PEDS ABDOMEN

General Surgery Seminar

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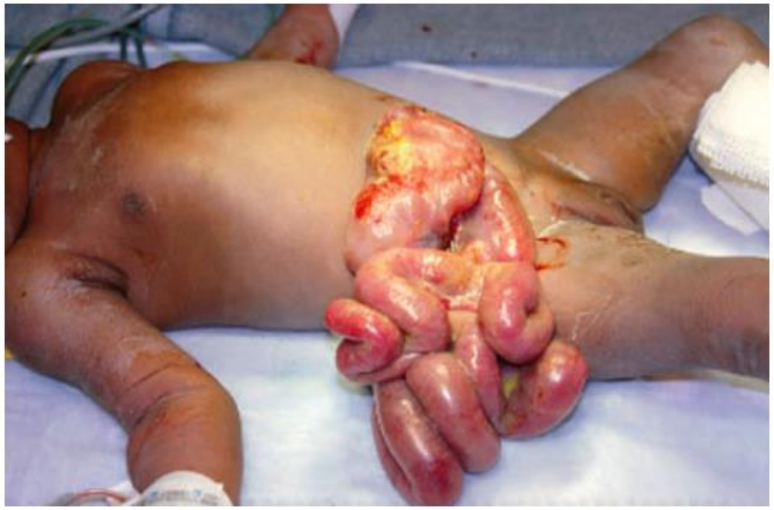
Case 1

A 2800g female is born via SVD at 38 weeks gestation. Vitals: T 37, HR 158, BP 80/50, RR 40. She has an abdominal wall defect. The bowel is thickened, edematous, and friable.



Case 1

What kind of defect is this?
Gastroschisis

What are the associated anomalies?
None.
10% of babies have bowel abnormalities such as atresia and stenosis. These are related to the bowel trauma and position in utero.

Case 1

What are the initial steps in managing this patient after delivery?

ABCs

Insert NG/OG

Cover bowels with saline soaked gauze and plastic

Administer IV fluids, Abx

Maintain normothermia

Case 1

What are the surgical management options?

1.Primary closure in OR. *Risk of abdominal compartment syndrome.*

2.Umbilical cord flap. *Temporary coverage.*

3.Placement of silastic silo. *This will prevent heat/water losses and keep bowel in sterile environment. Silo tightened over time to slowly reduce bowel.*

Case 1

When would you proceed directly to the OR?

- *Closing gastroschisis: defect closes around viscera causing ischemia and infarction*

Case 1

You have repaired the defect primarily and the baby was started on TPN. 3 weeks later, the NG is still draining 60 cc bile per day. The baby has not passed meconium. What do you do next?

A. Observation without intervention
B. Upper GI series
C. Contrast enema
D. Gastric emptying study
E. Initiate oral feeds

Case 1

You have repaired the defect primarily and the baby was started on TPN. 3 weeks later, the NG is still draining 60 cc bile per day. The baby has not passed meconium. What do you do next?

A. Observation without investigation

A period of ileus is expected and does not justify investigation. Mean time from operation to initiation of oral feeds in gastroschisis is 3-6 weeks.

Case 1

What is the most likely long-term complication of gastroschisis?

Short gut syndrome

Adhesive small bowel obstruction

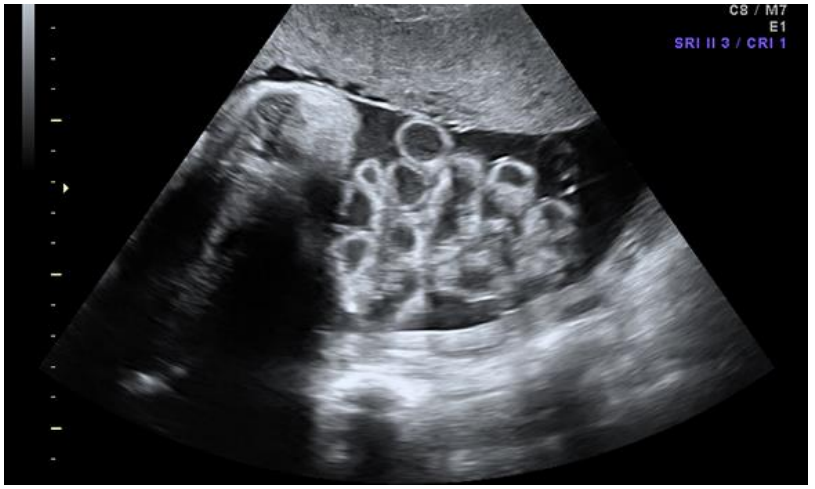
Gastroschisis

- Definition: full thickness, paraumbilical abdominal wall defect (usually < 4 cm) associated with evisceration of bowel
- No peritoneal sac covering bowel; bowel in direct contact with amniotic fluid which leads to serositis
- Liver is rarely involved

Gastroschisis

- Incidence: 1 in 3,000 – 8,000 live births
- Etiology: occlusion of right omphalomesenteric artery during embryogenesis results in disruption of umbilical ring and bowel herniation
- Possible factors: premature infants, low birth weight, young mothers (< 20 years old), ASA/ibuprofen/vasoconstrictive agents during 1st trimester, EtOH, smoking, recreational drugs

Gastroschisis



Diagnosis:

-Prenatal Ultrasound:

- < 4 cm paraumbilical abdominal wall defect
- Usually to the right of the midline
- Umbilical cord insertion site is normal

Gastroschisis

Management: Delivery Room

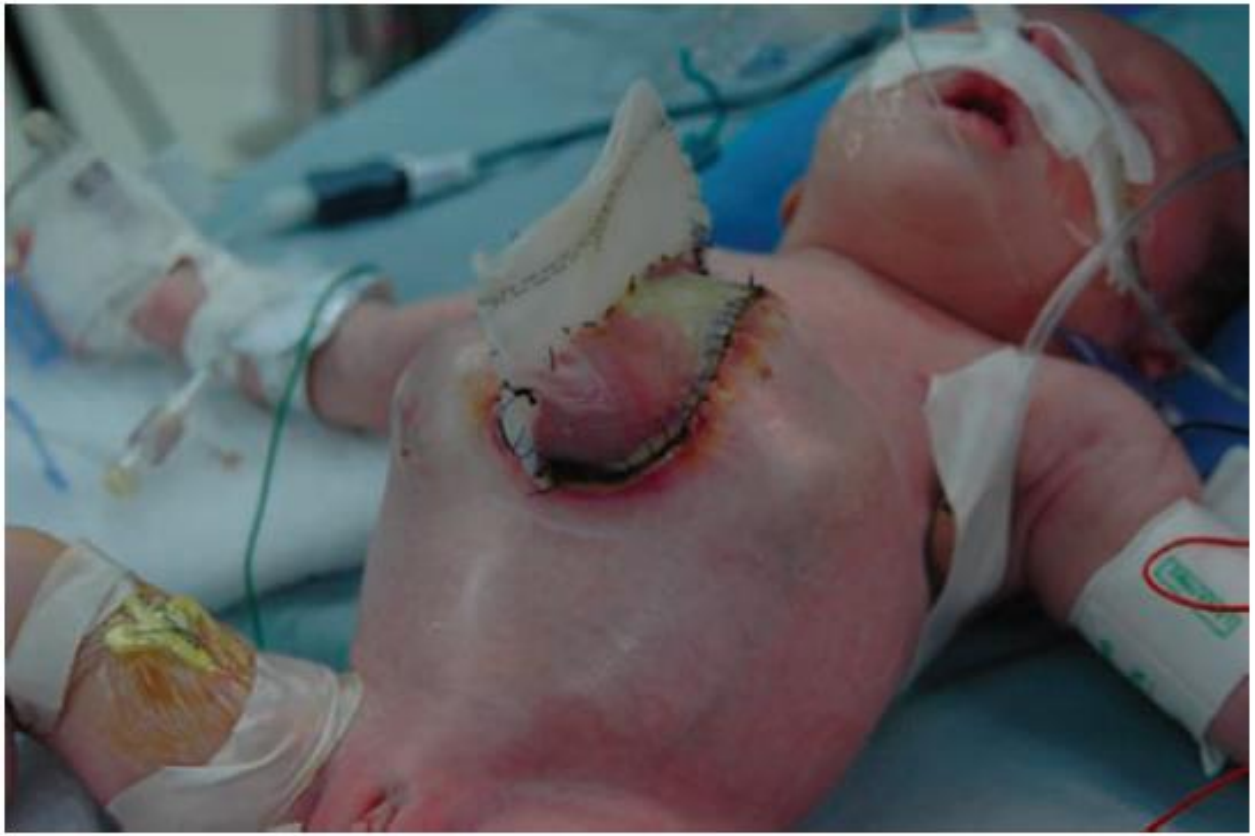
- Wrap bowel in sterile saline dressings covered with plastic wrap
- Insert OG to decompress stomach
- IV insertion – IVF and broad spectrum Abx
- Keep neonate in thermoneutral environment

Gastroschisis

Management: Operating Room
Primary Closure (70% success rate)
-Decompress bowel
-Extend defect by 1-2 cm
-Manually stretch abdominal wall
-Reduce bowel

Temporary closure
-Umbilical cord flap
Staged Closure
-Silastic silo placed at bedside to slowly reduce bowel

Gastroschisis



Gastroschisis

If associated with atresia:
1.Primary repair at time of abdominal wall closure –
OR–
2.Intestinal diversion (ostomy or long intestinal tube) followed by delayed repair

Gastroschisis

Prognosis:
-Survival rate >90%
-Favorable prognosis because typically not associated with other congenital anomalies
-Prolonged postoperative ileus
-25% complex cases associated with higher risk of in hospital mortality, short bowel syndrome, bowel obstruction, NEC, and TPN/tube feedings on discharge

Case 2

Male born at 34 wk 3 days gestation via C/S secondary to maternal and fetal distress. Baby was hypotonic. Apgars: 1 (1 min), 6 (5 min), 7 (10 min). Required PPV and was transferred to NICU. Consulted for abdominal wall defect.

Case 2

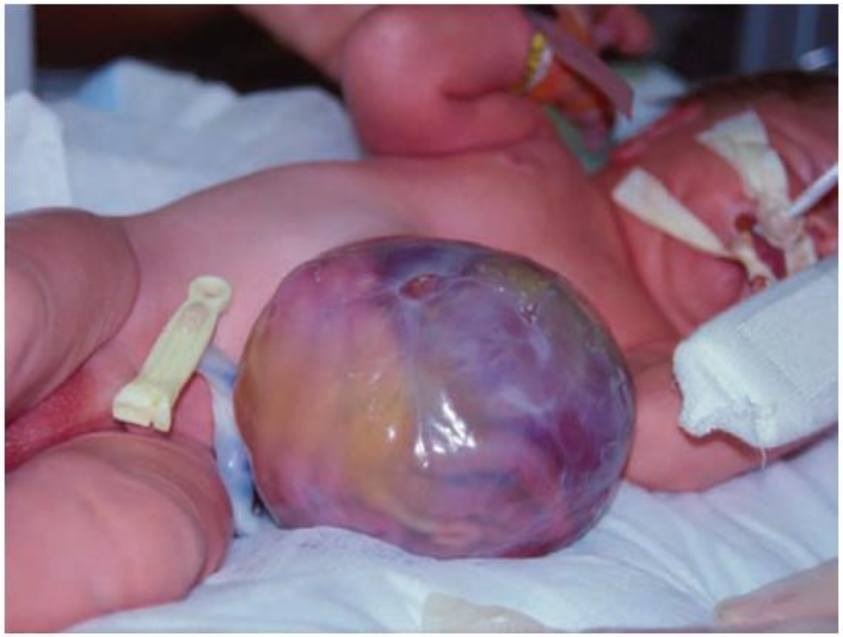
Prenatal hx:

- Born to 25 year old female
- Antenatal U/S showed a midline herniation with the sac containing bowel loops
- Fetal echo normal
- Spontaneous rupture of membranes at 28 wks – mother briefly admitted and treated with steroids and Abx

Case 2

Physical exam –
Large defect (~10cm) with intact amnion containing bowels and liver
What is the diagnosis?

