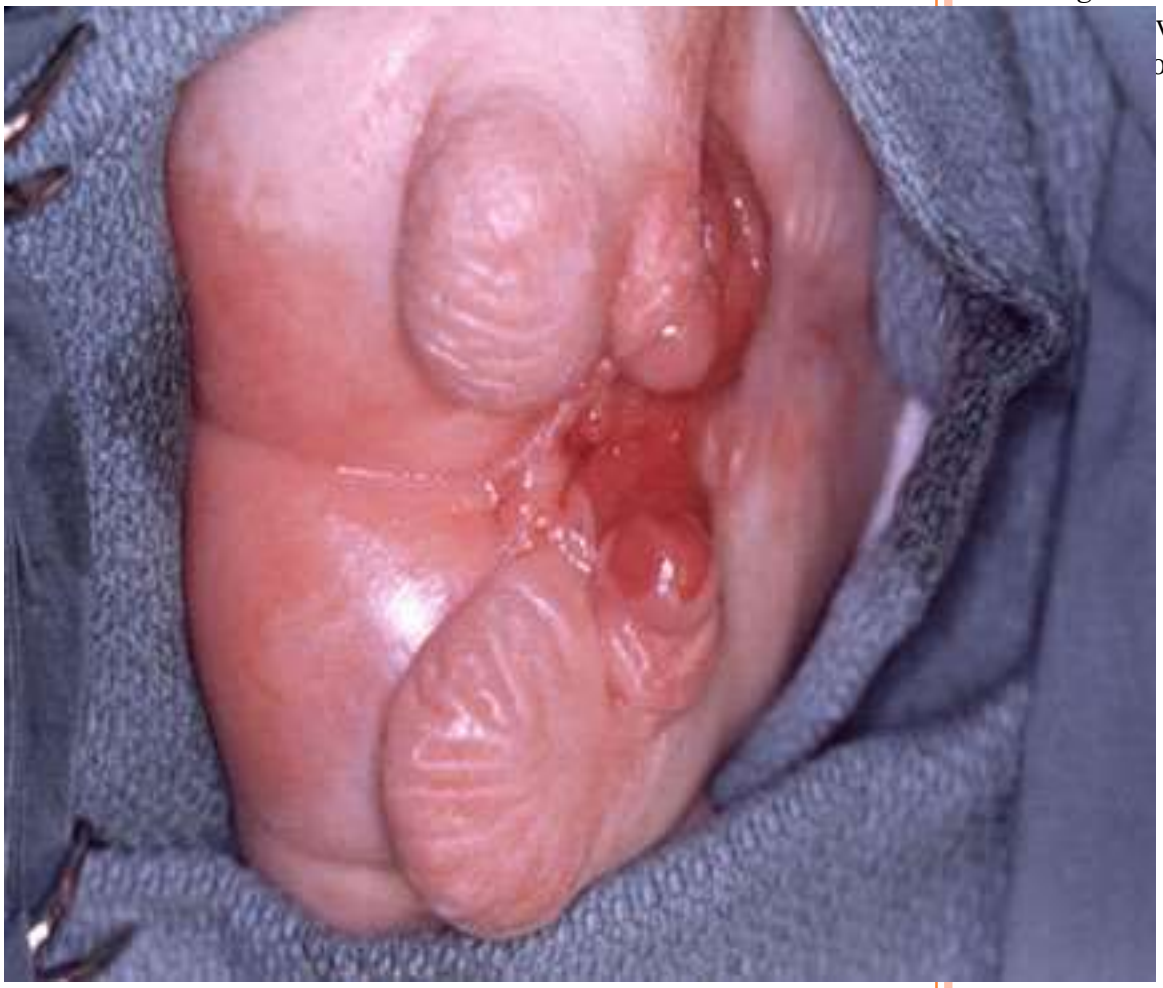
Case A. The flank wounds were skin grafted and closure of the mphalocele d.



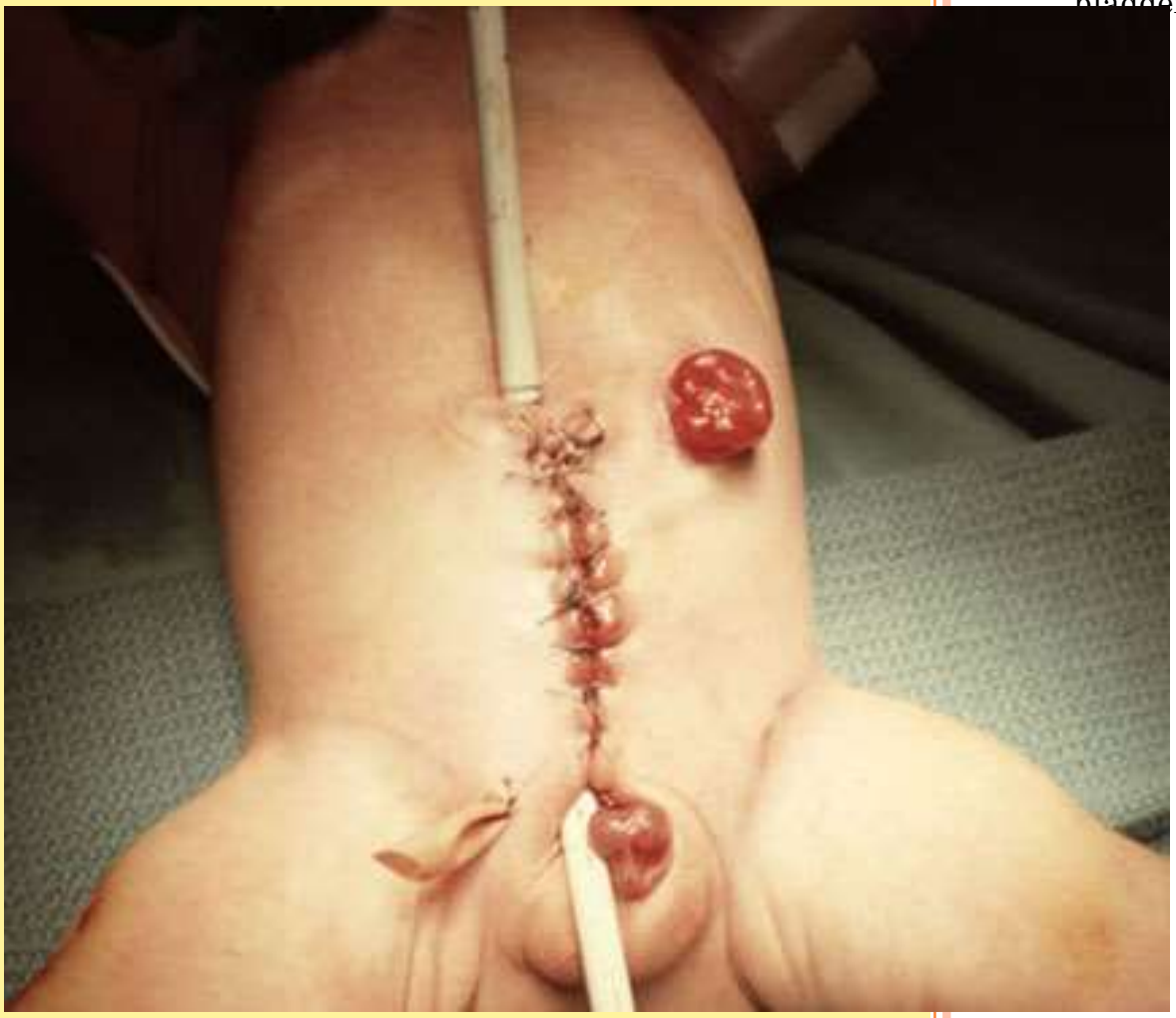
Baby with cloacal exstrophy.

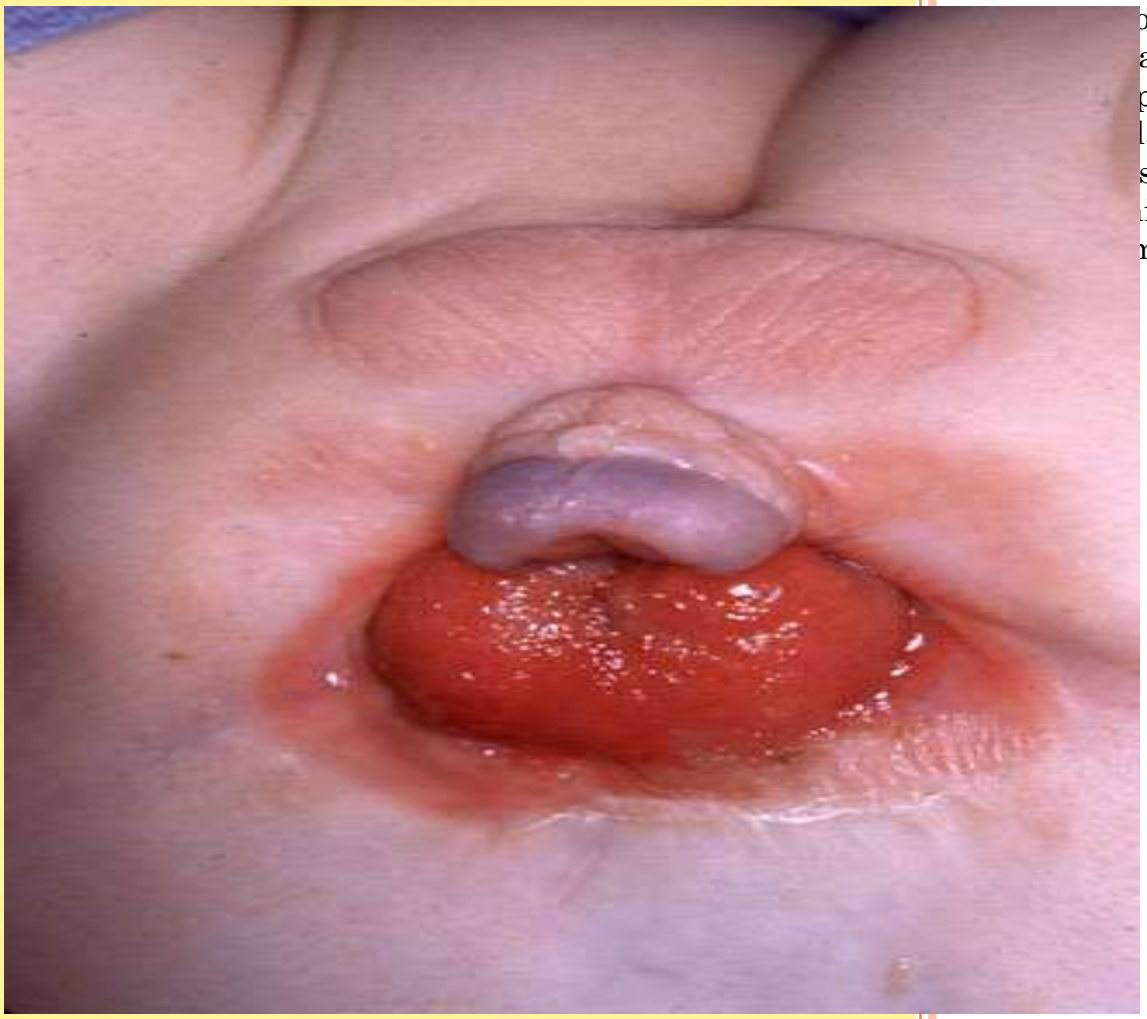


Note the bifid genitalia in this with cloacal ophy.



