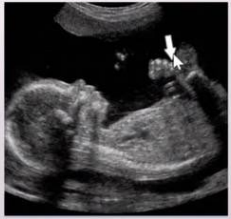
Congenital Abdominal Wall Defects (Omphalocele and Gastroschisis)

Gastroschisis

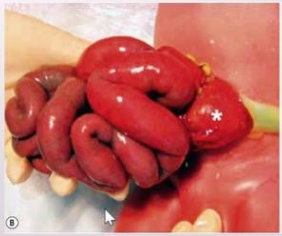
- 1 in 4000 live births
- Higher incidence in mothers younger than 21 years of age
- Diagnosis : AN US by 20 weeks' gestation

Bowel loops **freely floating in the amniotic fluid** and a defect in the abdominal wall to **the right of a normal umbilical cord**) **+abnormal maternal serum α-fetoprotein (AFP) level,** which is universally elevated +/-Intrauterine growth restriction (IUGR)

- Delivery should be in a tertiary centre



- Gastroschisis is associated with a variable degree of inflammatory thickening of the visceral bowel walls, which results in the characteristic appearance of "matted" intestines
- Associated with intestinal motility disorder , rotational disease , UDT 15-25% , bowel atresia
- Simple VS complicated (atresia ,Short bowel)



Management

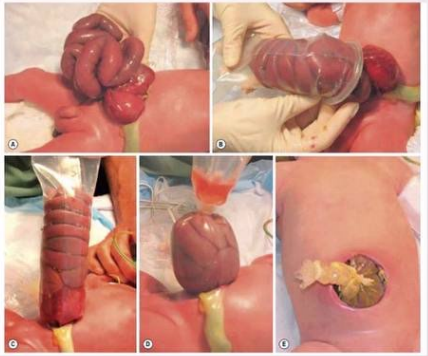
- Resuscitation

(NPO, NG, IVF, rectal tube to decompress)

- bowel should be wrapped in warm saline-soaked gauze and placed in a central position on the abdominal wall
- Surgery :

Either Primary closure or Staged closure

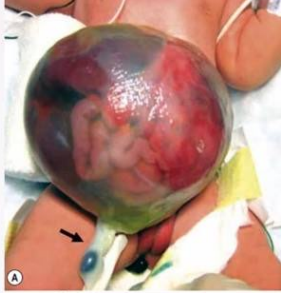
(with silo)



Long term outcome

- Long-term outcomes for infants born with gastroschisis are generally excellent
- Morbidities related to prematurity , bowel motility and length

Omphalocele



- 1 in 4000–6000.
- Associated with genetic defect and other anomalies (Trisomies 13, 18, 21, and 45 X ,Beckwith–Weideman, pentalogy of Cantrell , cardiac (14–47% incidence of anomalies) and central nervous (3–33% anomalies)
- Outcomes depend on associated anomalies
- Long term morbidities : gastroesophageal reflux disease (GERD), pulmonary insufficiency, recurrent lung infections or asthma, and feeding difficulty with failure to thrive

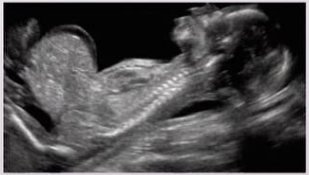
Diagnosis

Antenatally :

- ¬ 18-week US evaluation , elevated AFP

(prognostic factor :omphalocele diameter compared with abdominal circumference (O/AC, or omphalocele ratio), the femur length (O/FL), and the head circumference (O/HC), , organ contained inside the sac

- Deliver in a tertiary centre , at term , normal vaginal delivery (except if it is giant omphalocele and containing liver (to avoid shoulder dystocia , sac rupture and bleeding)



Management

- **Resuscitation**

NPO, NG, IVF, rectal tube to decompress)

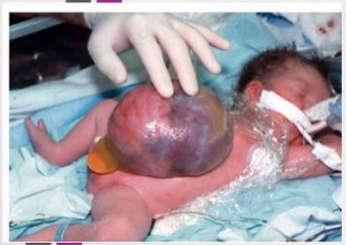
- **sac should be wrapped in warm saline-soaked gauze** and placed in a central position on the abdominal wall

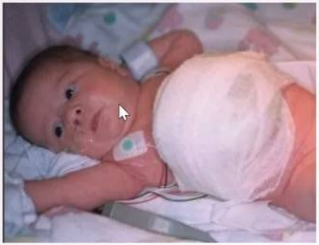
- **Surgery :**

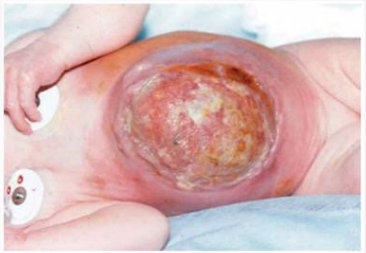
Primary closure : in small defect , consists of excision of the sac and closure of the fascia and skin over the abdominal content

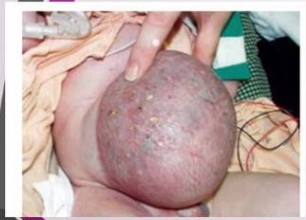
Staged closure using a mesh or using a silo with serial reduction then closure

paint and wait/ Scarification technique), in case of giant omphalocele , associated cormobidities)