Giant omphalocele



Case 2

Next steps:

-Stabilize (ABCs)

-Cover with gauze/plastic

-NG, IV Abx

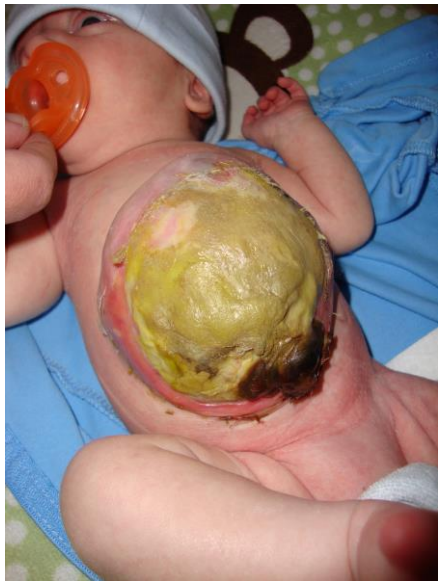
-Rule out associated congenital anomalies

- Echo, CXR, genetic testing

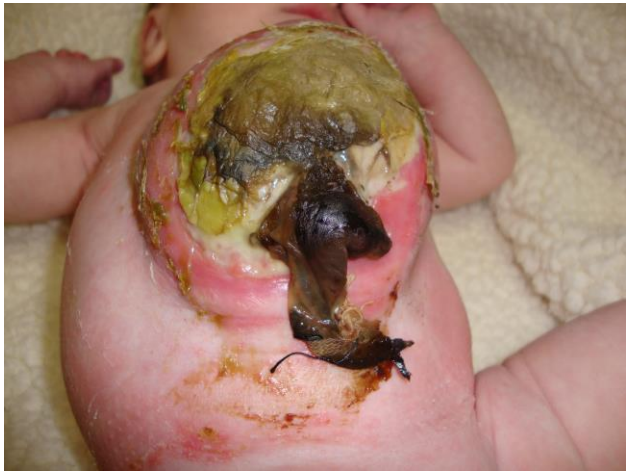
-Small omphalocele → repair

-Giant omphalocele → delayed repair

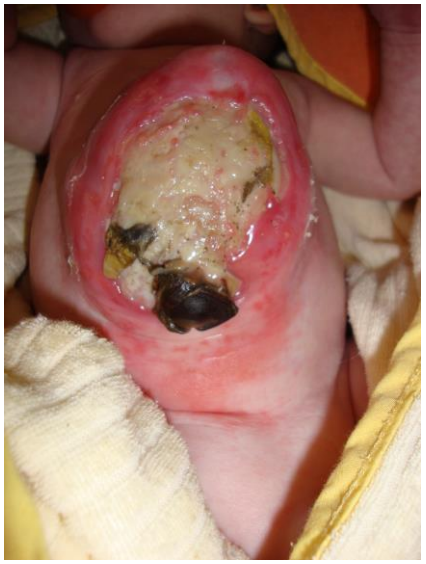
- Topical sclerosing agent



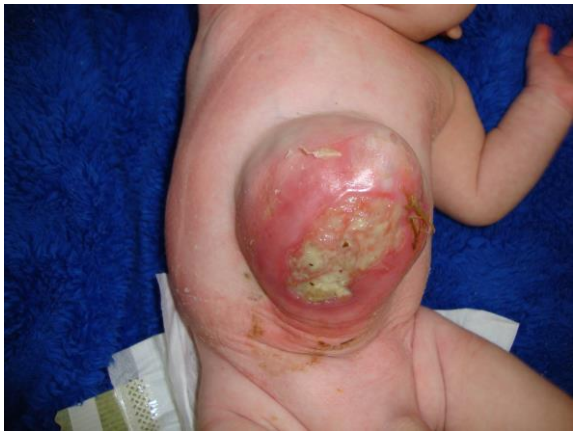
3 weeks



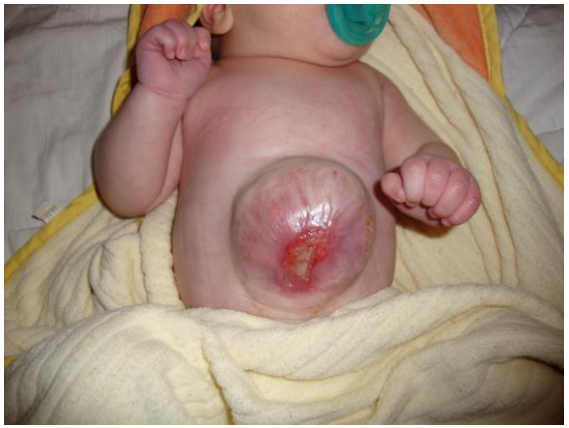
4 weeks



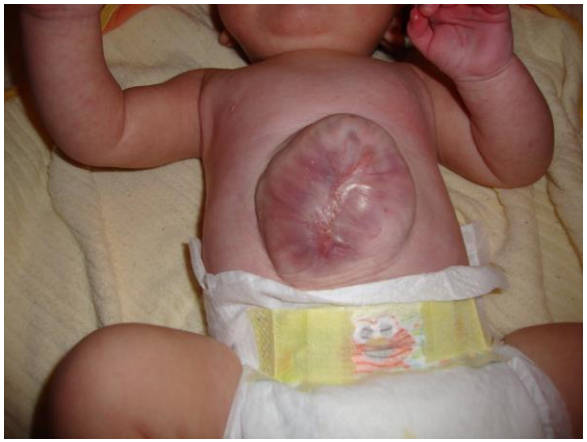
5 weeks



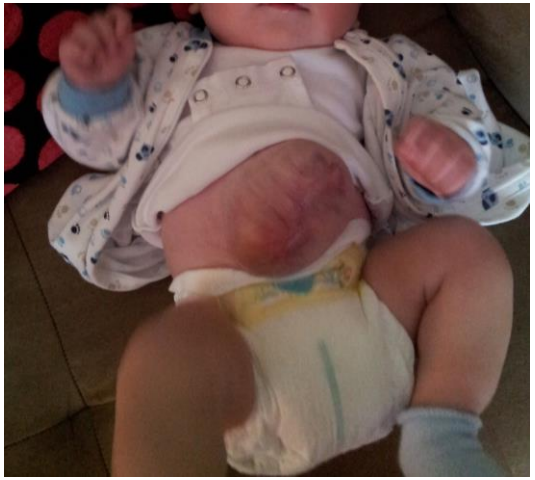
6 weeks



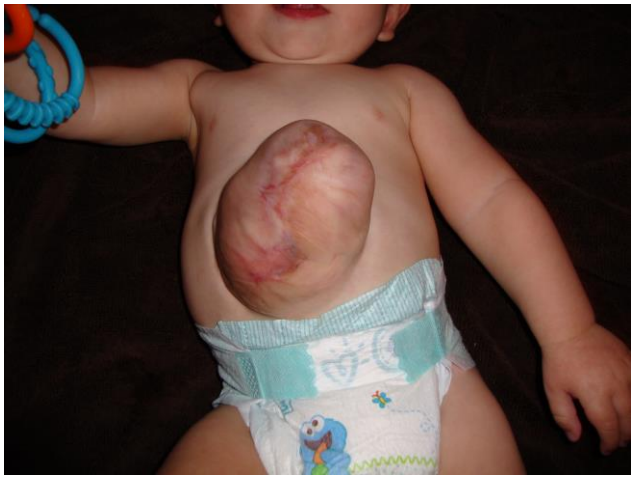
2 months



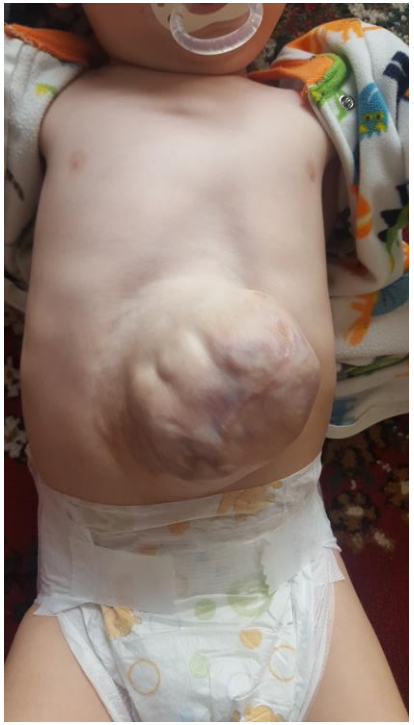
3 months



4 months



9 months



2.5 years

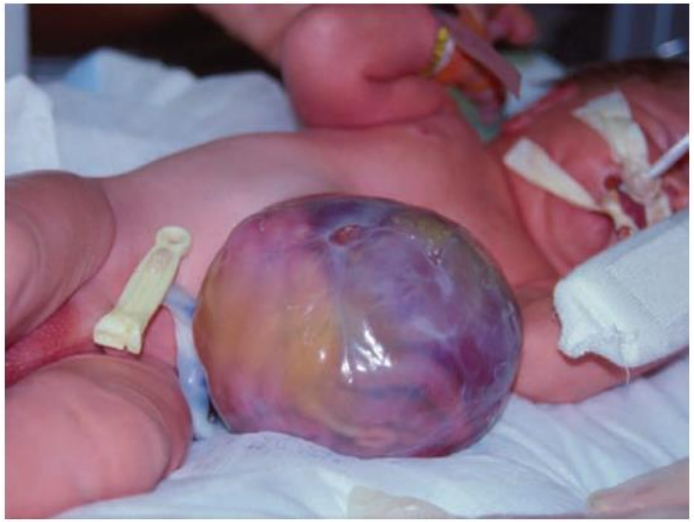
Case 2

Brought to the operating room at age 3 for laparotomy and primary repair of omphalocele.

Discharged POD 7

Omphalocele

- Definition: abdominal wall defect of varying size with herniated viscera contained in a sac
- Incidence: 1 in 6,000 – 10,000 live births



Omphalocele

Development of abdominal wall and GI tract depends on growth and fusion of cephalic, caudal, and lateral embryologic folds.

Omphalocele occurs when there is failure of migration and fusion of these folds.