Closure of the bladder exstrophy.





Baby with bladder
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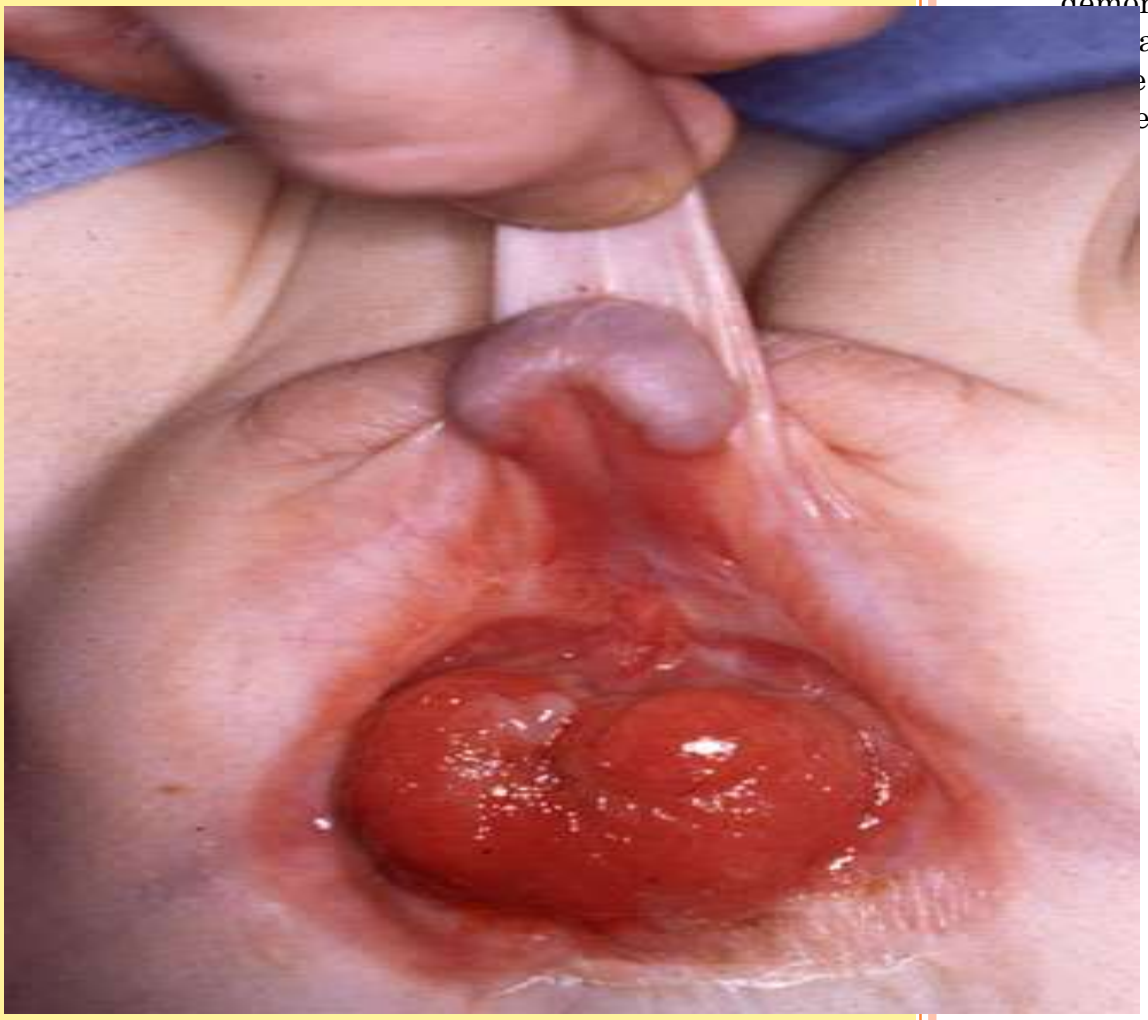
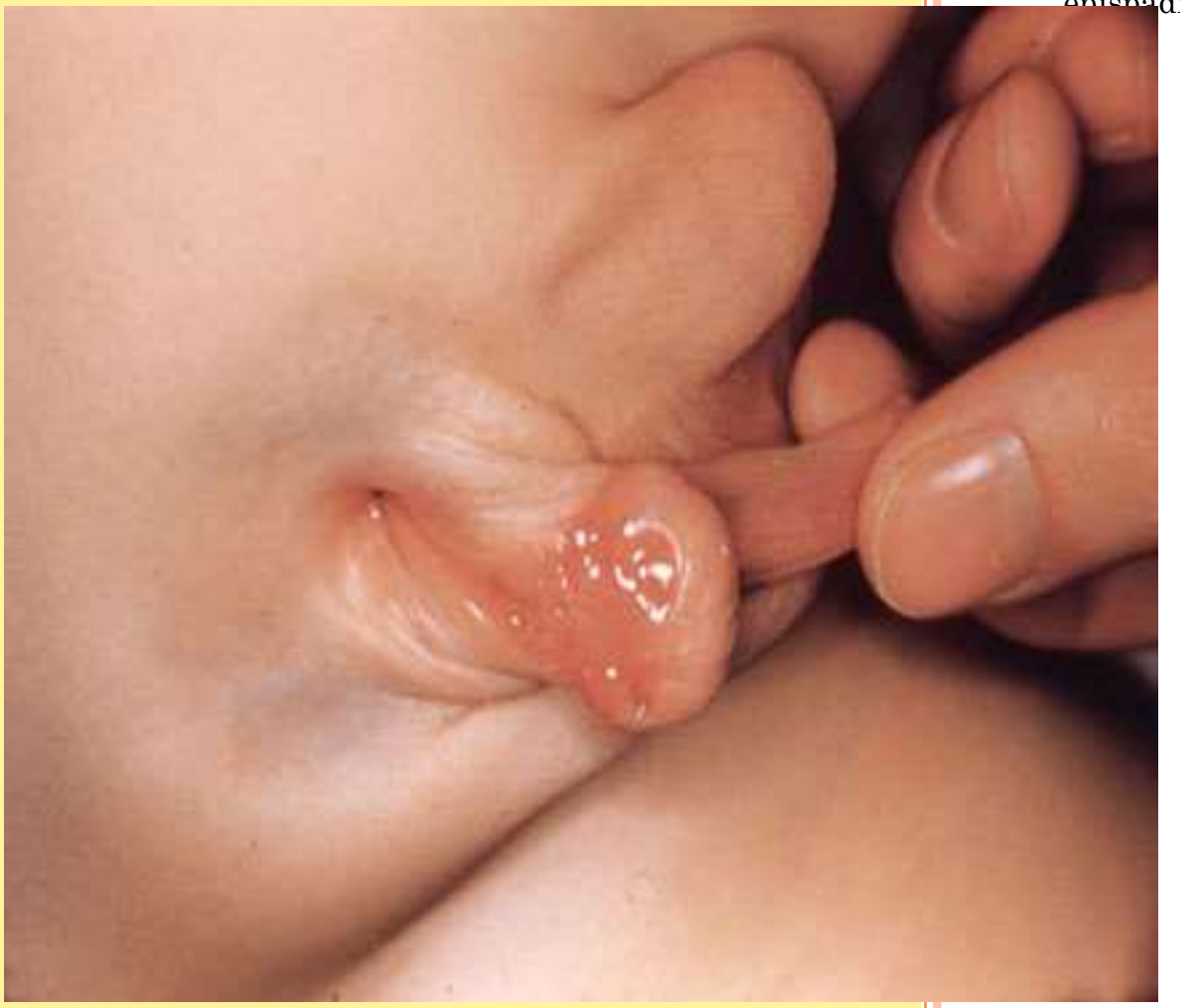
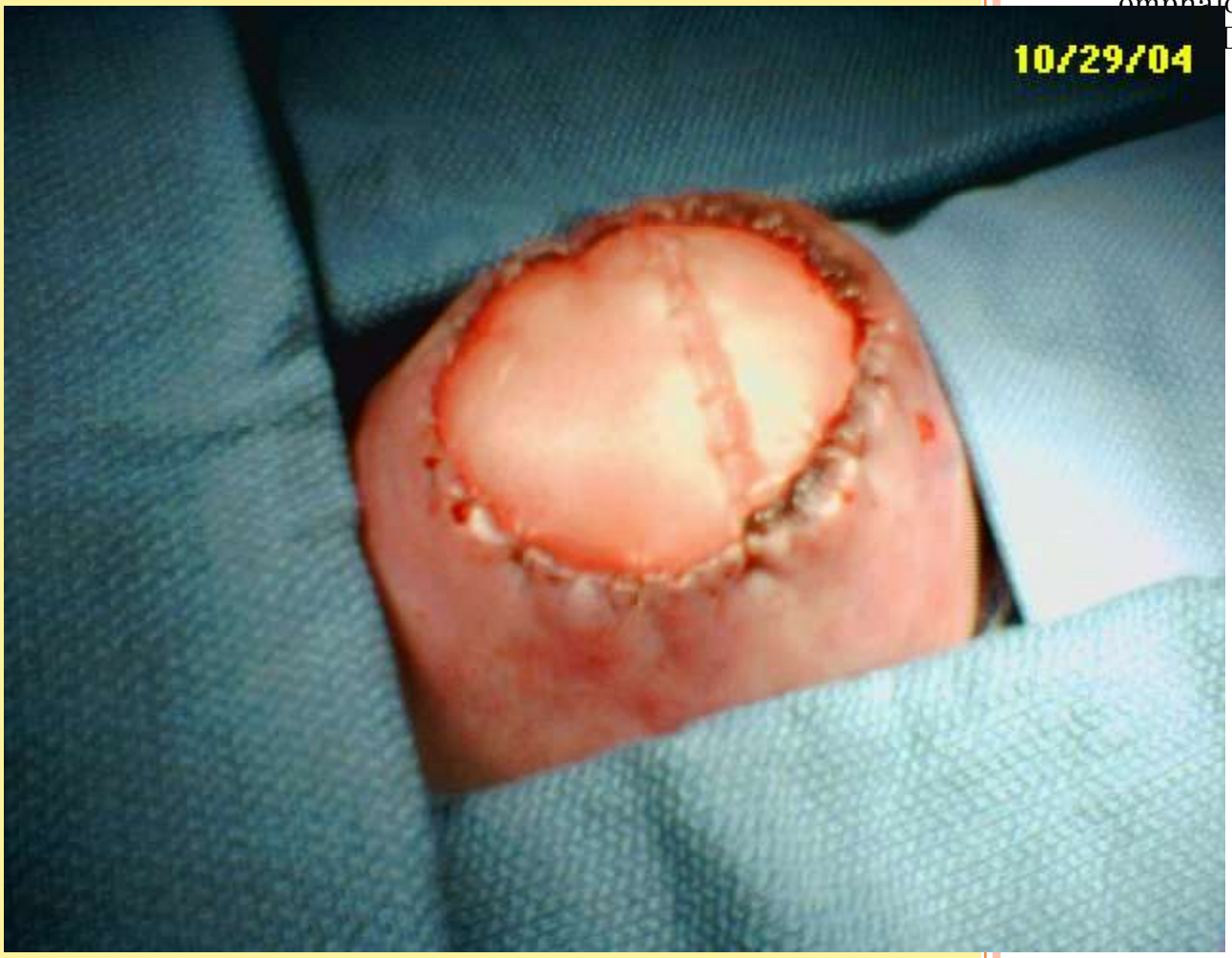
Another view demonstrating the adias shown previous e.