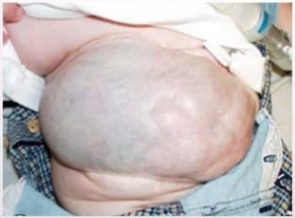


Table 48.1 Differentiating Characteristics Between Gastroschisis and Omphalocele

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

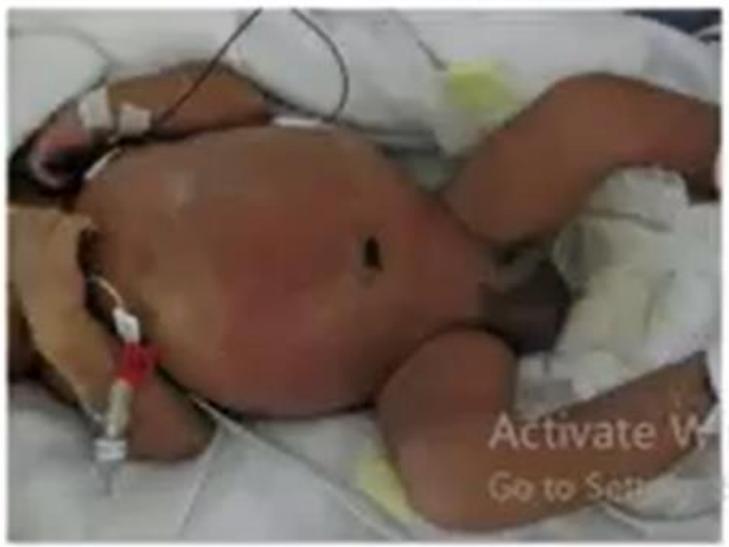
Necrotizing Enterocolitis (NEC)

- Disease of premature neonates.
- NEC affects about 10% of VLBW
- Incidence is inversely proportional to birth weight.
- The overall mortality of NEC probably approaches 30%
- Lower birth weight and younger gestational age correlate with higher risk of death.

Presentation

- NEC presents with feeding intolerance(vomiting or high gastric residuals), abdominal distention. Bleeding per rectum is a late sign .

- On exam : Abdominal distention
  - visible +/- palpable dilated bowel loops
  - skin discoloration
  - signs of peritonitis



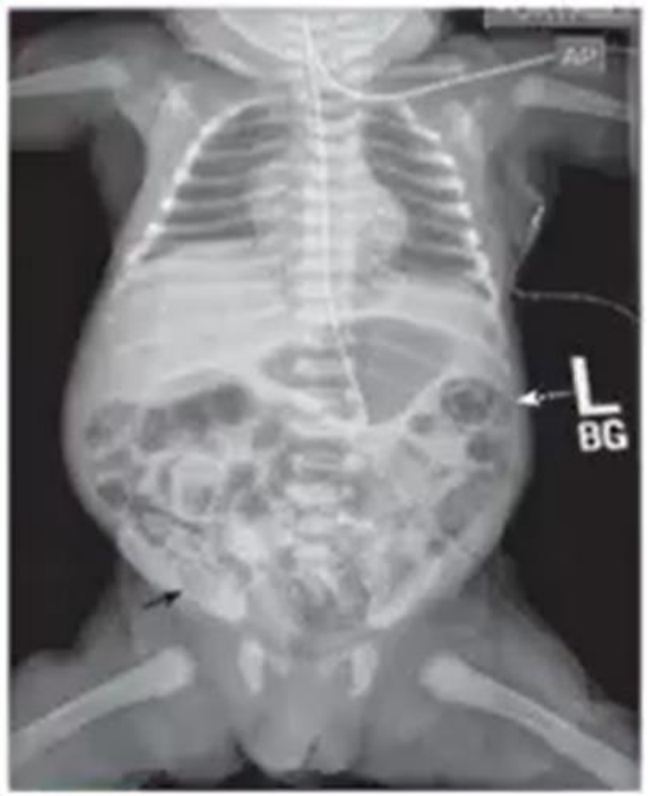
Diagnosis

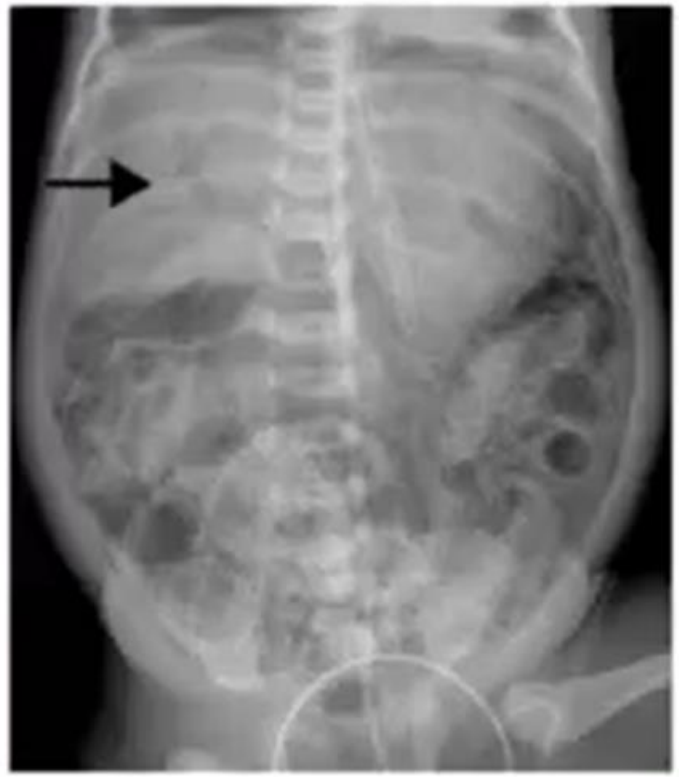
- Clinical and radiological
- History and physical examination
- Lab : High WBC, CRP , hyponatremia , thrombocytopenia , high lactate , metabolic acidosis .