Can be associated with other midline defects.
-Pentalogy of Cantrell

- Failure of cephalic fold: epigastric omphalocele, anterior diaphragmatic defect, sternal cleft, pericadial and cardiac defects

Omphalocele

Environmental and social factors play less of a role compared to gastroschisis

Karyotype abnormalities present in 30%
-Trisomy 13, 18, 21

50% associated with malformations
-Cardiac*, MSK, GI, GU

Beckwith-Wiedemann syndrome
-Omphalocele, macroglossia, hyperinsulinism

Omphalocele

Defect: small (2-5 cm) to large (>8 cm)

Liver containing or Non liver containing
“Giant omphalocele” – contains 75% of liver

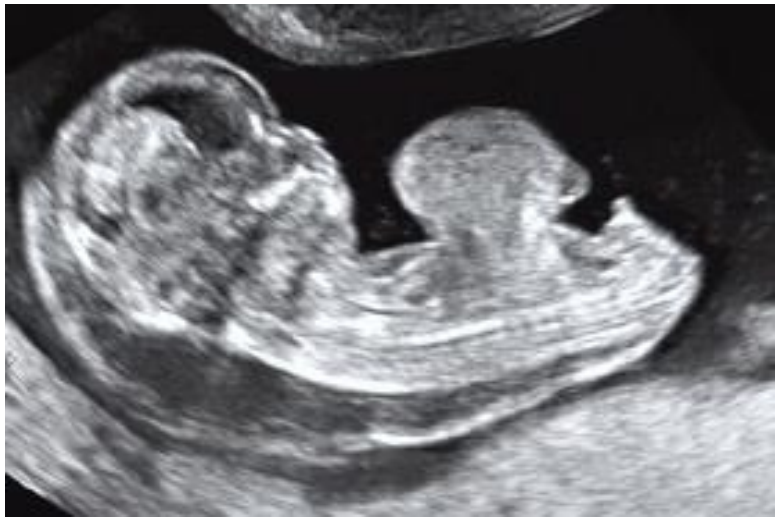
Sac: amniotic membrane, mesenchymal tissue (i.e. Wharton jelly), and peritoneum

Bowel: healthy (not exposed to amniotic fluid); malrotation is usually present

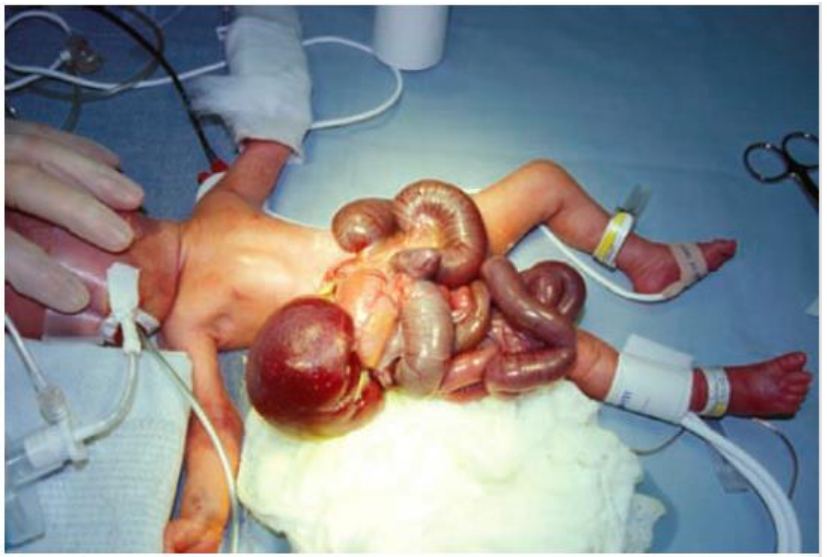
Omphalocele

Diagnosis:

Prenatal U/S
(2nd Trimester)



Physical Exam



Omphalocele

Management:

-C-section if giant omphalocele with herniation of liver

-External warmer/incubator

-NG/OG tube

-Cover with saline soaked gauze and plastic wrap

-IVF + IV Abx

-Pre-op work-up:

- CXR, echo, renal U/S, bloodwork

Omphalocele

Management: Surgical

-Reduce herniated viscera

-Primary fascial closure (60-70%)

-Resect sac

-May consider staged closure (silo)

-Limiting factor is intra-abdominal pressure

- Avoid abdominal compartment syndrome
- Intra-operative intragastric/intravesical pressure, end tidal CO2, CVP, regional oximetry

Omphalocele

Management: Giant Omphalocele (10%)
-Primary closure not possible due to poorly developed abdominal wall
-Promote epithelialization of sac with secondary closure of ventral hernia at later date

- Topical agents used include: mercurochrome, silver nitrate, silver sulfadiazine

Omphalocele

Post-op:

- Prolonged ileus is not common
- Increased frequency of gastroesophageal reflux
- Excellent prognosis if not associated with severe malformations
  - Increased mortality in infants with chromosomal syndromes and cardiac defects

Gastroesophageal reflux (GER)

Epidemiology:

Symptoms begin within 6 weeks of life

80% become symptom free by age 1

20% continue to be symptomatic

4% of these patients develop esophageal strictures

GER

Mechanism: inappropriate LES relaxation

Clinical manifestations:
-Regurgitation
-Coughing, wheezing, stridor secondary to aspiration
-Refusal to eat (secondary to esophagitis)
- May lead to failure to thrive and malnourishment

-Sandifer's syndrome
- Spastic posturing of head, neck, upper trunk associated with eating; disappears during sleep

GER

Diagnosis:

- Clinical
- Scintigraphy – technetium (gastric emptying)
- 24 hr pH monitoring
  - Indications: respiratory symptoms, intractable crying, reactive airways, recurrent pneumonia
- Endoscopy and biopsy
  - Indications: suspicion of esophagitis, dysphagia
- Manometry

GER

Treatment:
Positioning – prone with head elevated
Thicken foods
Medications

- Antacids
- H2 receptor antagonists: ranitidine
- Prokinetic agents: cisapride (increase LES, improve esophageal peristalsis and gastric emptying)
- Proton pump inhibitor: omeprazole

GER

Surgery:

- Intractable esophagitis or emesis that does not respond to PPIs
- Principles
  - Lengthening intra-abdominal esophagus
  - Accentuation of the angle of His
  - Increase in pressure barrier at the GE junction
  - Approximation of the crura

GER

- Nissen Fundoplication**
- Boix-Ochoa Technique
- Thal procedure
- Toupet procedure

Umbilical Hernia

Most common abdominal wall defect

Incidence:
10 x more common in African Americans
25-50% vs 5-10%

Increased risk in premature infants

Umbilical Hernia

Umbilical ring closes by contracture after cord ligated and umbilical vessels thrombose

Failure of recti to approximate and failure of round ligament to attach to both superior and inferior margins of the umbilical ring predispose fetus to developing umbilical hernia

Umbilical Hernia

Diagnosis:

- Physical exam – usually first noted after separation of umbilical cord remnant from umbilicus
- Usually asymptomatic and reducible

Umbilical Hernia

Management:
-Majority spontaneously close
-If hernia persists by age 4-5 then should be repaired
-Defect > 1.5-2 cm less likely to close and repair may be considered earlier (age 2-3)

-10% persist into adulthood
-Risk of incarceration or strangulation is rare but does increase in adults

Epigastric Hernia

Result from defects in linea alba

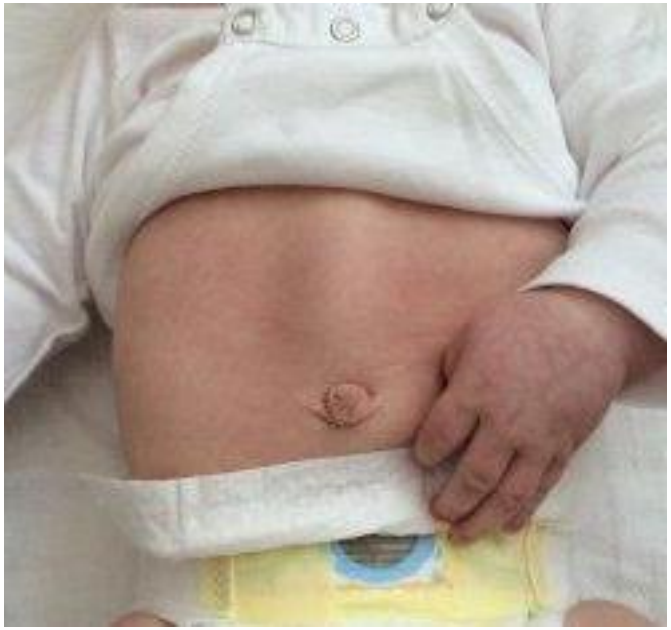
Incidence: 5%

Usually very small (0.5 – 1 cm palpable mass)

Do not resolve spontaneously – surgical repair required

Epigastric Hernia

Diastasis recti – rectus abdominis not fully developed



Case 3

13 day old boy presents with 1 day history of bilious emesis x 2 and BRBPR x 1

PMHx:
Born at 37 wks 3 days
Mother on methadone
NICU stay (no intubation) for a few days

Case 3

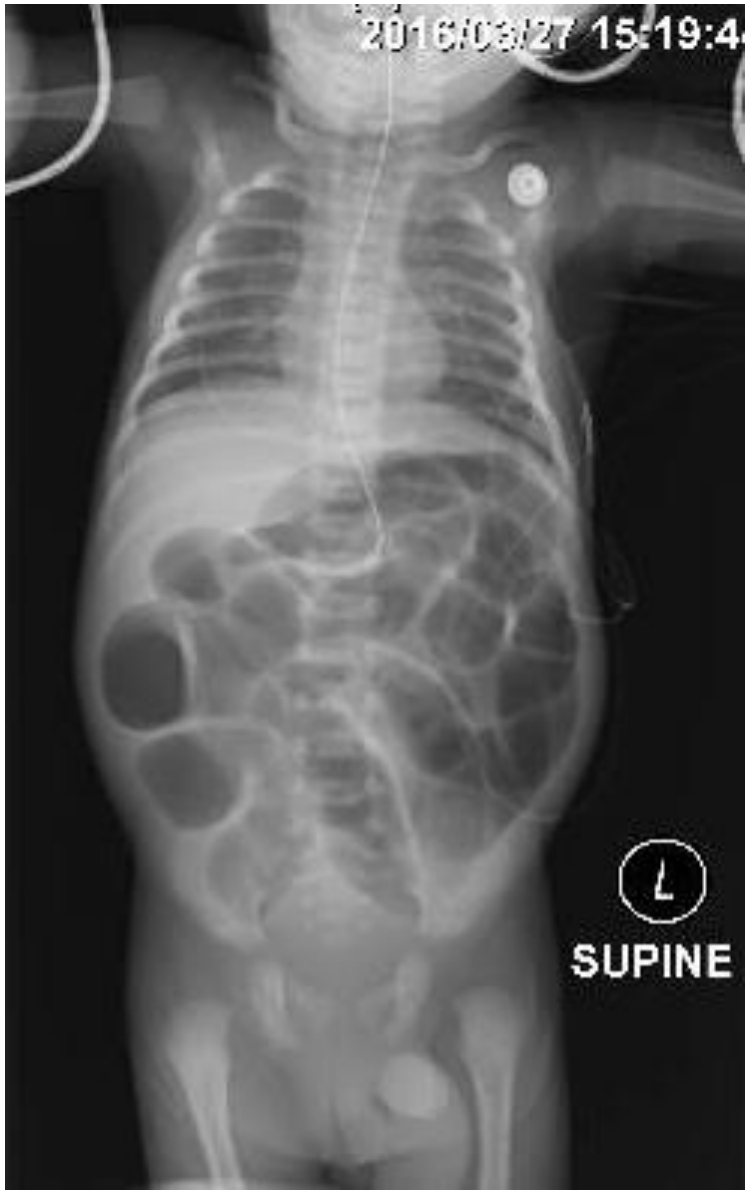
Differential diagnosis:

Next steps?