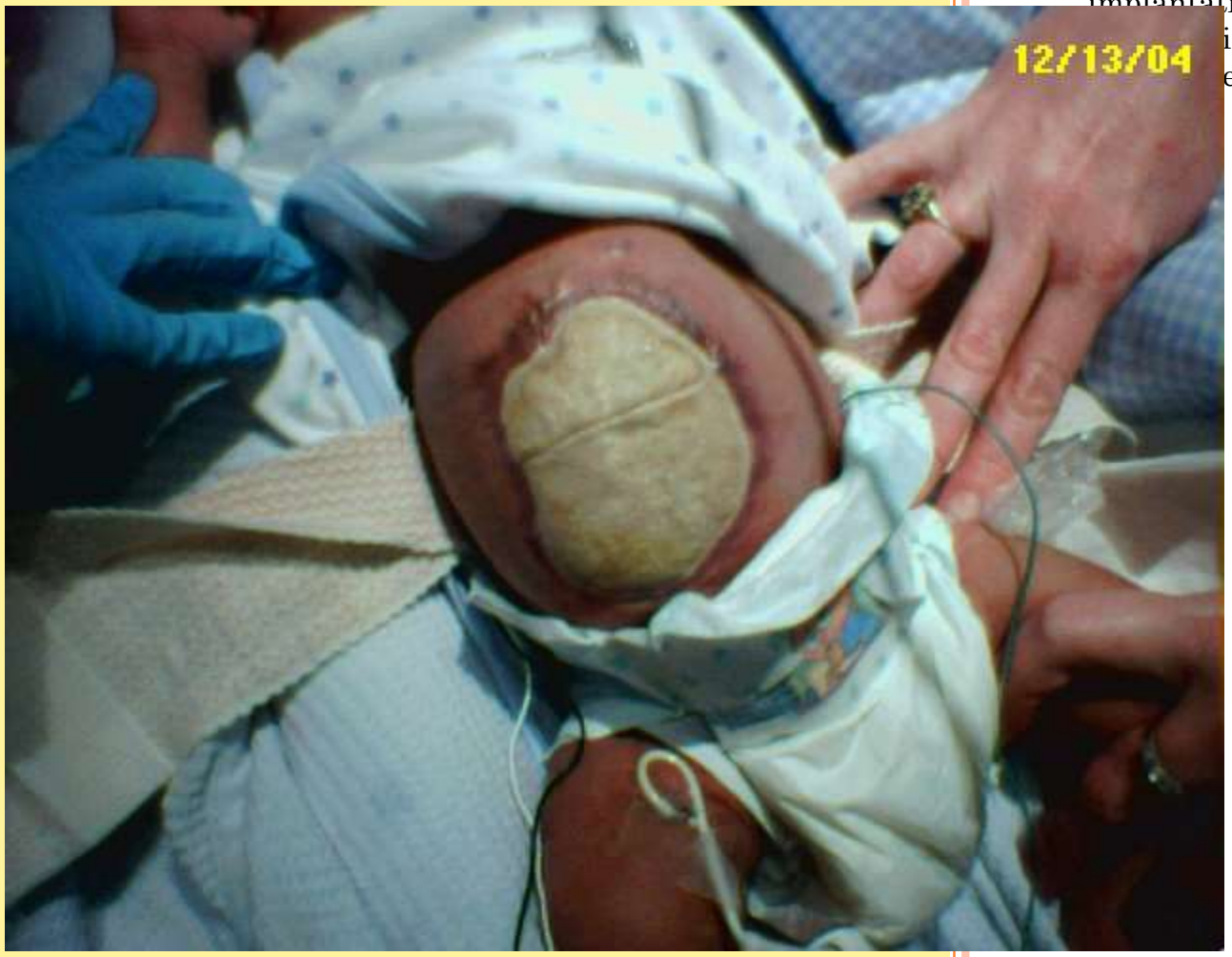
Baby with isolated epispadias.