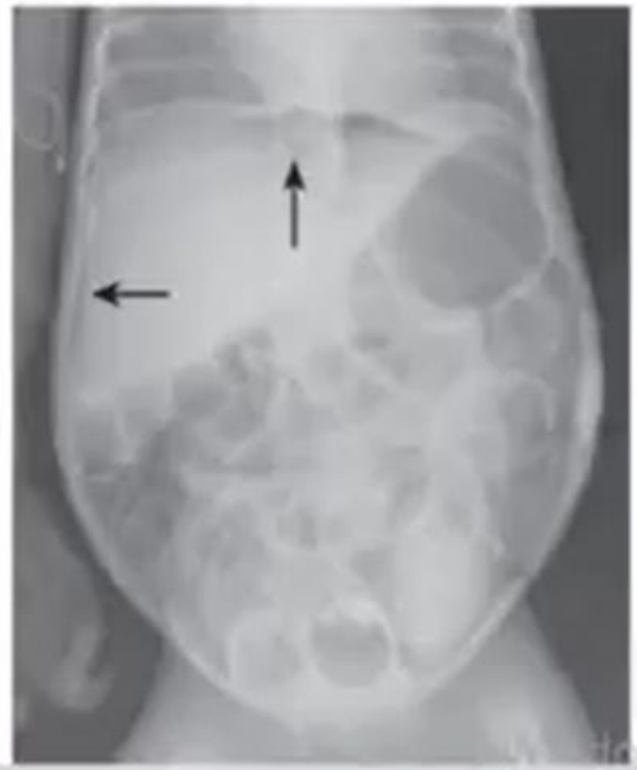
Table 33.1 Modified Bell Classification for NEC

	Clinical Findings	Radiographic Findings	Gastrointestinal Findings
Stage I	Apnea, bradycardia, and temperature instability	Normal gas pattern or mild ileus	Mild abdominal distention, stool occult blood, gastric residuals
Stage IIA	Apnea, bradycardia, and temperature instability	Ileus with dilated bowel loops and focal pneumatosis	Moderate abdominal distention, hematochezia, absent bowel sounds
Stage IIB	Metabolic acidosis and thrombocytopenia	Widespread pneumatosis, portal venous gas, ascites	Abdominal tenderness and edema
Stage IIIA	Mixed acidosis, coagulopathy, hypotension, oliguria	Moderate to severely dilated bowel loops, ascites, no free air	Abdominal wall edema, erythema, and induration
Stage IIIB	Shock, worsening vital signs and laboratory values	Pneumoperitoneum	Bowel perforation

- Abd Xray :Pneumatosis intestinalis is the classic radiographic finding in NEC.
- US

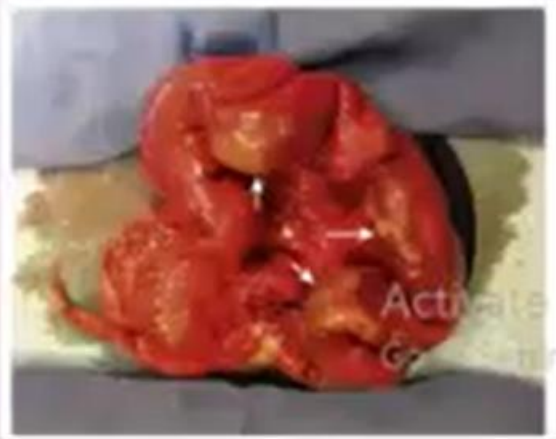






Management

- primary management is supportive (bowel rest, gastric decompression, IVF,IV antibiotic and parenteral nutrition, cardiopulmonary support if needed)
- Surgery is absolute indication in cases with pneumoperitoneum
- Surgical options : Laparotomy with resection /anastomosis laparotomy with resection/ stoma formation or peritoneal drainage (VLBW)