Case 3

O/E: T 37, HR 132, RR 54, BP 116/70
Abdomen distended, firm
Bulge in right groin
DRE - melena

Case 3



Case 3

pO2	L 69		86
pCO2	L 31		L 32
pH	L 7.32		L 7.33
HCO3	L 16		L 17
BE(B)	-8.4		-7.6
Sodium	H 149		141
Potassium	4.8		4.7
Chloride			
Glucose	6.2		9.6
Ionized Calcium	1.11		L 1.04
Lactate	H 3.3		1.2
Ionized Calcium pH7.4	L 1.07		L 1.01
Hematocrit	*65		*62
O2 Saturation (Calc)	*92.0		*96.0

3 hours later

Initial cap gas

Case 3

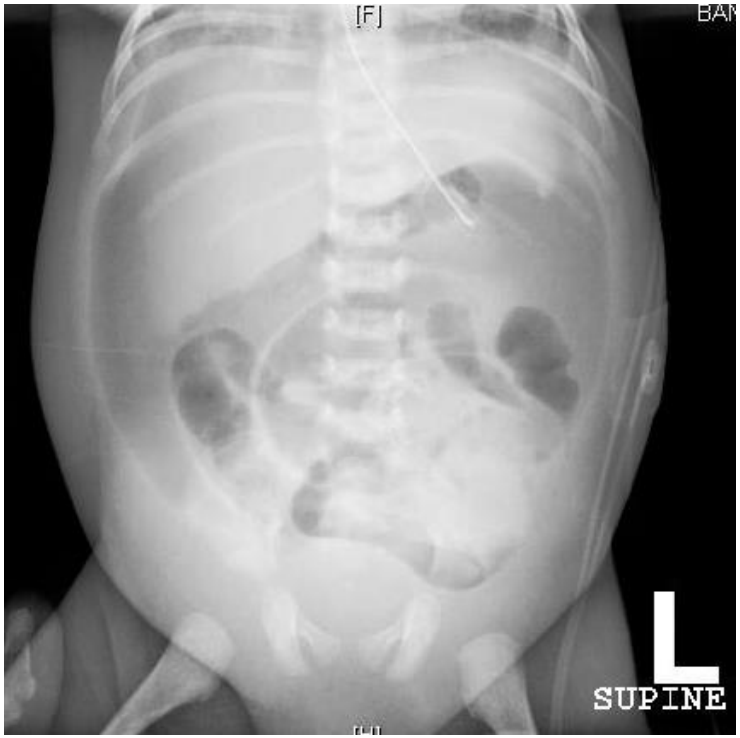
What is your management plan?
Resuscitate
NG tube
OR

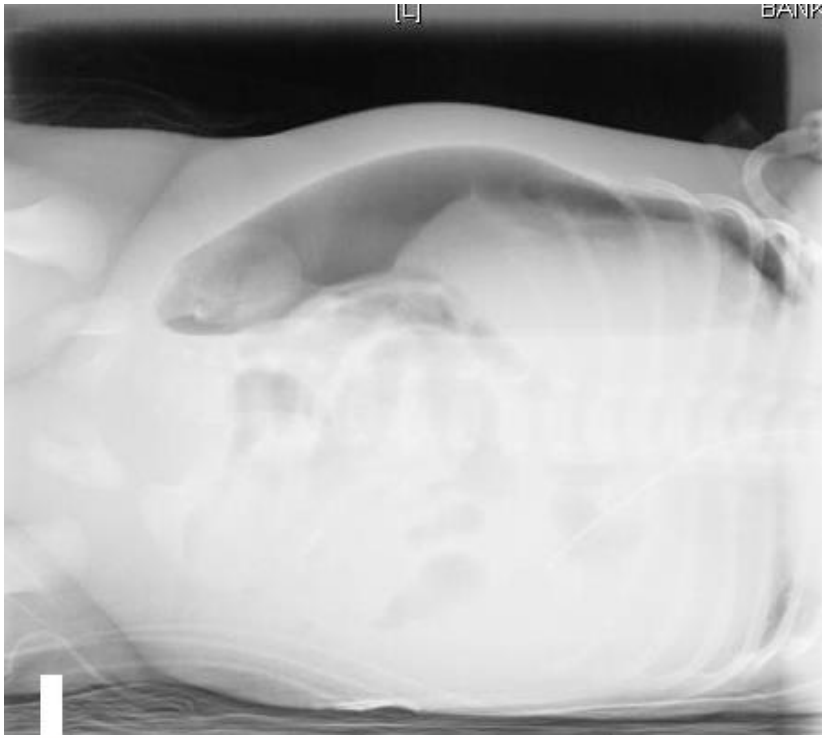
Case 3

- Laparotomy (omega incision)
- Dilated proximal small bowel with 5-8 cm incarcerated in right inguinal canal (10-15 cm from ileocecal valve) – reduced with mild traction, appeared ischemic
- Colon normal; no malrotation
- Right inguinal hernia repair (groin incision)
- Ischemic bowel resected, side to side stapled anastomosis

Case 3

On POD 8, you note that the baby's abdomen is distended and discolored.





Case 3

What next?
Exploratory laparotomy

Findings:
-3 tiny holes (1 mesenteric, 2 antimesenteric at staple line) closed primarily
-No intra abdominal contamination

-Next?
-Diverting loop ileostomy

Case 3

Patient was discharged 29 days after initial presentation (POD 21 from diverting ileostomy)

Admitted briefly for rehydration after increased ostomy outputs

Plan for closure of ileostomy in September

Inguinal Hernia

Most common elective pediatric general surgical procedure

Types:
-Congenital Indirect (99%)
-Direct (0.5%)
-Femoral (<0.5%)

Inguinal Hernia

Incidence: 1-3% of all children; 3-5% of premature infants

Increased incidence in those with connective tissue disorders (i.e. Ehlers-Danlos Syndrome or Marfan Syndrome)

8:1 male predominance

Right sided (56%) more common than left (27%)

Inguinal Hernia

Indirect Inguinal Hernia

-Abnormal patent continuation of peritoneum (processus vaginalis) through internal inguinal ring

-Hernia sac lateral to inferior epigastric vessels, anterior/medial to spermatic cord structures which are retroperitoneal

-Hernia sac descends along spermatic cord within cremasteric fascia

-Sac can reside within inguinal canal or descend through external inguinal ring into scrotum

Inguinal Hernia

Direct Inguinal Hernia

- Originates medial to deep inferior epigastric vessels
- External to cremasteric fascia
- Hernia sac protrudes directly through posterior wall of inguinal canal
- Can descend through external inguinal ring into scrotum

Inguinal Hernia

Signs and symptoms of incarcerated hernia:

- Non-reducible mass
- Inconsolable infant
- Feeding intolerance
- Pain
- Abdominal distention, vomiting, obstipation
- Edematous groin – reactive hydrocele

Inguinal Hernia

Operative Considerations

-Elective repair to prevent incarceration

- 70% of infants who require operative reduction of incarcerated inguinal hernia are <11 months old

-May attempt reduction of incarcerated hernia

- If remains unreduced for 1-2 hours then urgent operative reduction and repair

-Recurrence is low (0-1%)

Inguinal Hernia

Open
vs.
Laparoscopic

Journal of Pediatric Surgery (2006) 41, 1081–1084

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Journal of Pediatric Surgery

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Laparoscopic inguinal hernia repair—a prospective personal series of 542 children

Felix Schier*

Abstract

Purpose: This series prospectively evaluates a consecutive personal series of children undergoing laparoscopic hernia repair.

Methods: A total of 712 inguinal hernias were corrected laparoscopically in 542 children (396 boys and 146 girls, aged 4 days to 14 years, median 1.6 years). The internal inguinal ring was closed with a 4-0 nonabsorbable suture using 2-mm instruments. Patients were prospectively video-documented.

Results: There were no serious intraoperative complications. Operating time was comparable to open surgery. The contralateral inner ring was open on the left side in 16% of boys and 12% of girls, and on the right side in 18% of boys and 32% of girls. Direct hernias were found in 2.3%, femoral hernias in 1%, hernias en pantalon in 0.7%, and a combination of indirect and femoral hernia in 0.2%. Follow-up to date is 1-84 months (median 39 months). There were 4.1% hernia recurrences, 0.7% hydroceles and 0.2% testicular atrophies. Cosmesis is excellent.

Conclusions: Laparoscopic inguinal hernia repair can be a routine procedure with results comparable to those of open procedures. It is well suited for recurrences. The vas remains untouched. The visualization of structures is clear and leads to a defect-specific closure. The advantages of the laparoscopic approach include the following: its technical ease, it is an outpatient procedure, the cord structures remain untouched, the type of hernia is obvious, trocar placement is identical for any side or hernia type, clear visualization of the anatomy. Routine video documentation renders the diagnostic accuracy objective and absolute. Finally, recurrences are easier dealt with, be it from a previous open or from a laparoscopic approach. Although recurrences were slightly more frequent in the early stages, now they are closer to the rate with the open procedure.

Inguinal Hernia

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Incidence of metachronous contralateral inguinal hernias in children following unilateral repair – A meta-analysis of prospective studies☆

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Operative exploration of asymptomatic contralateral groin controversial
60-70% patent processus vaginalis at age < 2
However, risk of contralateral hernia following open unilateral repair is 6%

- Reserve for patients with associated disorders, high suspicion of bilateral clinical hernia, underlying risk to anesthesia, etc.

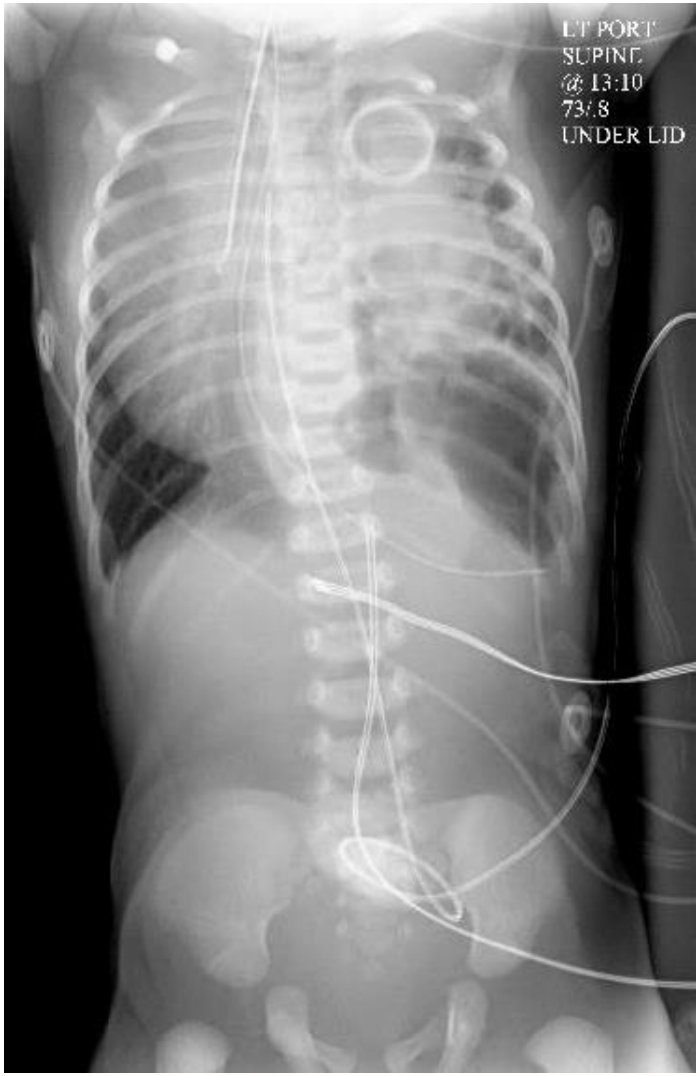
Case 4

Male born at 39 wk 6 days gestation via C/S due to failure to progress and fetal distress. The pregnancy was normal other than polyhydramnios on U/S.