Closure of a giant omphalocele with Derm patch



Two months after implantation: ization of erm



Eight months
after implantation:
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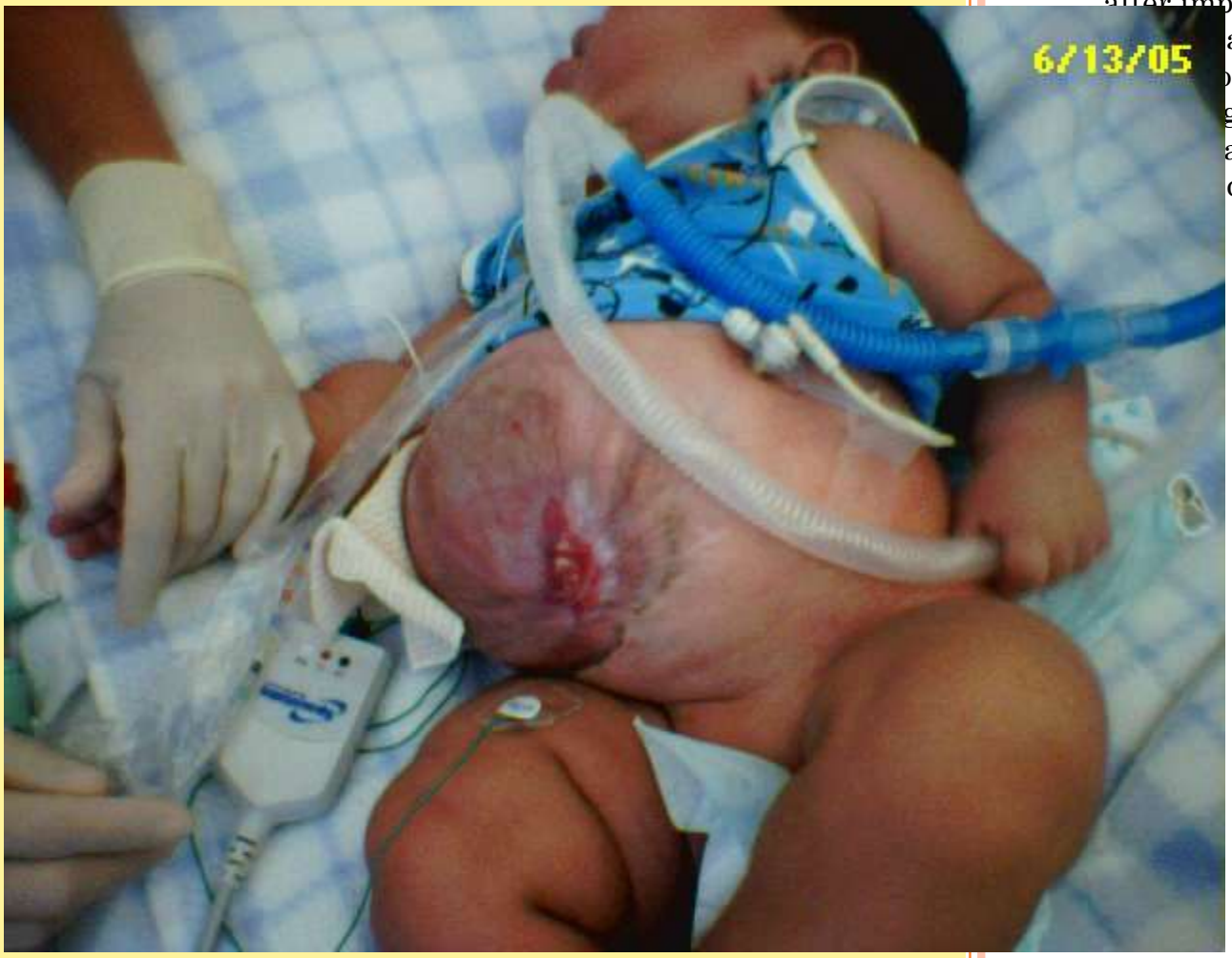
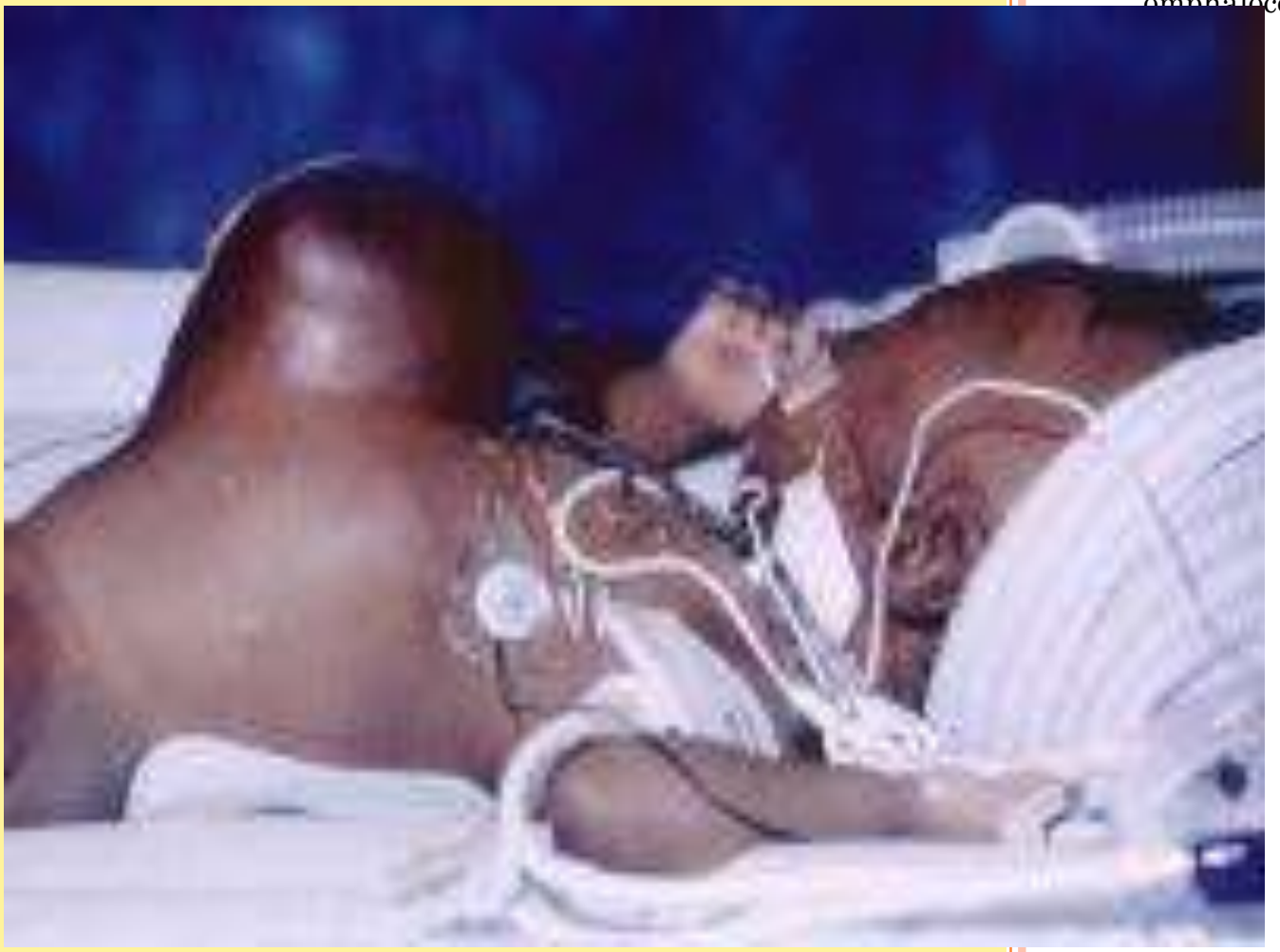
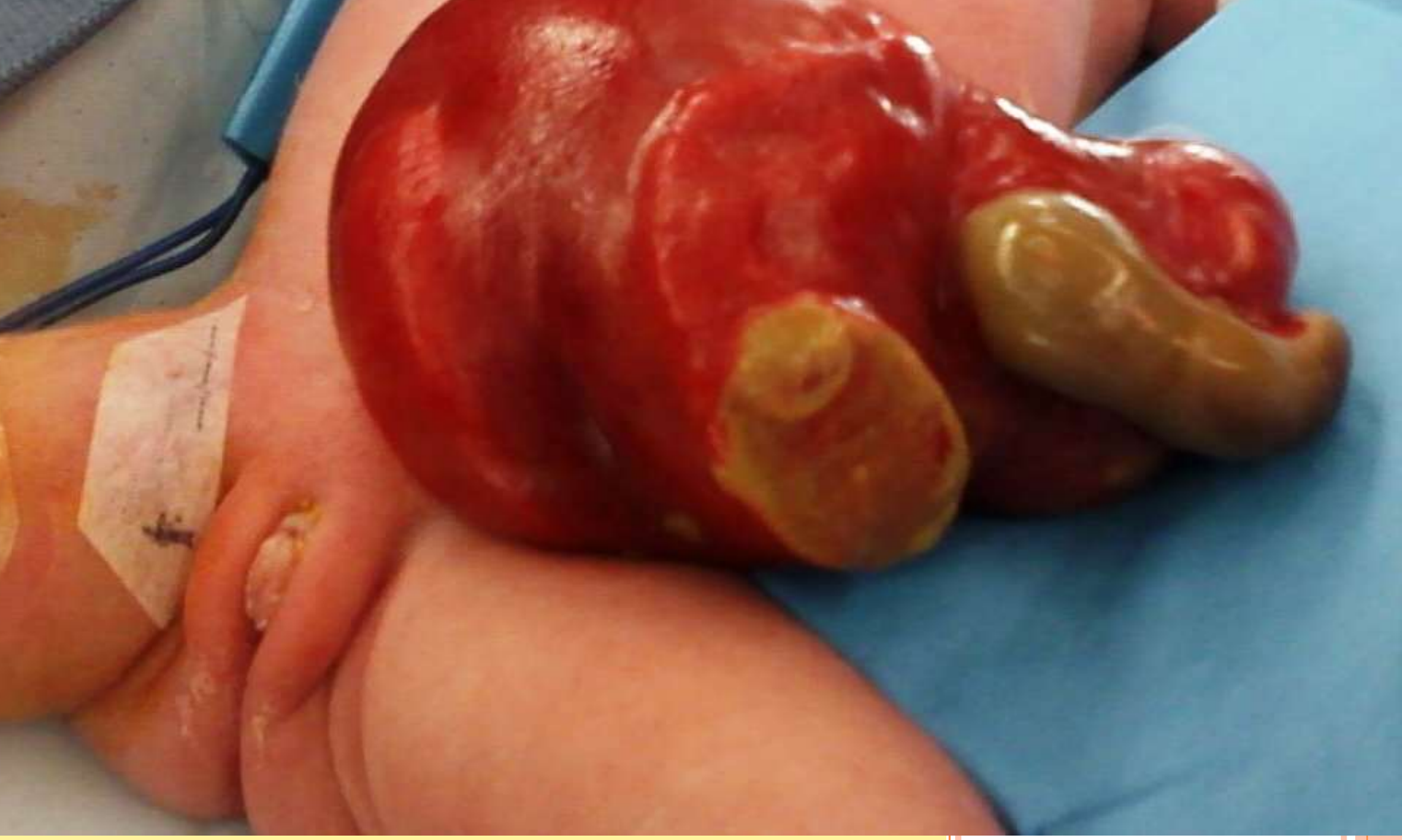
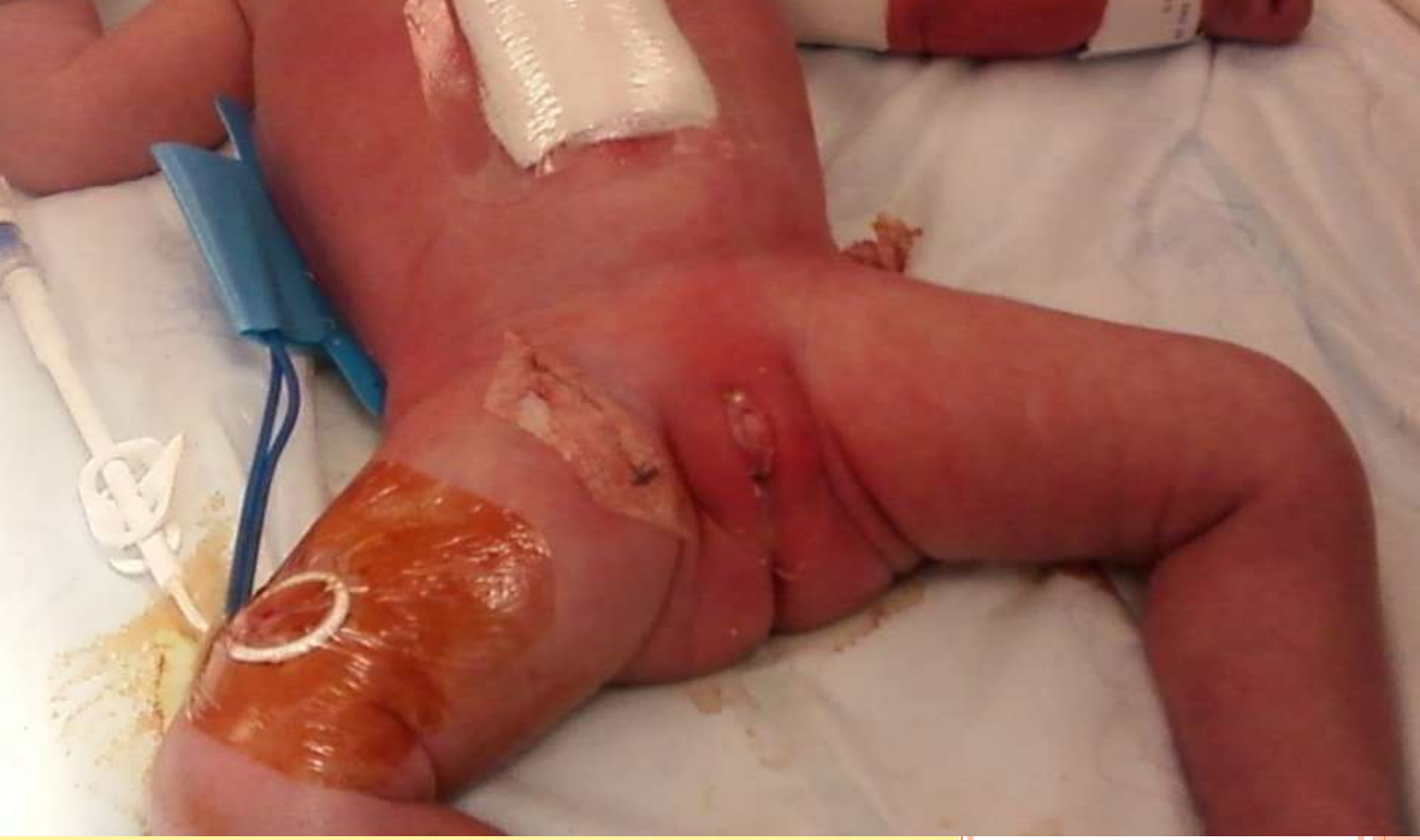


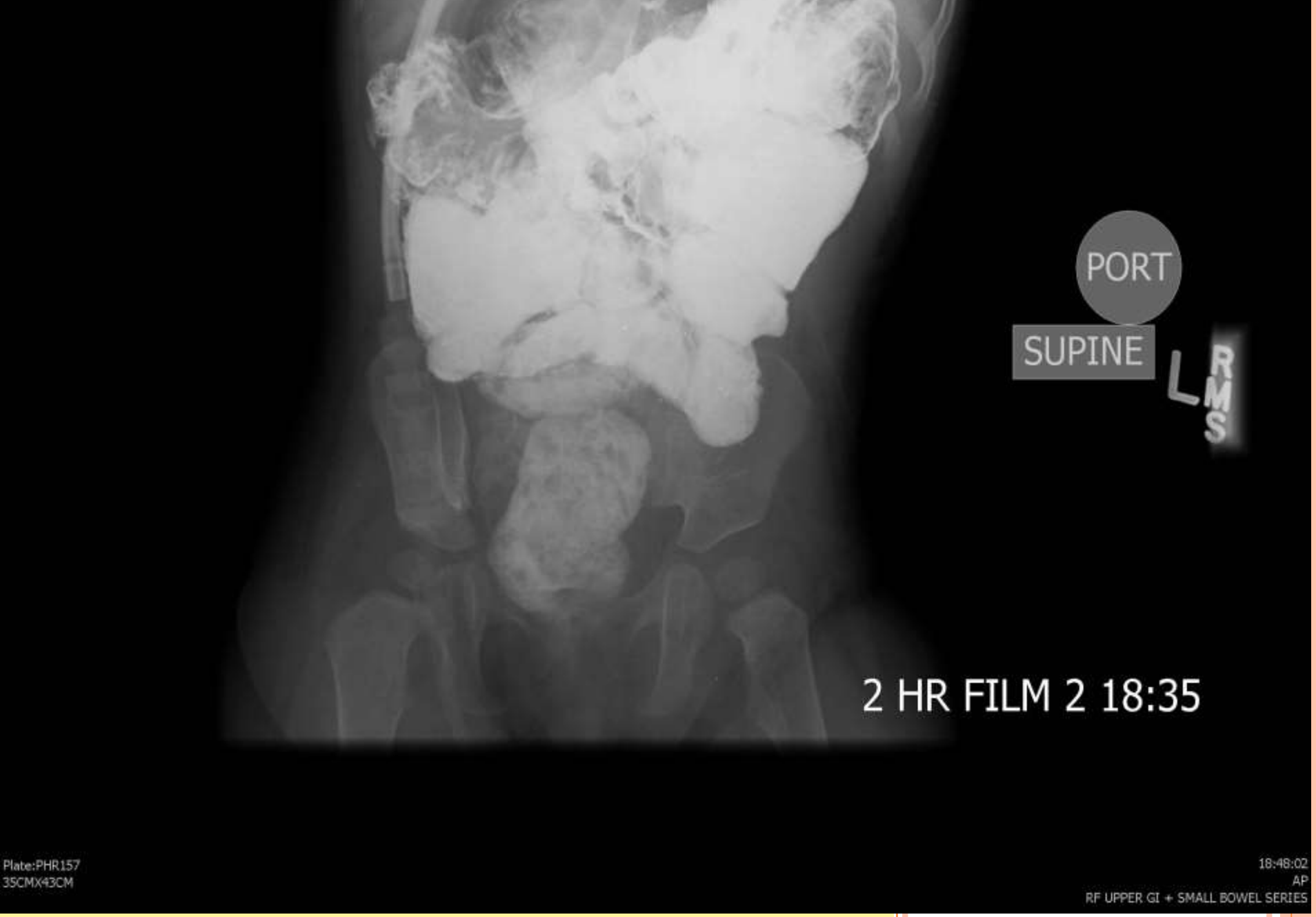
Baby with an omphalocele.