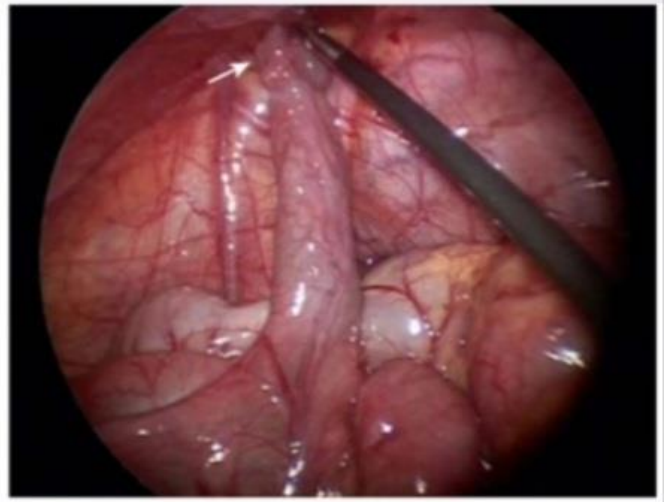


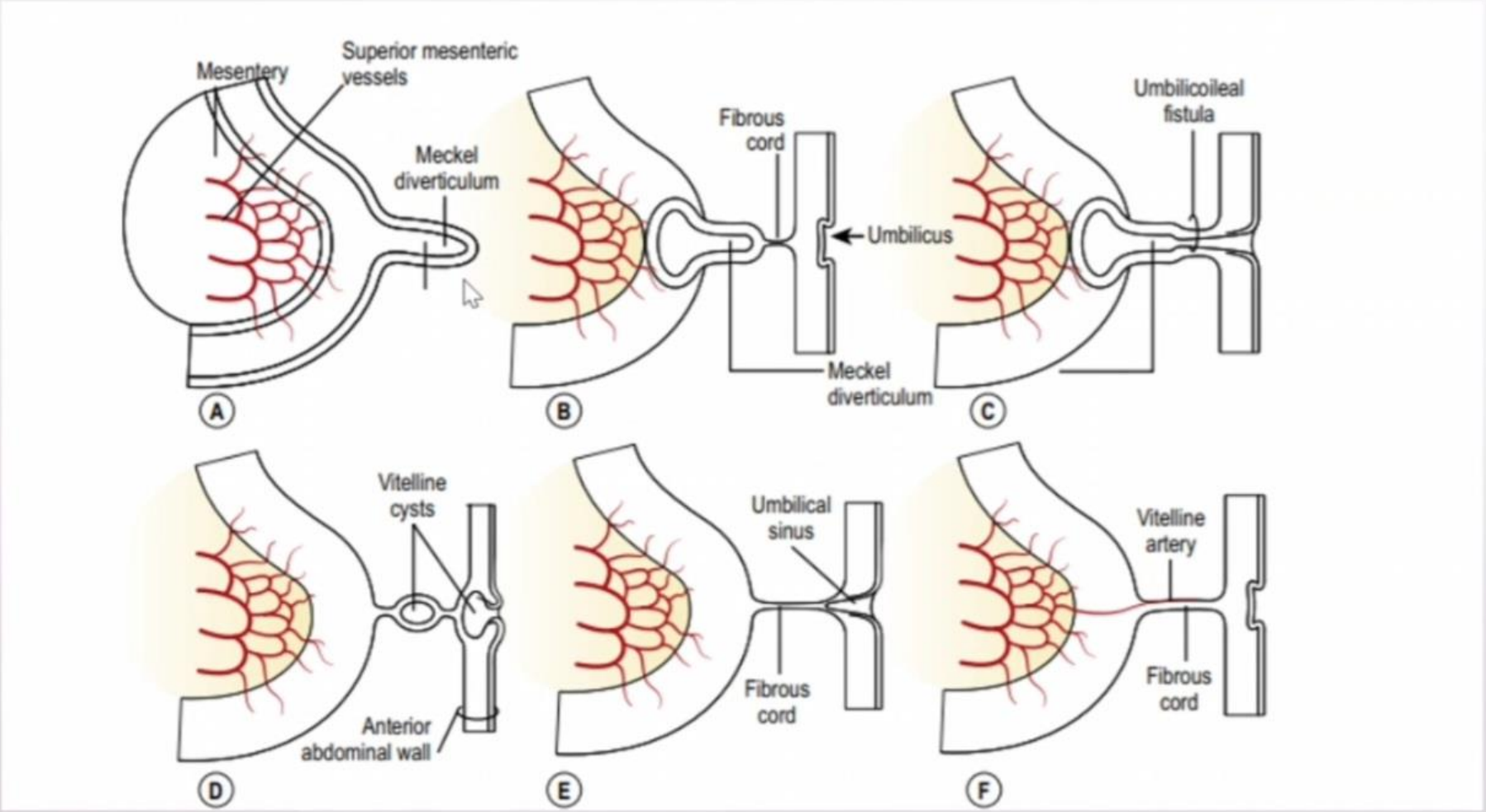
Outcome

- Recurrence 10 %
- Mortality ¬30%(inversely proportional to birth weight and gestational age) Medical NEC carries a mortality of 20%, surgical NEC mortality is 35- 50%)
- Intestinal failure :NEC is the leading cause of pediatric intestinal failure (IF) resulting in more than 1/3 of IF patients
- Stoma complications
- Intestinal stricture
- Neurodevelopmental delay :intellectual delays,moderate-to-severe developmental delay with speech and motor impairment

Meckel diverticulum

- True incidence of Meckel diverticulum is unknown because most patients are asymptomatic.
- Estimated at approximately 2%,
- ¬ 4% will become symptomatic
- M:F of 2:1
- Rule of 2s : occurs in 2% of the population
  - 2:1 male-to-female ratio
  - discovered by 2 years of age
  - located 2 feet (60 cm) from the ileocecal valve
  - commonly 2 cm in diameter and 2 inches (5 cm) long
  - contain two types of heterotopic mucosa (Gastric is the most common followed by pancreatic)