The baby is blue, HR 53. Apgar at 1 minute is 2. The abdomen is scaphoid. Baby is resuscitated with bag mask ventilation and nasal intubation.

CXR shows the following:

Case 4



Case 4

What is the diagnosis?
Congenital diaphragmatic hernia

Case 4

Initial steps in management:
Intubation
NG or OG
IVF
NICU

Rule out associated anomalies
-Echo: most common cardiac association is left heart hypoplasia

Case 4

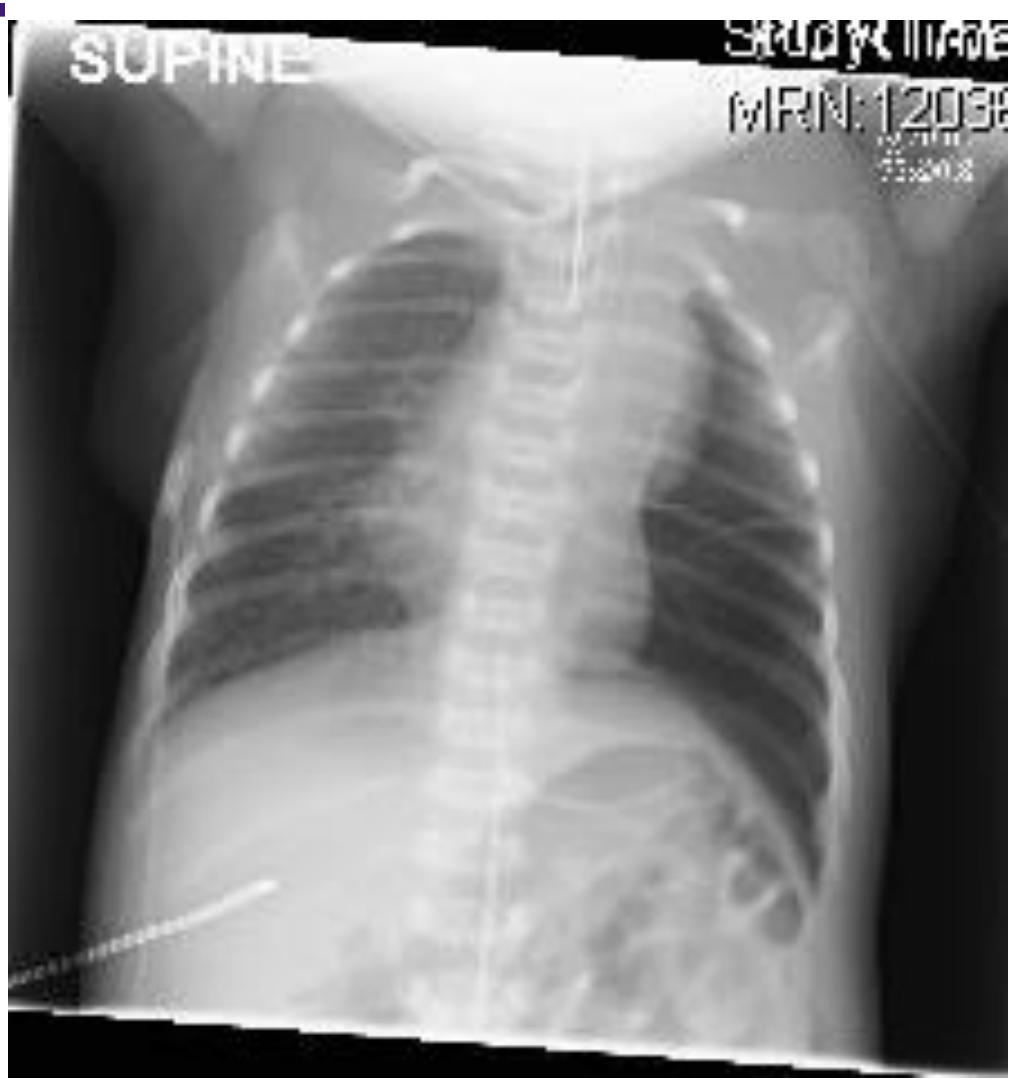
When would you proceed with surgical repair and what would you do in the operating room?

Case 4

At DOL 3 – laparotomy and repair of left CDH

1. Left subcostal incision
2. Small bowel, cecum, ascending, transverse, descending colon, left kidney and spleen in chest cavity; liver intra abdominal
3. Viscera reduced
4. Large defect spanning entire width of posterior diaphragm; diaphragm folded with adhesions
5. Posterior diaphragm released – primary repair
6. Viscera inspected – normal

Case 4



Congenital Diaphragmatic Hernia (CDH)

Definition: opening in diaphragm allowing viscera to herniate into chest

Incidence: 1 in 5000 births

Etiology: unknown

Left side predominance: 80% vs 20%

Location: posterolateral (90%) vs anteriomedial (10%)

CDH

Embryology:
-*Septum transversum* fuses dorsally with mesoderm of mediastinum
-*Pleuroperitoneal canals* connect pleural space and peritoneal cavity
-Closure of pleuroperitoneal canal → fetal diaphragm (8th wk gestation)
-Defective formation of pleuroperitoneal membrane or post-hepatic mesenchymal plate results in CDH

CDH

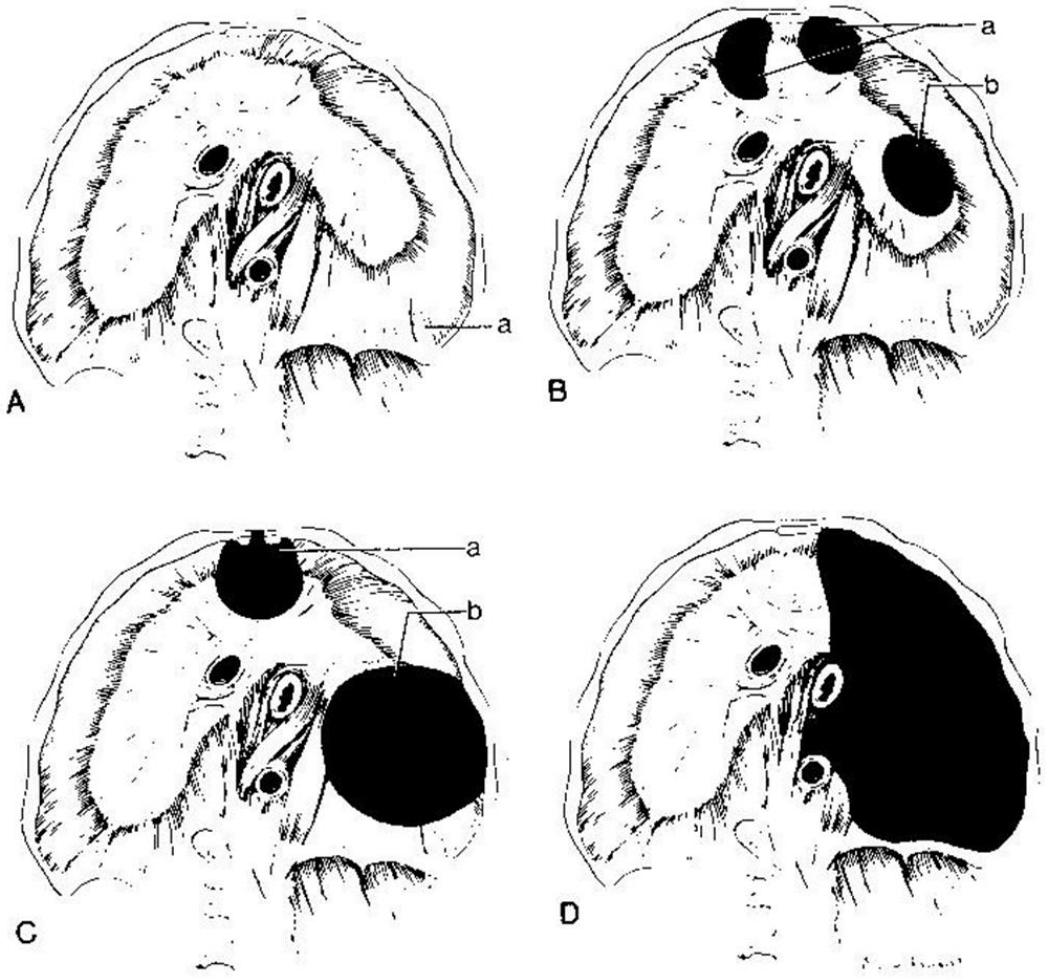


Figure 24–1. The inferior surface of the diaphragm and common locations of congenital diaphragmatic hernia. *A*, The normal diaphragm and the site (a) of the lumbosacral triangle, a potential area of weakness between diaphragmatic muscle fibers originating from the 12th rib and those originating from the lateral arcuate ligament. *B*, Retrosternal hernia (a) produced by failure of the sternal and costal contributions of the diaphragm to fuse at the site where the internal mammary artery traverses the diaphragm. Small posterolateral hernia (b) produced by failed closure of the pleuroperitoneal canal during embryologic development. *C*, Diaphragmatic defect and cleft sternum (a) associated with the pentalogy of Cantrell, which results from embryologic failure in the development of the septum transversum. Large posterolateral defect (b) with only a thin rim of posterior diaphragm. *D*, Agenesis of the left hemidiaphragm with absence of the left diaphragmatic crura.

CDH

Lung Development: 4 stages
1.Pseudoglandular (5-17 wks) – major bronchi, terminal bronchi formed
2.Canalicular (16-25 wks) - respiratory bronchioles, alveolar ducts, pulmonary vessels
3.Terminal sac (24 wks-birth)
4.Alveolar (late fetal life – childhood)

CDH

- Visceral herniation during pseudoglandular stage → Ipsilateral pulmonary hypoplasia
  - Decrease in pulmonary mass and weight
  - Reduction in bronchiole divisions
  - Reduction in number of alveoli and respiratory bronchioles
  - Hypolastic pulmonary vascular tree with abnormal muscularization of pulmonary arterioles
  - Surfactant deficiency

CDH

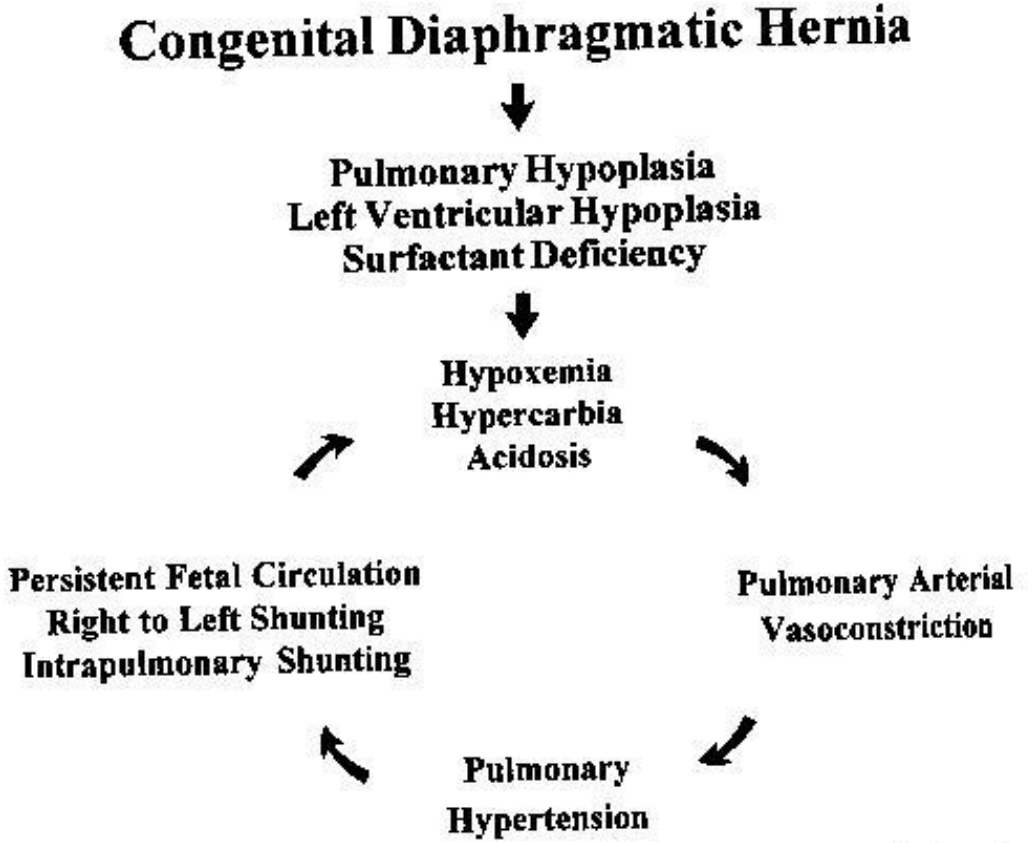


Figure 24–3. The pathophysiology of acute respiratory failure in neonates with congenital diaphragmatic hernia.