Complicated gastroschisis.




PORT
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2 HR FILM 2 18:35
Plate:PHR157
35CMX43CM
18:48:02
AP
RF UPPER GI + SMALL BOWEL SERIES

MALROTATION

Errors of Midgut Development and rotation

- **Non-rotation:** leaving the major part of the colon on the left side and the small intestine to the right of the midline
- **Incomplete rotation:** the coecum is situated in the sub-hepatic region
- **Reversed rotation:** the final 180º rotation occurs in a clockwise manner so that the colon is lying posterior to the duodenum and the superior mesenteric artery
- **Hyper-rotation:** the rotation continues to 360º or 450º so that the coecum rests in the region of the splenic flexure.

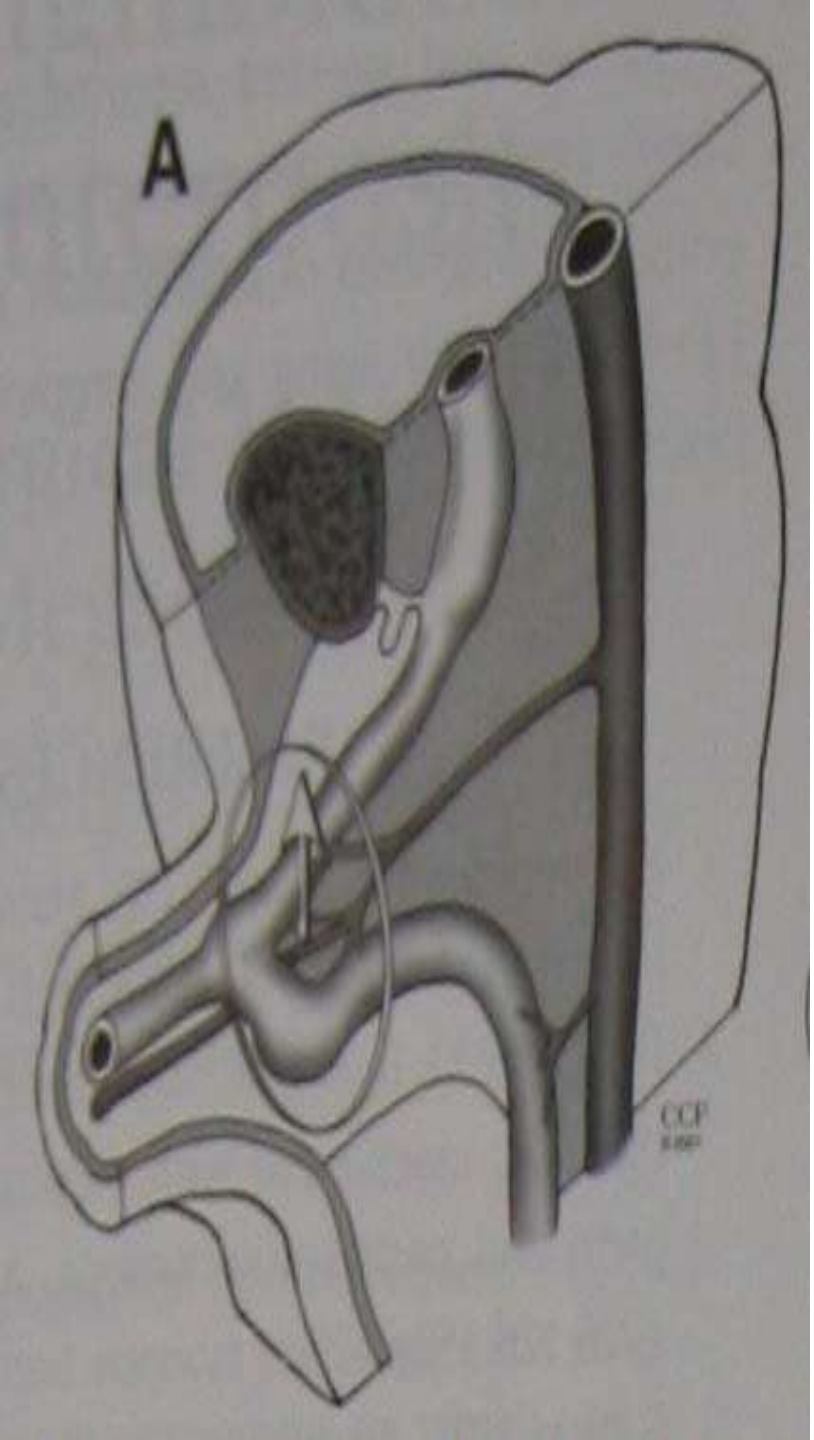
- Normal rotation of the human intestine requires transformation from a simple, straight alimentary tube into the mature fixed and folded configuration present at birth.

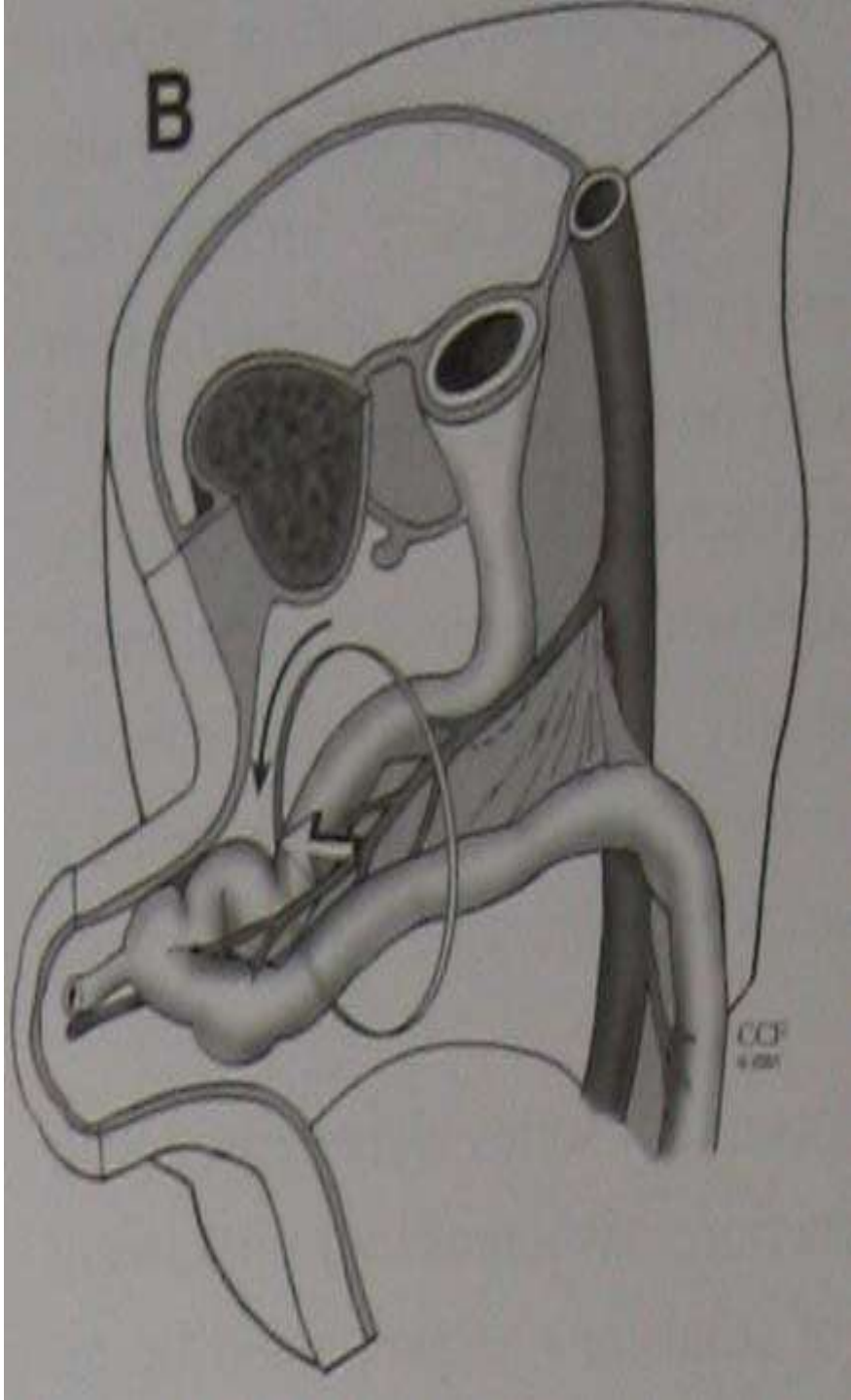
- The duodenojejunal junction becomes fixed in the left upper abdomen while the cecum is anchored in the right lower quadrant.

- The midgut, defined as the portion of the intestine supplied by the superior mesenteric artery, is thus suspended from a wide mesenteric base.

EMBRYOLOGY

- The development of the midgut begins with the differentiation of the primitive intestinal tract into the foregut, midgut, and hindgut at the fourth week of gestation.
- The mature alimentary tract and all associated digestive organs are formed from this primitive tube.
- The most accepted model of midgut maturation involves four distinct stages:
  - (1) herniation.
  - (2) rotation.
  - (3) retraction.
  - (4) fixation
- The intestinal loop can be divided into the cephalic (duodenojejunal) limb and the caudal (cecocolic) limb, which rotate separately but in parallel.