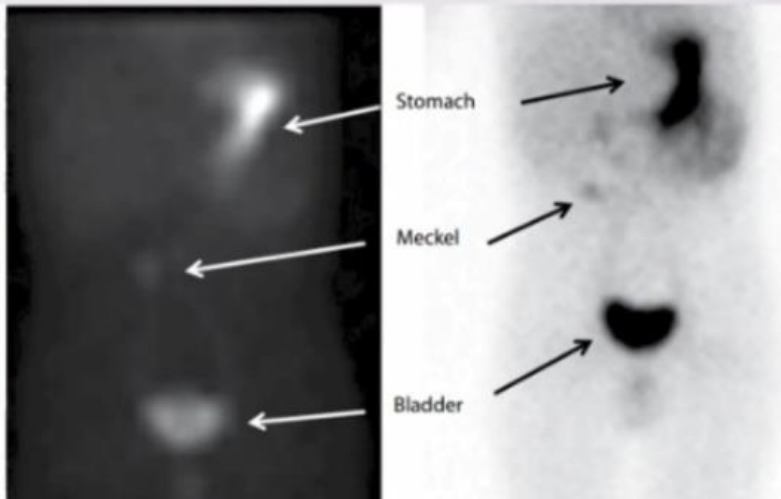


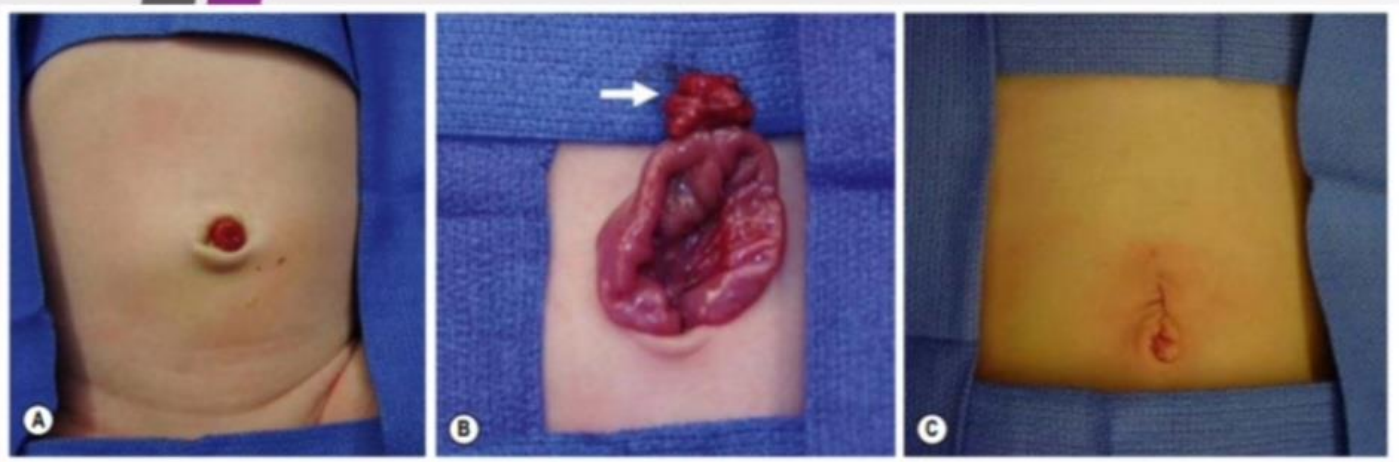

Mesentery
Superior mesenteric vessels
Meckel diverticulum
A
Fibrous cord
Umbilicus
Meckel diverticulum
B
Umbilicoileal fistula
C
Vitelline cysts
Anterior abdominal wall
D
Umbilical sinus
Fibrous cord
E
Vitelline artery
Fibrous cord
F

- The three most common presentations in children are **intestinal bleeding (30–56%), intestinal obstruction (14–42%), and diverticular inflammation (6–14)**
- Less common signs include a cystic abdominal mass and a newborn with an umbilical fistula resulting from a patent vitelline duct, In elderly, neoplasia can develop within the Meckel diverticulum. (Carcinoid is the most common tumor)

Diagnosis

- In patients presenting with obstruction or inflammation, the diagnosis of a Meckel diverticulum is not usually definitively determined preoperatively
- US and CT might be helpful
- In case of bleeding diverticulum , technetium-99m pertechnetate radionuclide study (Meckel scan) , false negative 25%