CDH

Associated anomalies (50%):

- Cardiac (60%): VSD, ASD, heart hypoplasia
- Neural tube defects
- Pulmonary sequestration
- Renal and genital anomalies
- Trisomies 13, 18, 21
- Midline defects – omphalocele, cleft palate, EA

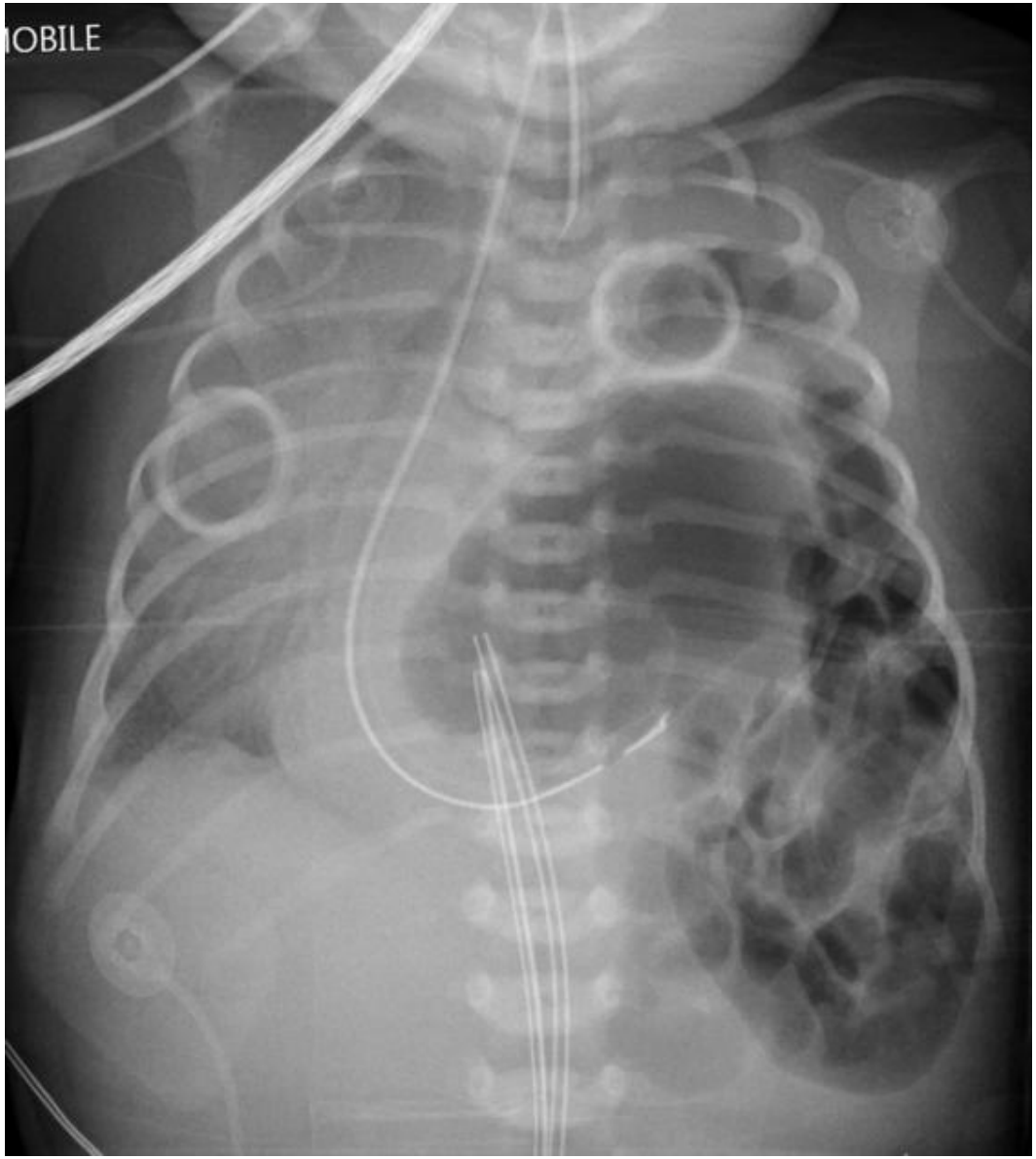
CDH

Diagnosis:
-Antenatal ultrasound
- Can be identified as early as 25 wk gestation
- Polyhydramnios (80%)
- Bowel/liver/spleen in chest

-On exam: respiratory distress, decreased air entry, bowel sounds in chest, scaphoid abdomen

CDH

Diagnosis: CXR



CDH

Management:

-ABCs
  - Endotracheal intubation, neuromuscular blockade, PPV
    - If conventional mechanical ventilation fails, consider: HFV, HFOV, nitric oxide administration, ECMO

-Insert NG

-Maintain normothermia

-Echo → rule out cardiac anomalies

-Repair when stable

CDH

Surgical considerations:
-Transabdominal approach preferred with subcostal incision
-Bowel, liver, spleen carefully reduced from thoracic cavity
-Excise hernia sac if present (20%)
-Close small defects primarily with 2-0 or 3-0 permanent sutures
-If large defect, use mesh
- Higher risk of recurrence

CDH

Post-Op Care:
-Chest tube to water seal to avoid overexpansion/pneumothorax of contralateral lung
-Continue mechanical ventilation

- Initial decrease in chest compliance
- Surgical stress can precipitate pulmonary vasoconstriction → pulmonary hypertension

CDH

Outcomes:

60-80% survival

- Highly variable
- Depends on associated anomalies
- Depends on severity of lung hypoplasia

CDH

Indications for fetal surgery:
-Severe pulmonary hypoplasia
-0% survival with optimal postnatal care

-Predictors of high mortality on U/S

- Early cardiac ventricular disproportion
- Reduced lung area to head circumference ratio
- Hepatic herniation into chest
- Polyhydramnios diagnosed before 25 weeks gestation
- Left ventricular hypoplasia

CDH

Fetal tracheal occlusion:

- Causes lung growth and gradual reduction of herniated viscera
- Mediated by mechanical and hormonal growth stimuli triggered by trapped fetal lung fluid (unable to exit into amniotic cavity)
- Goal is for baby to be born without lung hypoplasia or associated pulmonary hypertension
- Minimally invasive procedure; experimental

Case 5

7 week old girl born at term. Hospitalized a few days after birth due to hyperbilirubinemia. Her pediatrician is concerned as she still appears visibly jaundiced.

She is thriving, feeding well, and gaining weight appropriately. She passed meconium after delivery and now passes 2-3 yellow stools per day.

Case 5

O/E: Jaundiced. Alert, VSS. Abdomen slightly distended. Liver palpable 4 cm below costal margin.

Bloodwork is normal other than elevated total (250) and direct (15) bilirubin

Case 5

What is the differential diagnosis?
Congenital TORCH infections
Neonatal giant cell hepatitis
Alpha 1 antitrypsin deficiency
Alagille syndrome
Biliary atresia

Jaundice that persists longer than 4 weeks and jaundice due to conjugated hyperbilirubinemia is pathologic

TABLE 44-1. Causes of Jaundice and Cholestasis in Infancy

Disease	Age at Onset	Clinical Features	Diagnostic Test	Treatment
Unconjugated Hyperbilirubinemia				
Hemolytic disease	Birth–2 d	Severe jaundice, early	Positive Coombs's	Phototherapy exchange transfusion
Physiolgic jaundice	3–7 d	Increased with neonatal stress	Nonspecific	Phototherapy
Breast milk jaundice	1–8 wk	Benign	Nonspecific	D/C breast feeding ± phenobarbital
Congenital hemolytic disorders	1–8 wk	Progressive	Red cell fragility, specific tests for G6PD or pyruvate kinase levels	Variable, supportive
Metabolic	1–8 wk	Variable	Specific for disease (e.g., Crigler-Najjar, hypothyroidism)	Specific for disease
Conjugated Hyperbilirubinemia				
Inspissated bile syndrome	1st wk	Hemolytic disorders with exchange transfusion	US	Supportive, ± biliary tract irrigation
Bacterial infections	1st wk	Signs of sepsis	Blood culture + US	Supportive
Vascular cause	1st wk	Shock; congenital heart disease	US, echocardiogram	Supportive
Biliary atresia	After 1st wk	Well otherwise	US, liver biopsy, HIDA scan, open cholangiogram	Kasai portoenterostomy
Choledochal cyst	After 1st wk	May have sepsis, palpable mass	US, HIDA scan, open cholangiogram	Supportive/ reconstruction
Paucity of bile ducts	After 1st wk	Syndromatic form associated with Alagille's syndrome	US, liver biopsy, HIDA scan, open cholangiogram	Supportive/ ursodeoxycholic acid
Metabolic	After 1st wk	Varies with syndrome, galactosemia, α_1-antitrypsin deficiency, tyrosinemia, cystic fibrosis	Metabolic screening, HIDA scan, may require open cholangiogram	Specific to syndrome
Infection	After 1st wk	Generally ill	TORCH screen, HIDA scan, liver biopsy may require open cholangiogram	Specific to syndrome
Total parenteral nutrition	After 1st wk	Short gut syndrome, NEC	None specific, US, liver biopsy, HIDA scan	Enteral feeds
Idiopathic	After 1st wk	Systemically ill	Liver biopsy, US, HIDA scan	Supportive

D/C, discontinue; G6PD, glucose-6-phosphate dehydrogenase; HIDA, hepato-iminodiacetic acid; NEC, necrotizing enterocolitis; US, ultrasonography.