A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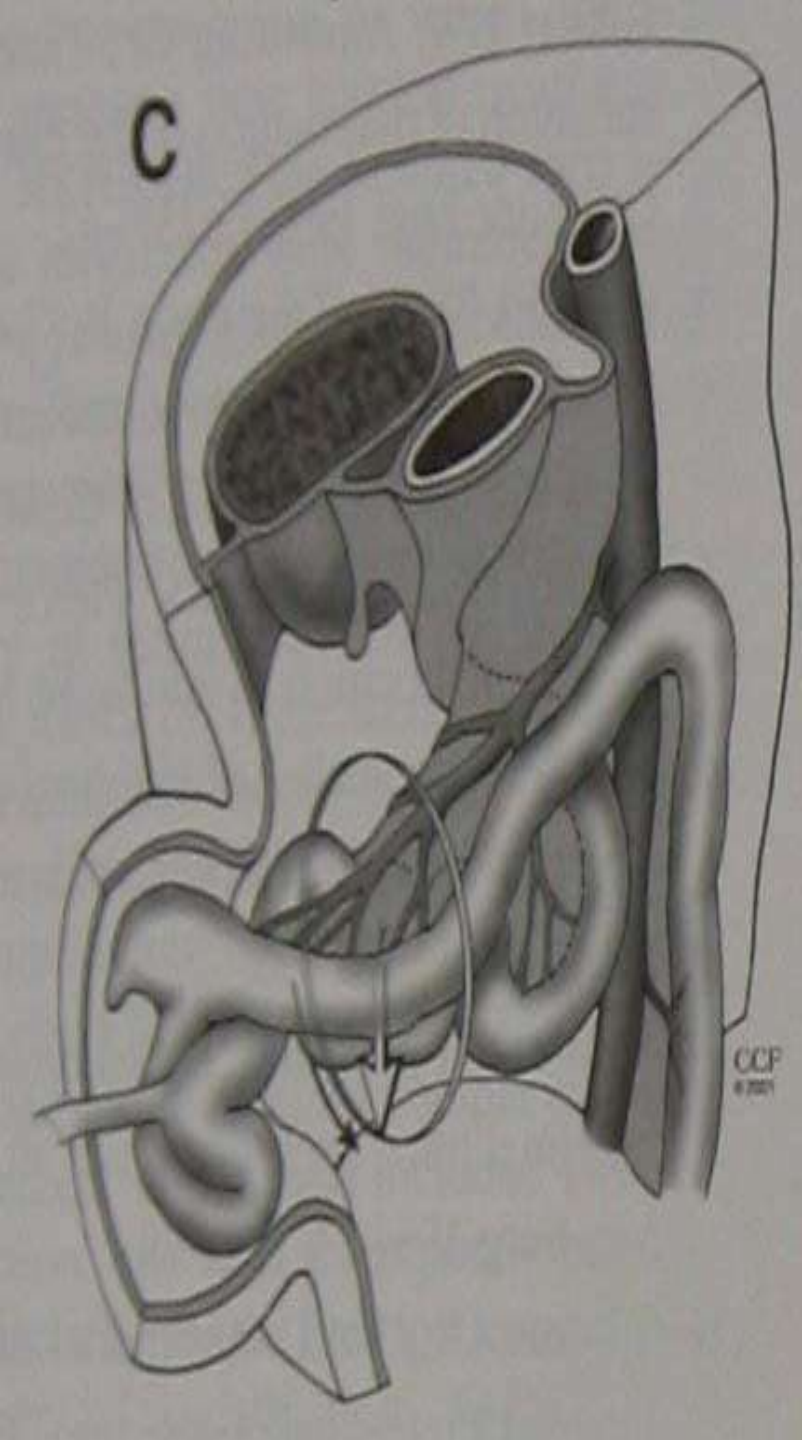
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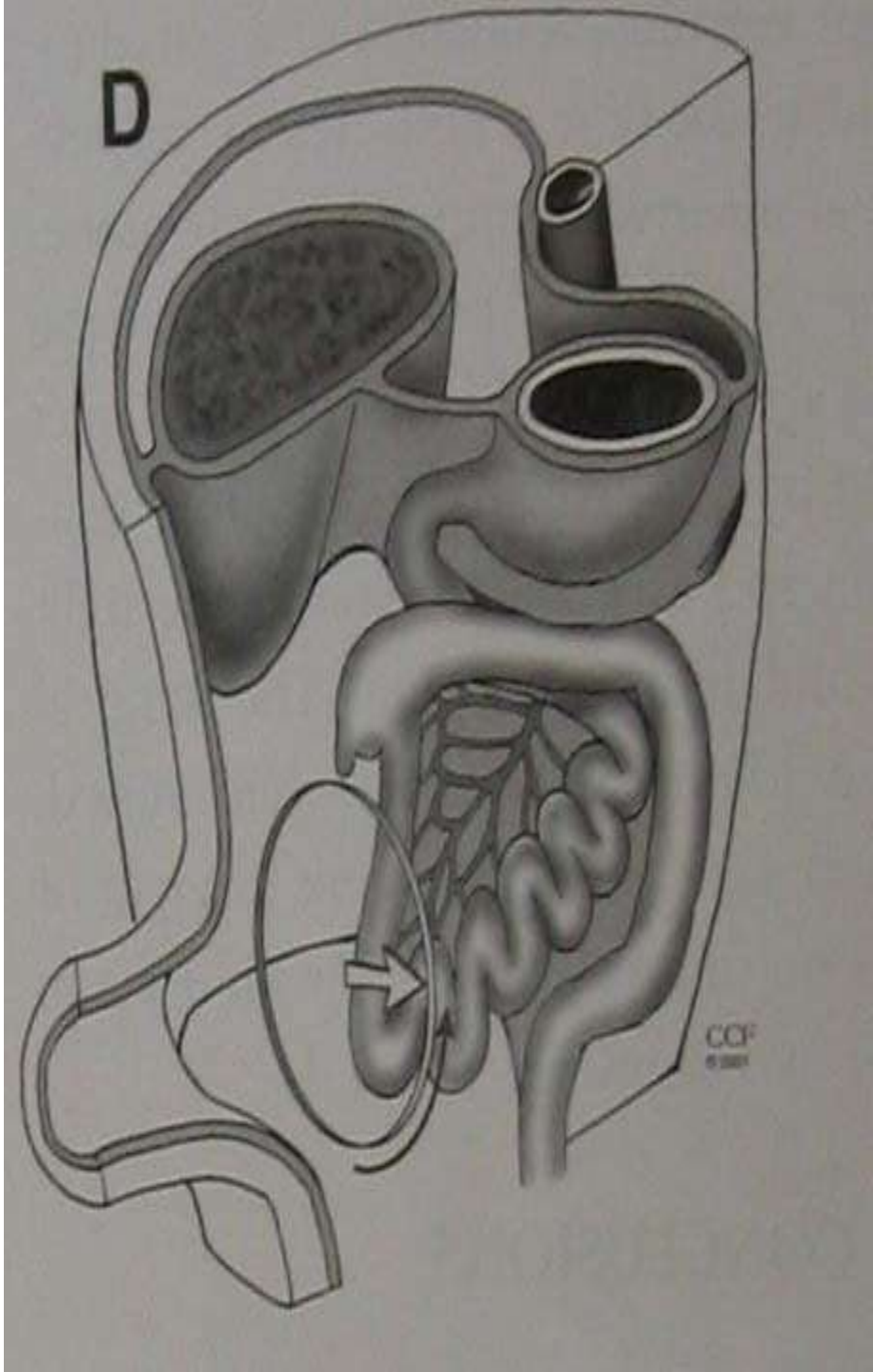
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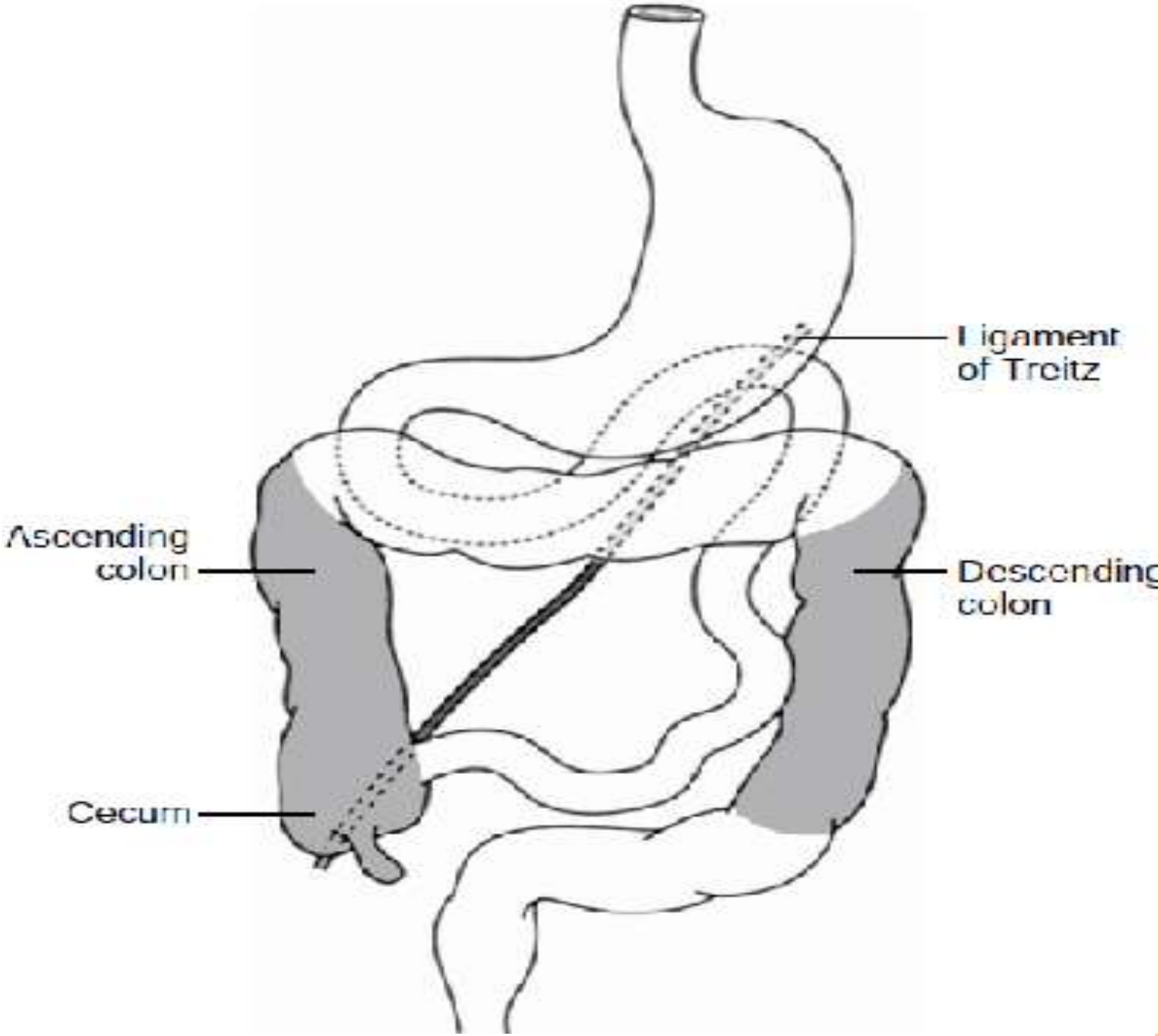
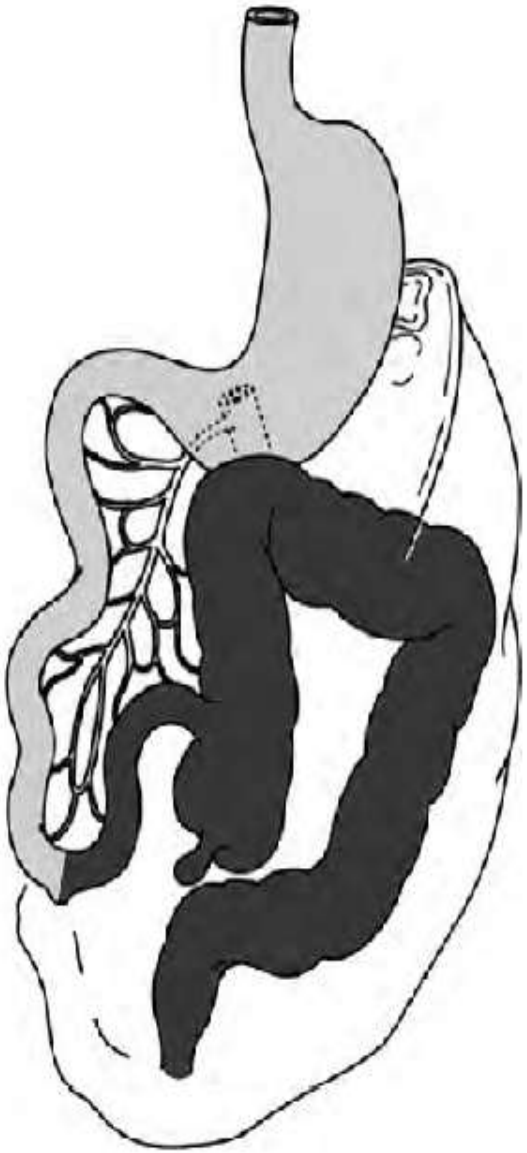
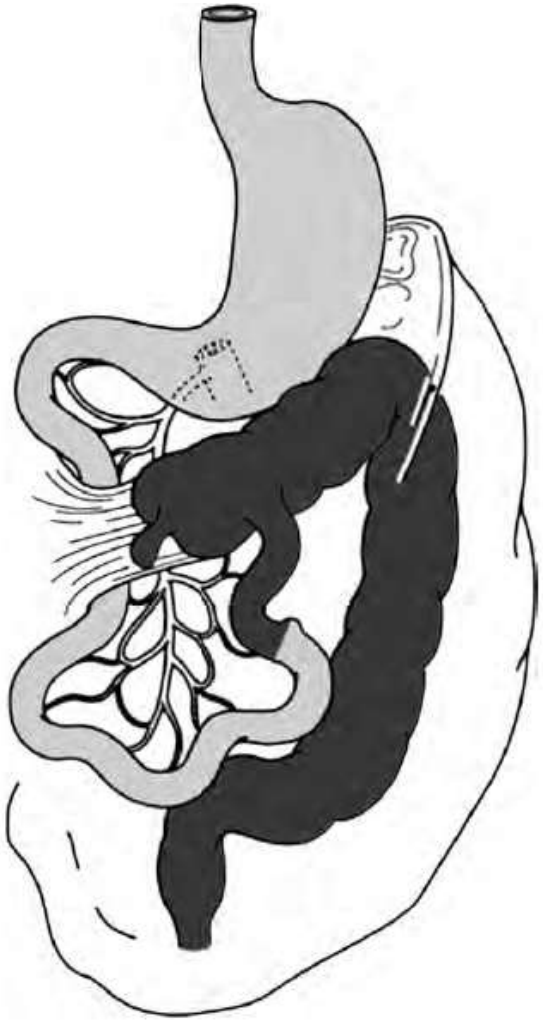