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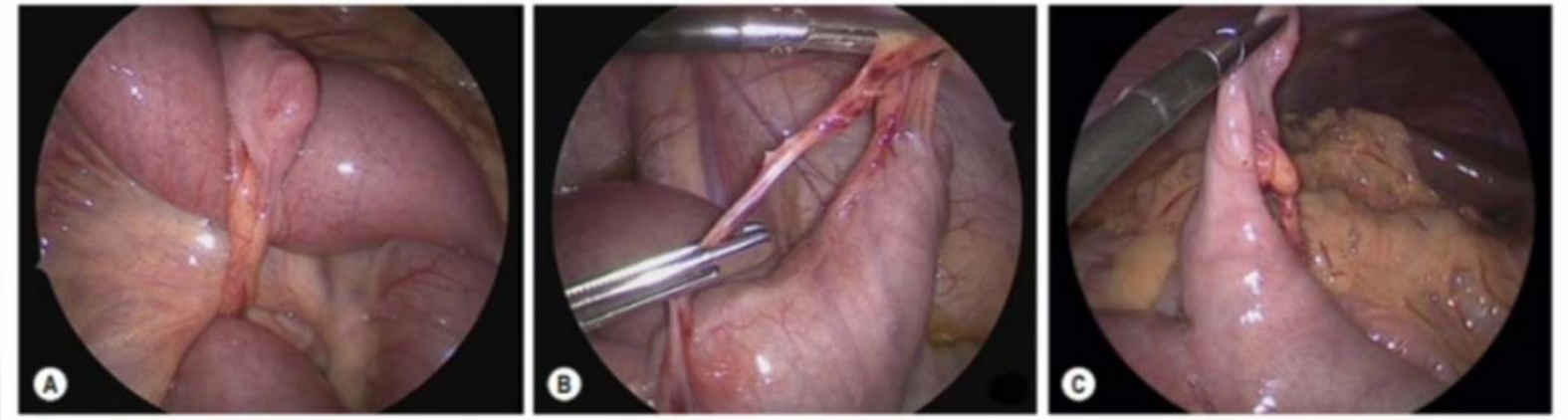
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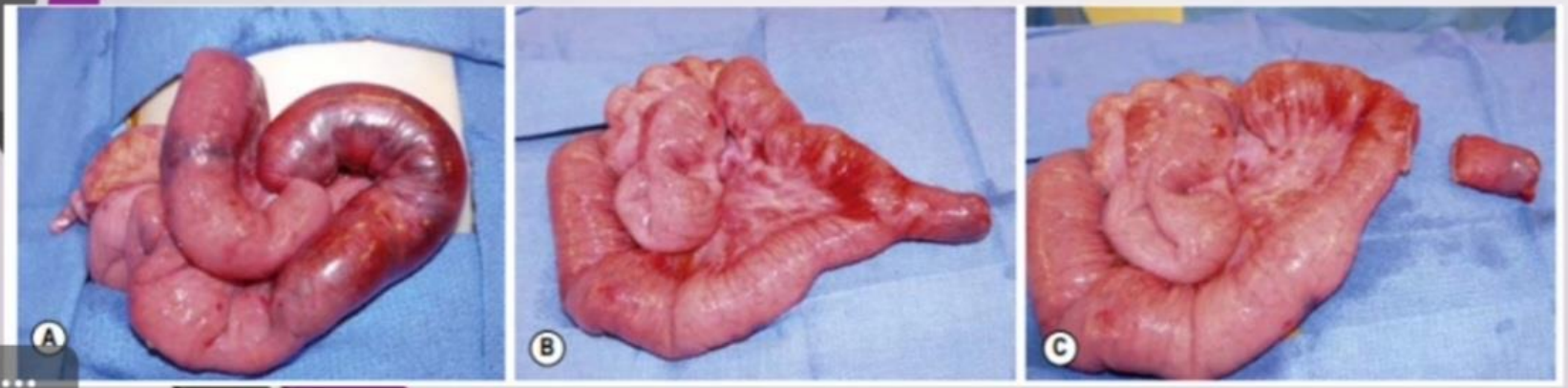
A
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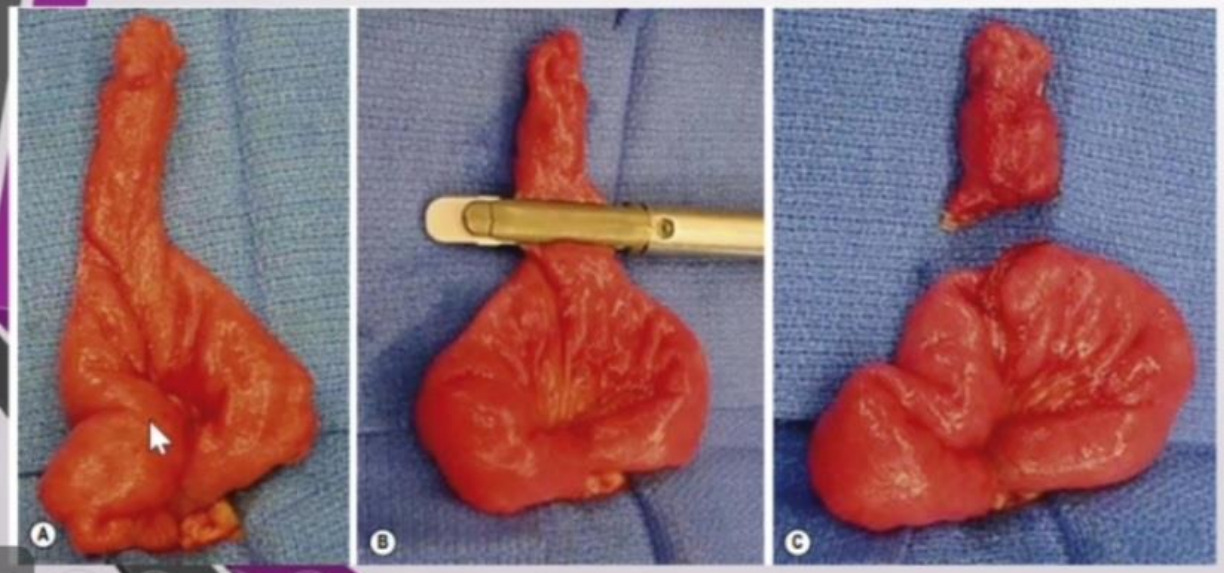
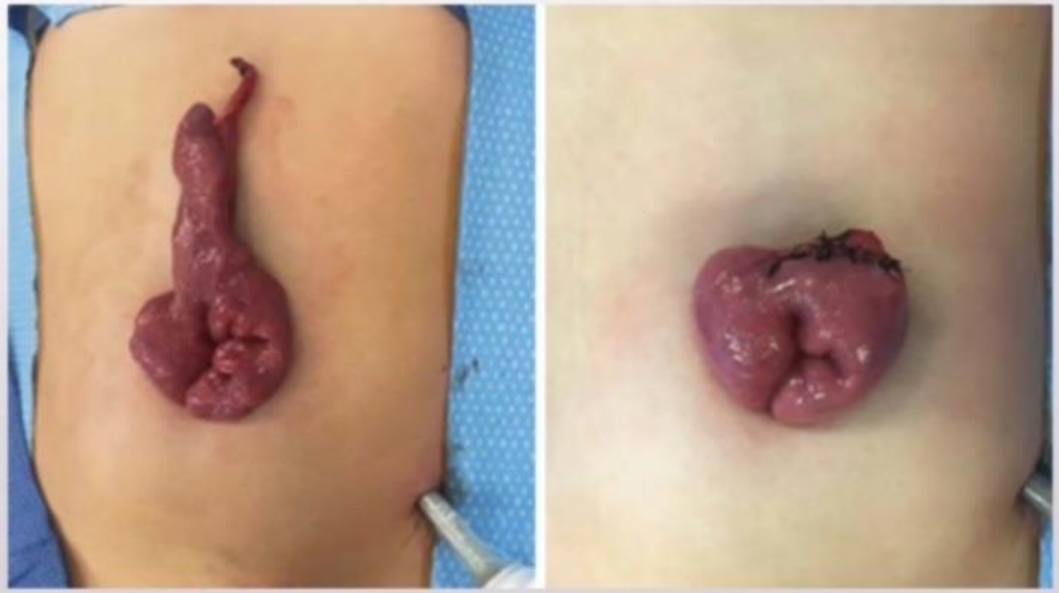
C