Case 5

Which investigations would you order?
Bloodwork
-Liver enzymes, bilirubin, serum alpha 1 antitrypsin, hepatitis work up
U/S
Nuclear scintigraphy (i.e. HIDA scan)***
-Uptake by liver without excretion for 48 hours is highly suspicious for biliary atresia

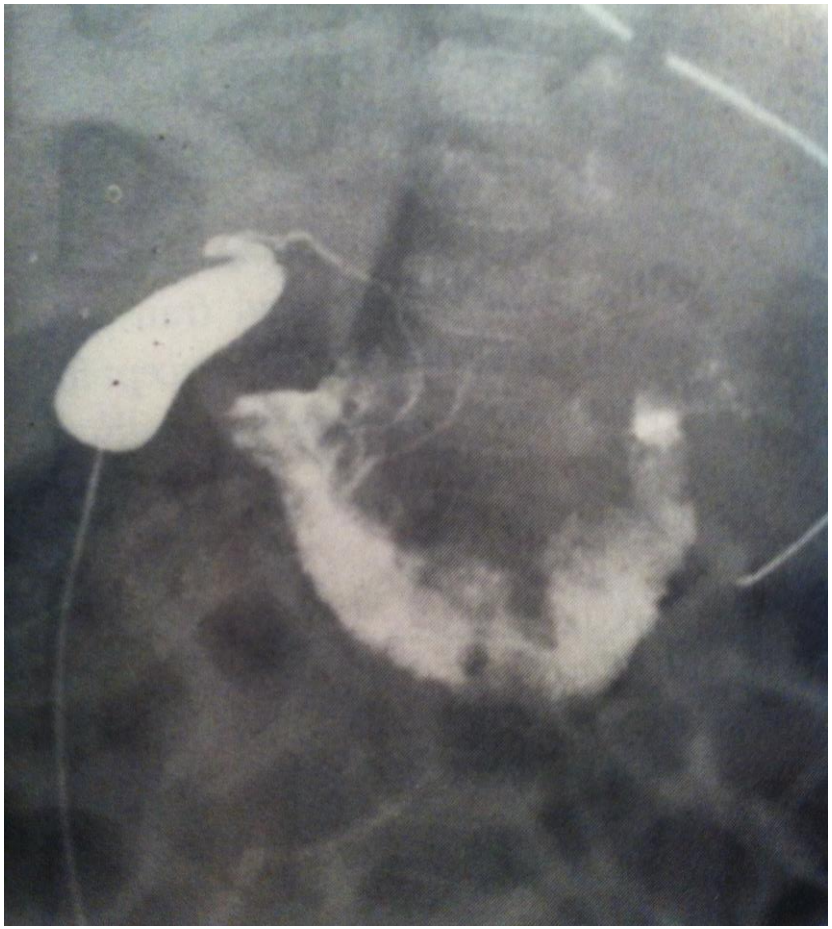
Case 5

Tests performed are inconclusive. Baby is now 8 weeks and jaundice continues. You decide to operate on the baby.

How will you proceed?

Case 5

Intraoperative cholangiogram:



Case 5

Next step?

Kasai portoenterostomy

- Excision of biliary tree
- Anastomosis of jejunum to portal plate

Liver biopsy – determine prognosis

Biliary Atresia

Definition: progressive obliteration of normally developed extrahepatic biliary tract

Biliary hypoplasia - diminutive but patent biliary tree secondary to neonatal hepatitis or alpha 1 antitrypsin deficiency

Incidence: 1 in 10, 000 births

Mechanism unknown

Most common cause of chronic cholestasis in pediatric population

Biliary Atresia

Subtypes:

A. Complete obliteration of biliary ducts

B. Obliteration of proximal ducts with patent distal ducts

C. Patent proximal ducts with distal fibrosis

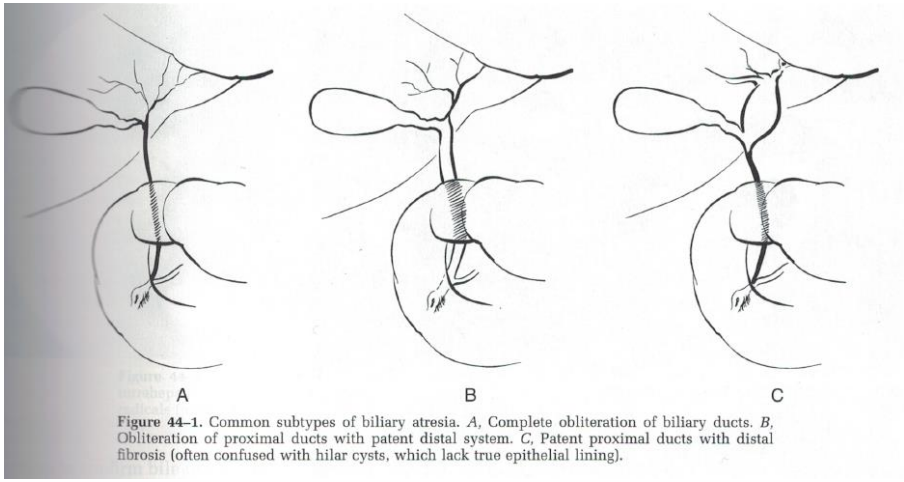


Figure 44–1. Common subtypes of biliary atresia. *A*, Complete obliteration of biliary ducts. *B*, Obliteration of proximal ducts with patent distal system. *C*, Patent proximal ducts with distal fibrosis (often confused with hilar cysts, which lack true epithelial lining).

Biliary Atresia

Presentation and Diagnosis:

- Progressive neonatal jaundice during first few weeks of life
- Dark urine
- Acholic stools
- Late findings: failure to thrive, feeding intolerance, portal hypertension, fat soluble vitamin deficiency

Biliary Atresia

Investigations:

-Conjugated hyperbilirubinemia

-Serum alpha 1 antitrypsin

-Radioisotope scanning (technetium 99m)

- Biliary atresia will have prompt uptake and no excretion into gut because of obliterated extrahepatic bile ducts

-Cholangiography

- Ruled out If bile ducts patent from liver to duodenum

-U/S

- Diminutive or absent gallbladder without associated intrahepatic duct dilatation

Biliary Atresia

Management:
Surgery
-Early biliary drainage within 2-4 months of age may be associated with reversal of liver injury and increase long term survival

Medical therapy – post-op management of chronic liver disease

Biliary Atresia

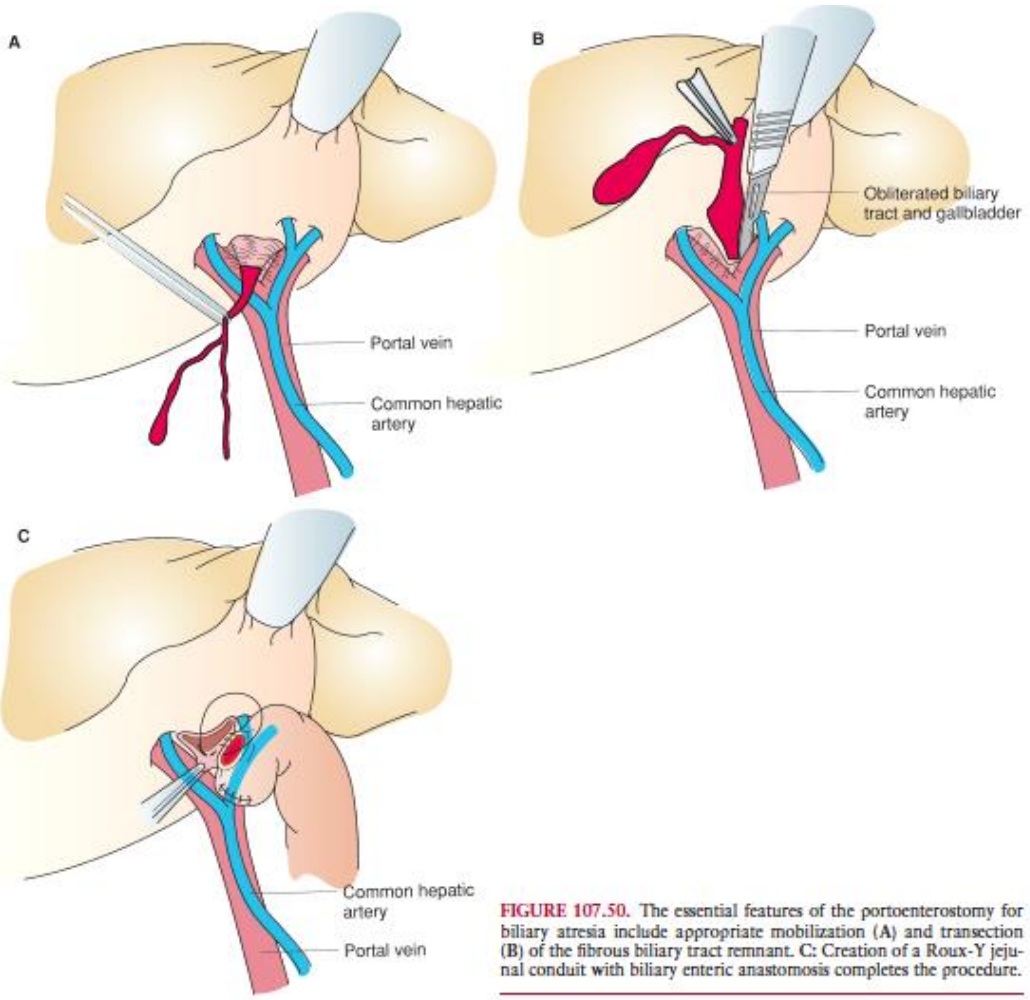


FIGURE 107.50. The essential features of the portoenterostomy for biliary atresia include appropriate mobilization (A) and transection (B) of the fibrous biliary tract remnant. C: Creation of a Roux-Y jejunal conduit with biliary enteric anastomosis completes the procedure.

Biliary Atresia

Portoenterostomy:

- Developed in Japan by Morio Kasai (1950s)
- Biliary drainage to arrest or reverse parenchymal liver injury
- Probability of bile flow depends on age at time of operation
  - 65-75% chance of bile flow if <2 months of age

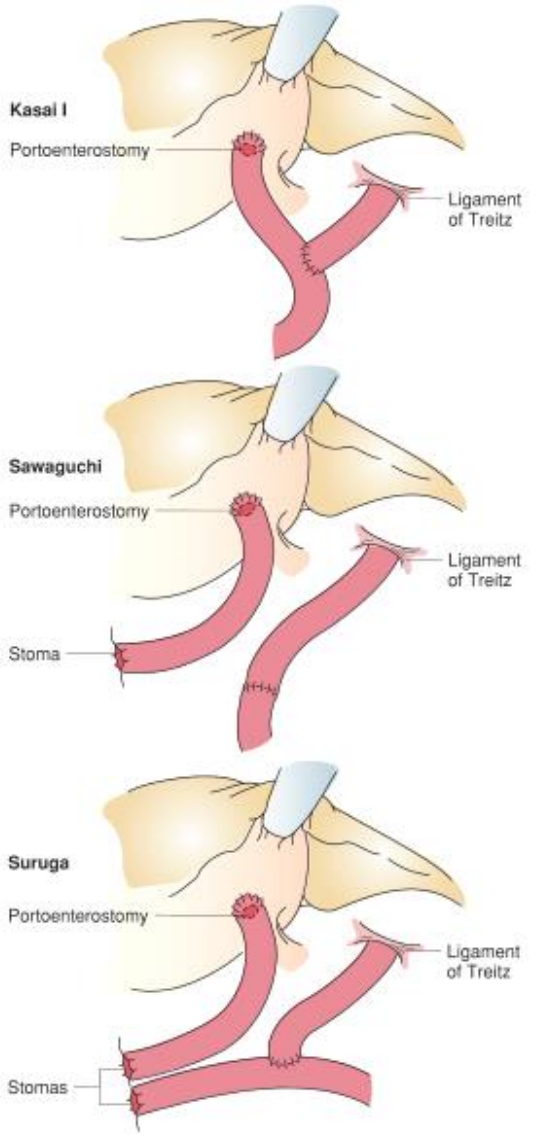


FIGURE 107.51. Several types of conduits have been used for biliary drainage following portoenterostomy. There are relatively few data to help in selecting from among them, and current trends emphasize simplicity. These are the three most commonly used conduits. The primary importance of this issue is that the reoperative surgeon must be familiar with the anatomic variations.