Ligament of Treitz
Ascending colon
Descending colon
Cecum

Non rotation



Incomplete rotation



PRESENTATION

- The incidence of malrotation has been estimated at 1 in 6000 live births.
- Up to 75% of patients present during the first month of life.
- Another 15% will present within the first year.
- Volvulus, intestinal gangrene, and mortality have been noted regardless of the patient's age or chronicity of symptoms.
- Bilious vomiting remains the cardinal sign of neonatal intestinal obstruction, and malrotation must be the presumed diagnosis until proven otherwise.

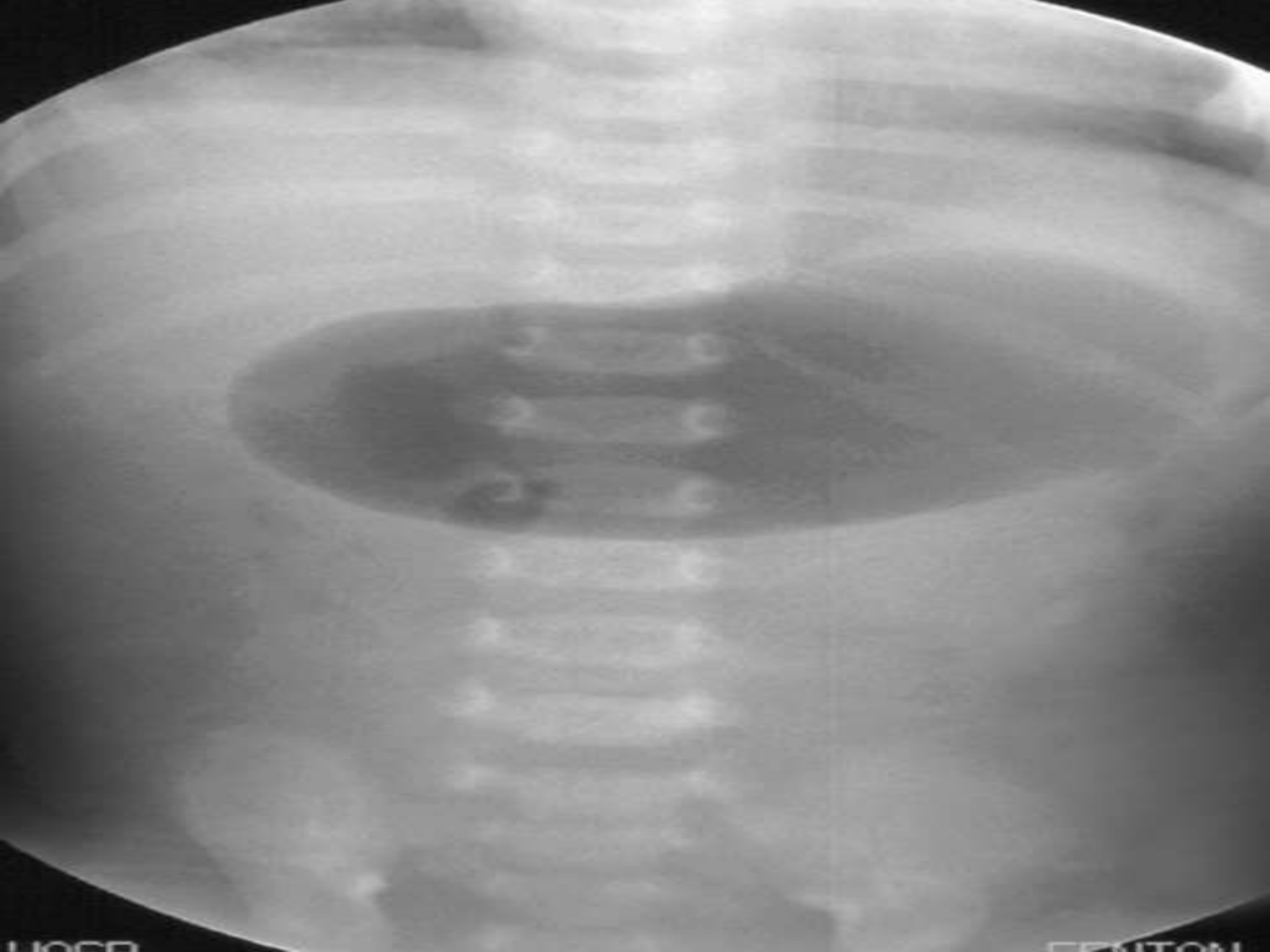
- Other signs in the neonate include abdominal pain and distention.
- The inconsolable infant may rapidly deteriorate as metabolic acidosis quickly advances to hypovolemic shock.
- Late signs include abdominal wall erythema and hematemesis or melena from progressive mucosal ischemia.
- Many other cases will present less dramatically.
- Failure to thrive, gastro esophageal reflux, early satiety, and mild abdominal discomfort are routinely reported.

Table 32-1	Incidence of Associated Anomalies (by percent) with Malrotation
Intestinal atresia	5-26%
Imperforate anus	0-9%
Cardiac anomalies	7-13%
Duodenal web	1-2%
Meckel's diverticulum	1-4%
Hernia	0-7%
Trisomy 21	3-10%

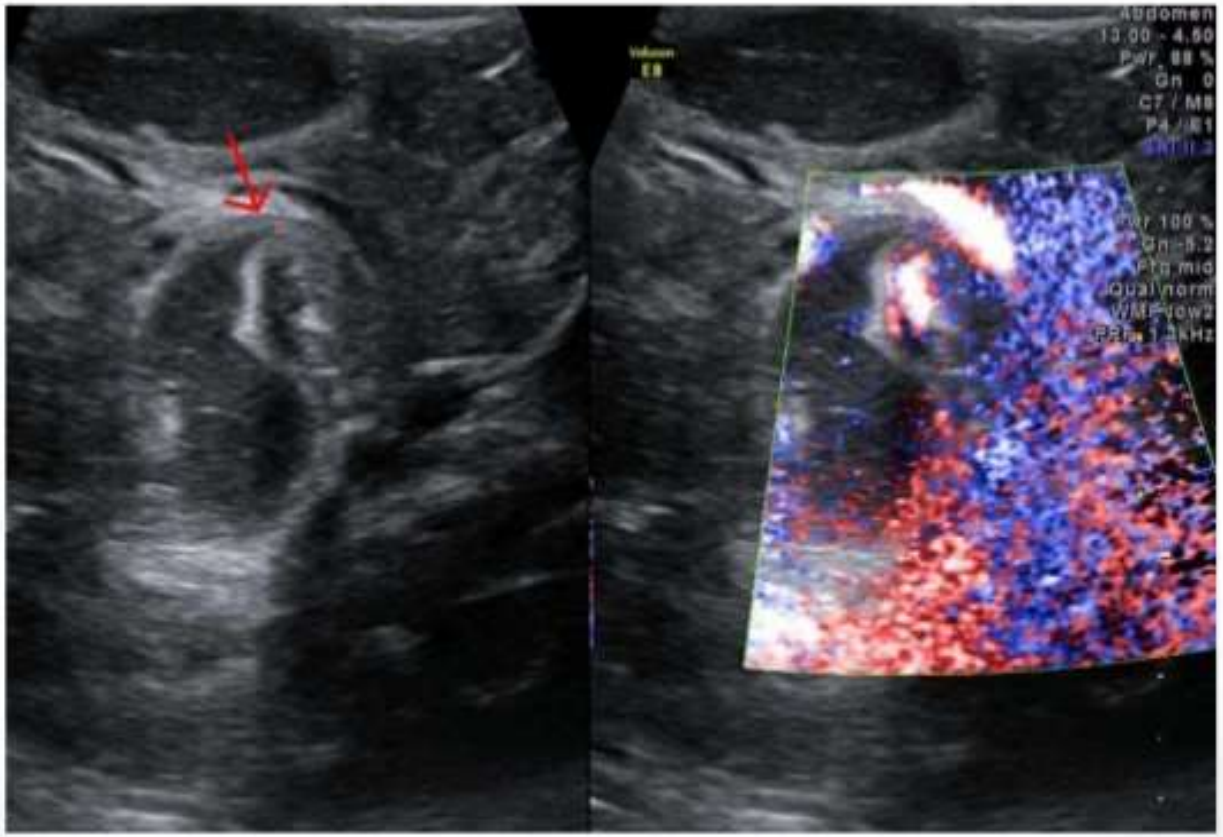
Rare: esophageal atresia, biliary atresia, mesenteric cyst, craniocynostosis, Hirschsprung's disease, intestinal duplication.