Management

- Stabilize the patient in case of bleeding
- Surgery : open or laparoscopic diverticulum resection or segmental bowel resection + anastomosis





Biliary atresia

- Biliary atresia (BA) is a relatively rare obstructive condition of the bile ducts causing neonatal jaundice
- It is a sclerosing cholangiopathy that represents the most common cause of end-stage liver disease and the most common indication for liver transplantation in children
- The incidence of BA varies around the world (Europe: 1 in 18,000 live births; France: 1 in 19,500 live births; UK and Ireland: 1 in 16,700 live births; Japan: 1 in 9640 live births
- The highest recorded incidence is in French Polynesia (1 in 3000live births).
- There is a slight female preponderance

- It is an isolated disease of term infants in 85% of cases. In the remainder of affected patients, it occurs as part of a syndrome, the most common of which is BASM (biliary atresia, splenic malformation (asplenia or polysplenia) and malrotation).

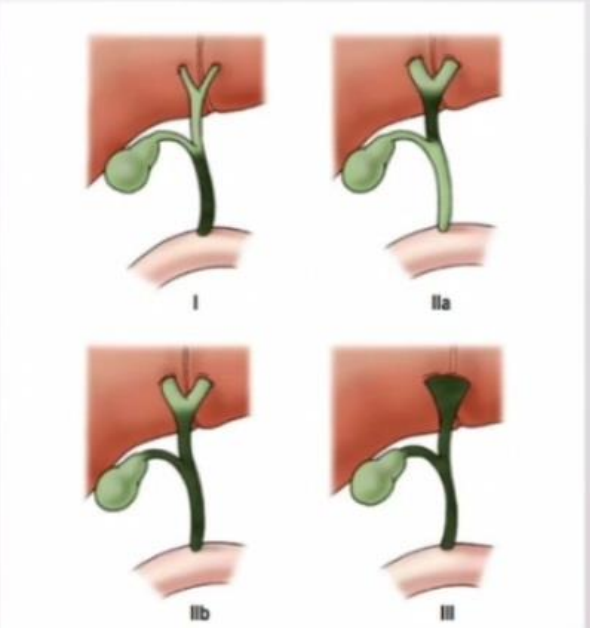
- The etiology is multifactorial (intrauterine or perinatal viral infection, immunologically mediated inflammation and other autoimmune/ genetic factors, exposure to toxins, abnormal ductal plate remodeling, a vascular or metabolic insult)

- BA is classified according to anatomic and cholangiographic findings.

Type I is atresia of the common bile duct

type IIa is atresia of the common hepatic duct, type IIb is atresia of the common bile duct and the common hepatic duct

Type III is atresia of all extrahepatic bile ducts up to the porta hepatis



Presentation

- Signs suggestive of BA are jaundice, pale stools, and hepatomegaly.
- Anemia, malnutrition, and growth retardation ensue because of malabsorption of nutrients and fat-soluble vitamins.

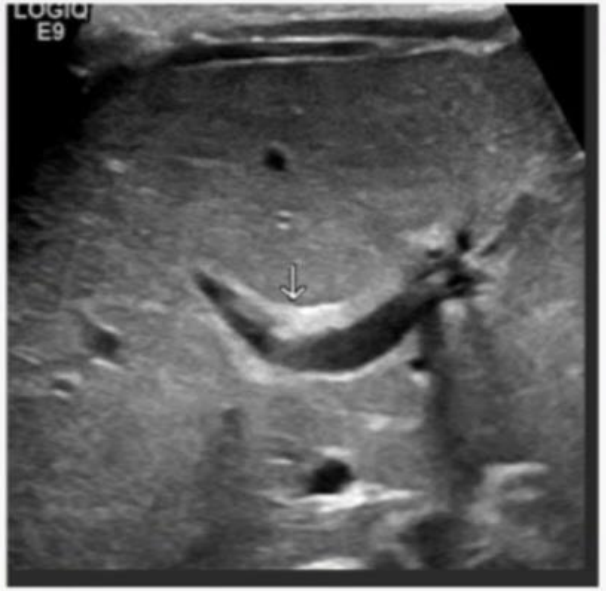
Box 43.1 Diagnosing Biliary Atresia

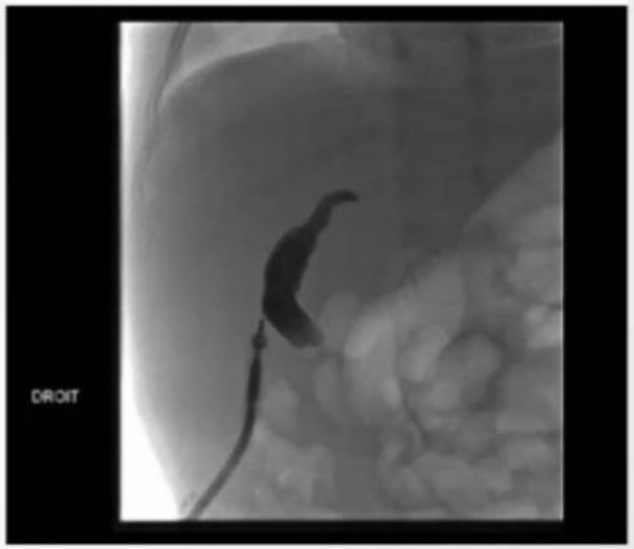
Routine Assessments

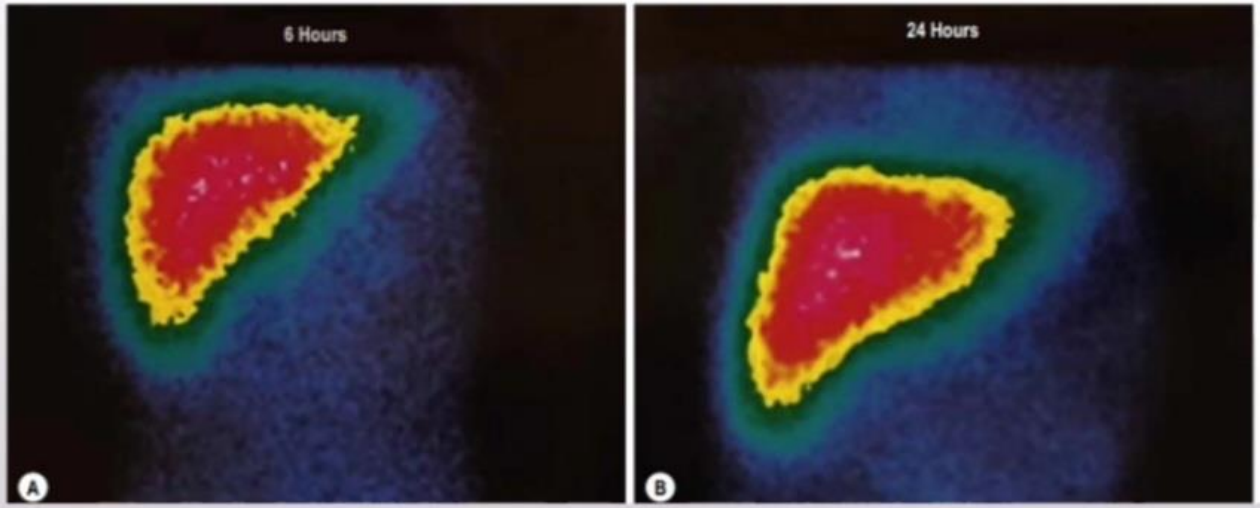
Stool color
Consistency of the liver on palpation
Conventional liver function tests plus γ-glutamyl transpeptidase
Coagulation (prothrombin time, activated partial thromboplastin time)
Ultrasonography
Hepatobiliary scintigraphy

Specific Investigations

Histobiochemical
Hepatitis A, B, C serology
TORCH titers
α1-Antitrypsin
Serum lipoprotein-X
Serum bile acids
Confirmation of extrahepatic bile duct patency
Duodenal fluid aspiration
Endoscopic retrograde cholangiopancreatography (ERCP)
Near-infrared reflectance spectroscopy
Needle biopsy
Direct observation (open or laparoscopic)
Surgical cholangiography

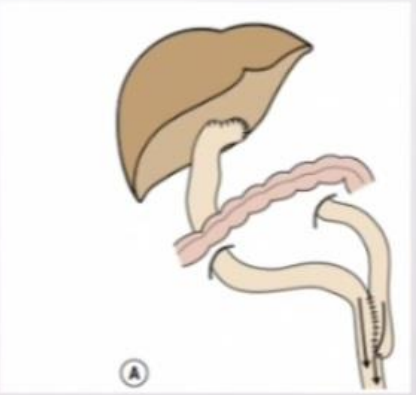






Surgery

- ROUX-EN-Y LIMB AND ENTEROTOMY FOR PORTOENTEROSTOMY (kassi procedure)



- Liver tx
- The indications for liver transplantation following portoenterostomy are: (1) lack of bile drainage; (2) signs of developmental retardation or its sequelae; and (3) presence of socially unacceptable complications/side effects.

outcome

- Classically, the major determinants of satisfactory outcome after portoenterostomy are
  (1) **age at** initial operation
  (2) successful achievement of postoperative bile flow
  (3) presence of microscopic ductal structures at the porta hepatis
  (4) the extent of liver parenchymal disease at the time of diagnosis
  (5) technical factors involving the portoenterostomy anastomosis
  (6) CMV status , syndromic or isolated

- Following a successful Kasai operation, pigmented stool is usually seen within 2–3 weeks

- Such success is typically seen in 2/3 of patients, but is maintained into adulthood in only 1/2 of the patients with initial jaundice clearance.
- liver transplantation will be required in 2/3 of patients at some point in their life.

Post op complications

- Cholngitis
- Fat , protein , and mineral malabsrbtion
- Failure to thrive
- Portal hypertension
- HEPATOPULMONARY SYNDROME AND PORTOPULMONARY HYPERTENSION
- INTRAHEPATIC BILE LAKE CYSTS
- HEPATIC MALIGNANCY