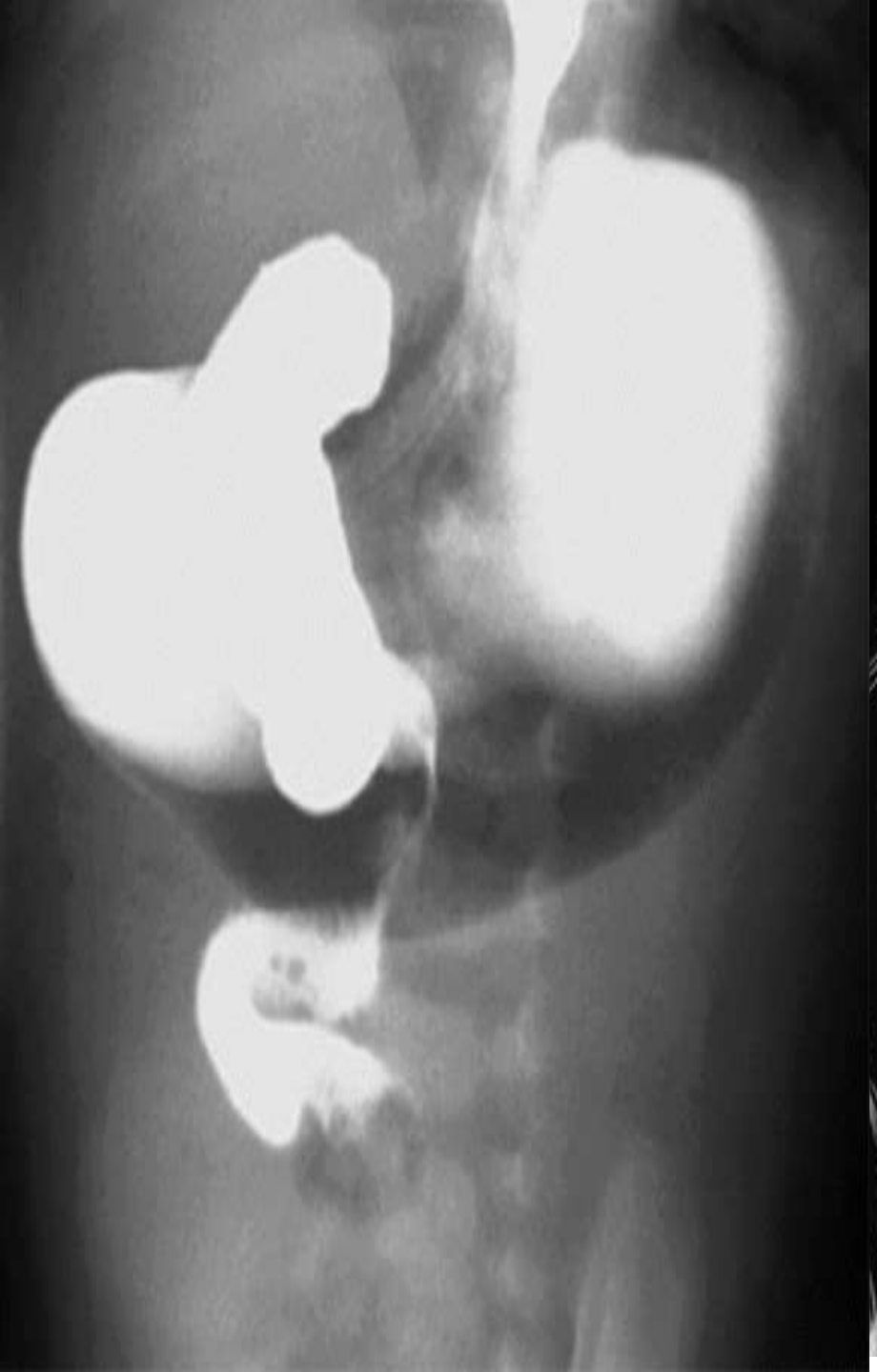
DIAGNOSIS

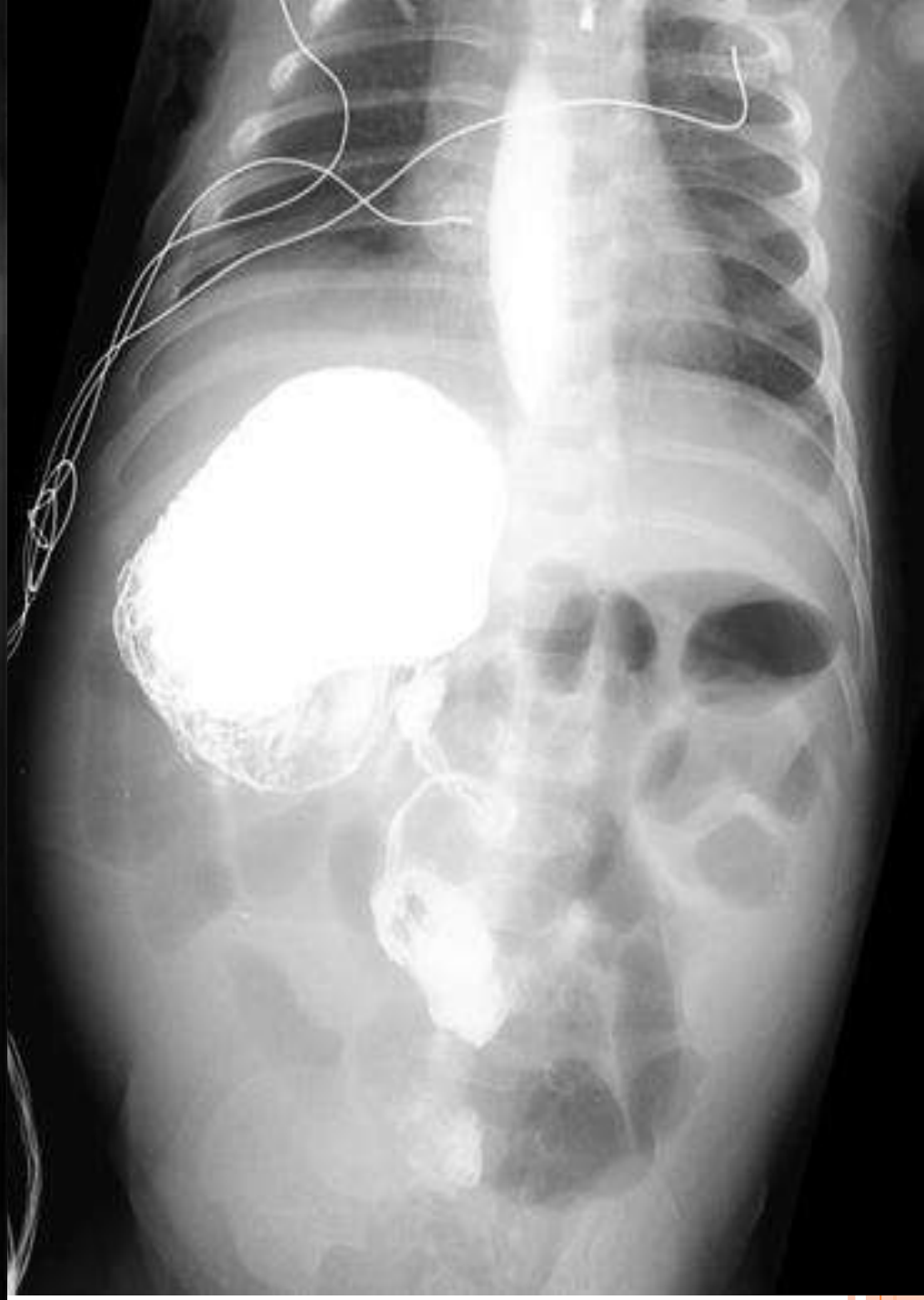
- Plain X-Ray.
- Doppler study.
- Upper contrast study.
- CT- scan.



Colour doppler: whirlpool sign



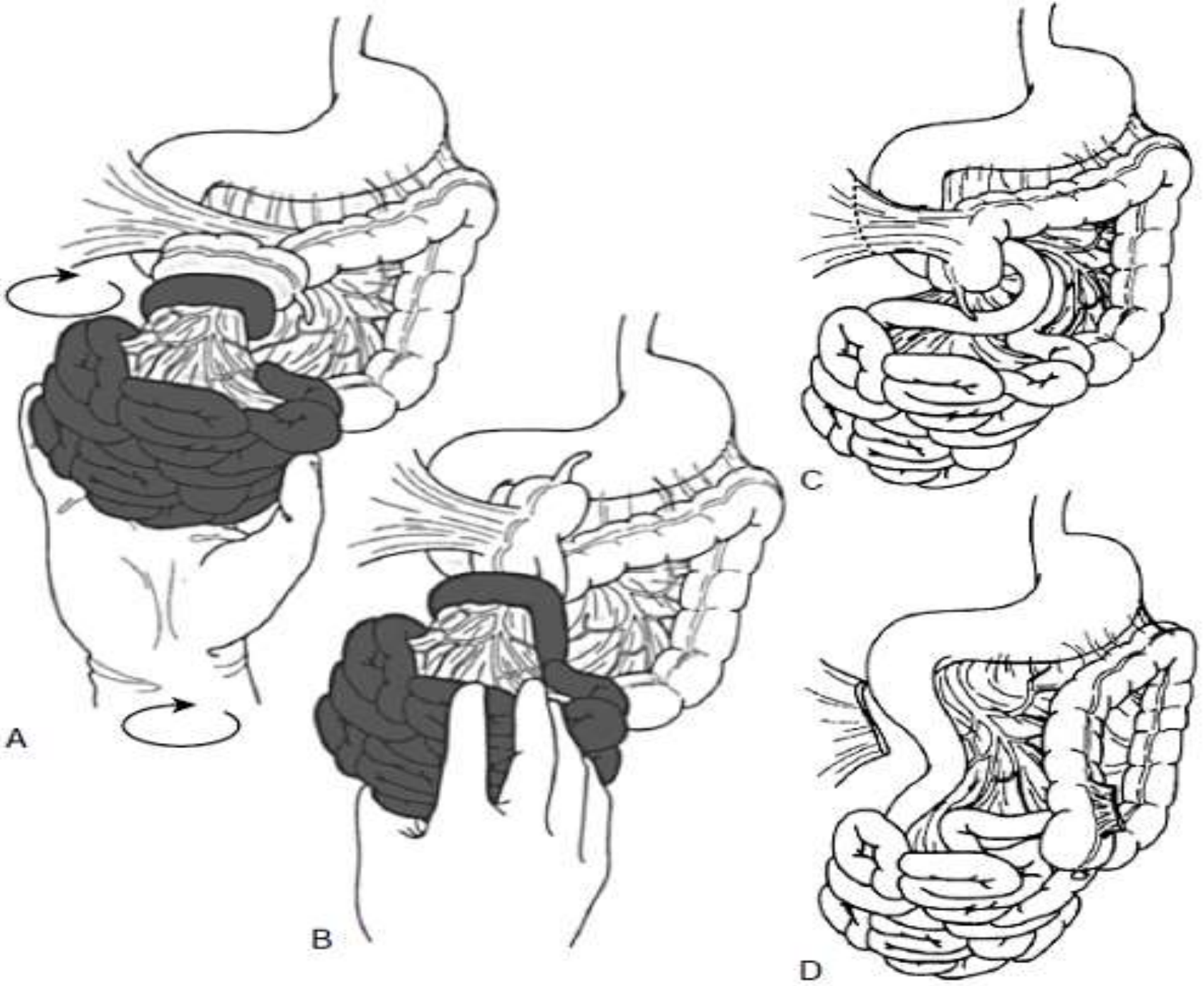




RT

Treatment: (Ladd's procedure)

- ***The aim of surgery:***
- 1. Entry into abdominal cavity and evisceration (open)
- 2. Counterclockwise detorsion of the bowel (acute cases)
- 3. Division of Ladd's cecal bands
- 4. Broadening of the small intestine mesentery
- 5. Incidental appendectomy
- 6. Placement of small bowel along the right lateral gutter and colon along the left gutter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LAPAROSCOPY

- The laparoscopic treatment for intestinal rotation anomalies in neonates, infants, and children with or without midgut volvulus has been proposed by several authors since van der Zee's original report in 1995.
- Reverse trendelenberg position
- 4 port technique used.

Summary

- Rotational anomalies are the result of arrest of normal rotation of the embryonic gut
- Early diagnosis and surgical intervention reduces morbidity and mortality .
- Must have a high index of suspicion for infants with bilious vomiting.
- Malrotation, regardless of age and presence of symptoms is treated surgically with Ladd procedure