Biliary Atresia

Post-operative cholangitis (40-50%)
-Fever, leukocytosis, decreased bile flow
-Medical management: IVF, antibiotics

1/3 – do not require transplant
1/3 – liver failure after 5 years, requiring transplant
1/3 – liver failure post-op, requiring transplant

5 year survival rate ~50%

Biliary Atresia

Liver Transplantation:
Indications:

- Progressive hepatic failure despite portoenterostomy
- Growth retardation
- Complications of portal hypertension

5 year survival 75-95%

Consider effects of lifelong immunosuppression

- Risk of infection and treatment related malignancies

Case 6

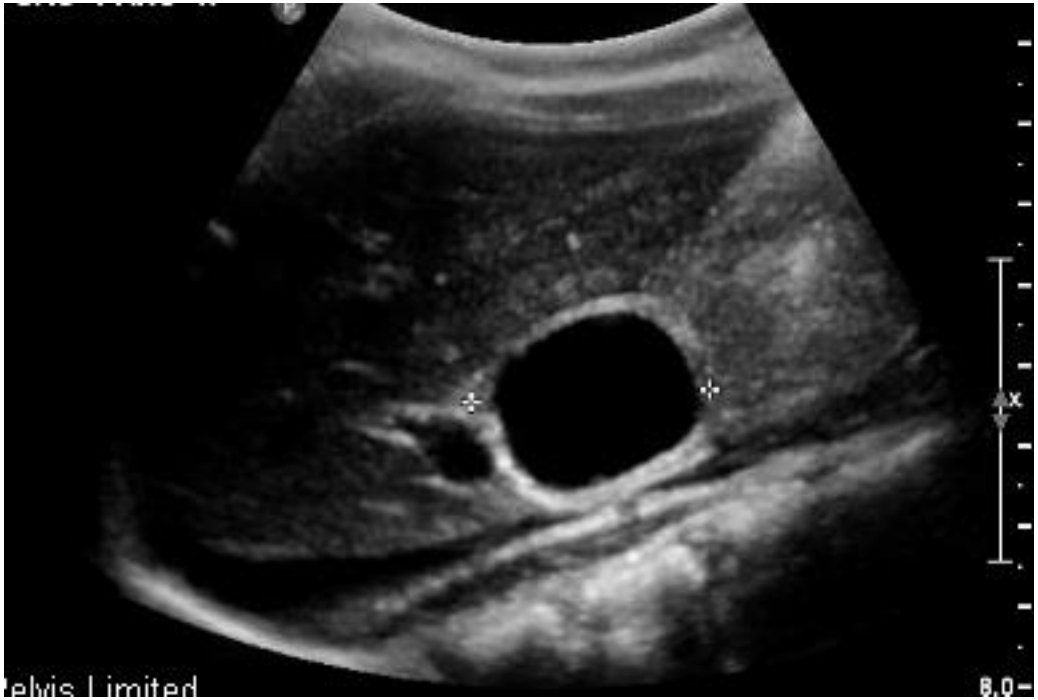
3 year old female referred to outpatient clinic for ”cystic lesion” on U/S of RUQ. History of abdominal pain since 3 months of age. Treated for constipation with daily PEG.

Case 6

Physical exam:
VSS
Abdomen non distended, soft, non tender
No palpable masses
No jaundice

Case 6

U/S:
4.1x 2.4 cm
cyst originating
from biliary tree



Involves extrahepatic ducts – tapers proximally and distally
Normal intrahepatic ducts

Bloodwork normal

Case 6

What is the diagnosis?

How would you manage this patient?

Case 6

Operation: Da Vinci assisted choledochal cyst resection, cholecystectomy, Roux en Y hepaticojejunostomy

Discharged POD 3

Choledochal Cysts

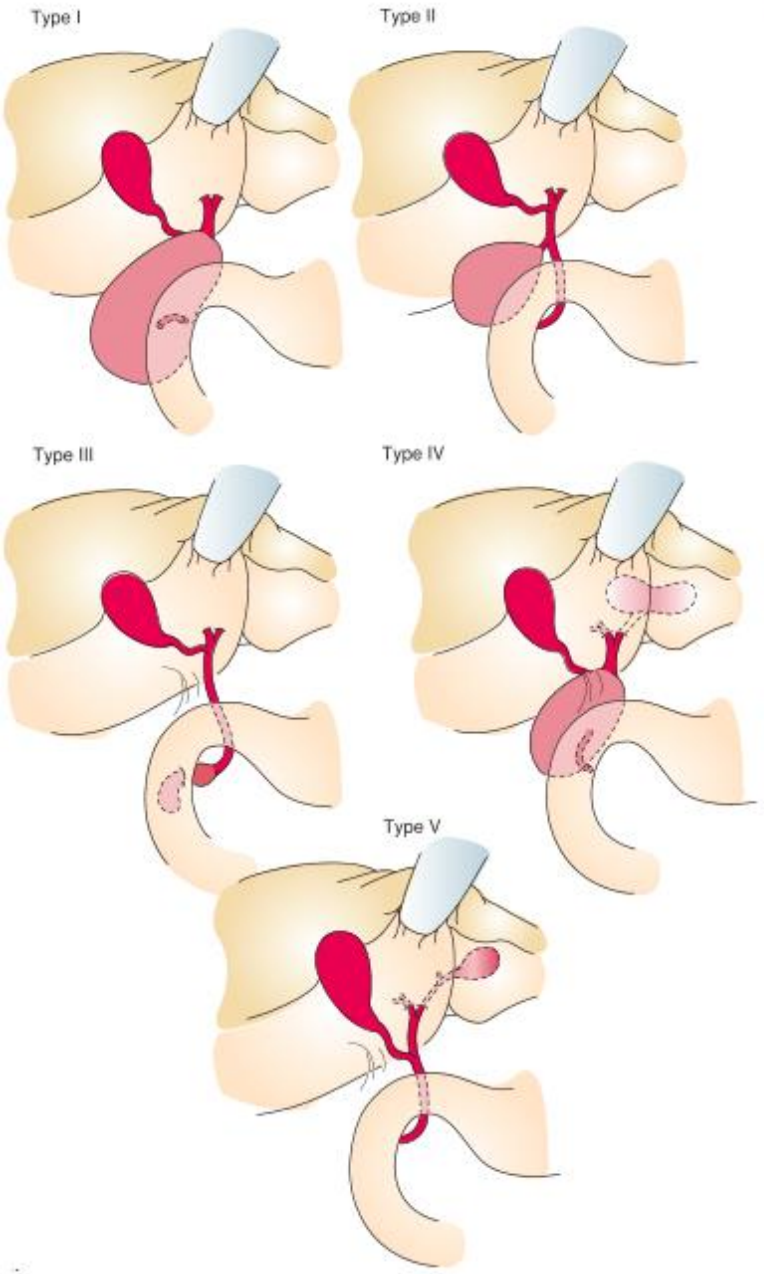
Definition: cystic dilatations of the bile ducts

Incidence: 1 in 100, 000

Possible mechanisms:
-Abnormal recanalization of primitive bile duct cords
-Inflammation caused by reflux of pancreatic secretions into CBD

Choledochal Cysts

Type I (50-85%): CBD dilated with normal intrahepatic duct
Type II (2%): isolated CBD diverticulum
Type III (1-5%): intraduodenal CBD cyst dilation
Type IV (15-35%): multiple cysts
A: intra and extra hepatic
B: extrahepatic only
Type V (20%): one or more cyst dilations of intrahepatic ducts with no extrahepatic duct involvement



Choledochal Cysts

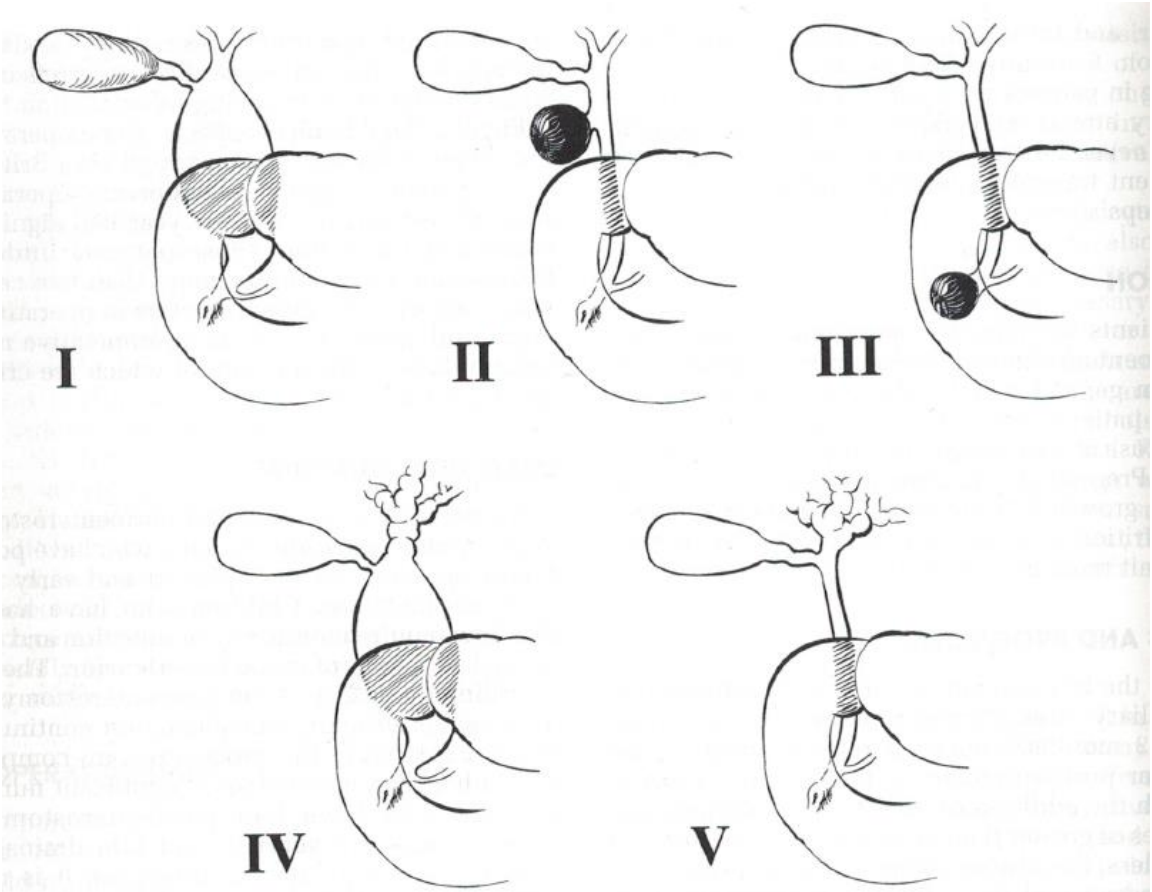


Figure 44–6. Classification of choledochal cyst.[41, 42]

Choledochal Cysts

Presentation and Diagnosis:
Jaundice
Abdominal pain
Abdominal mass

Imaging:
U/S
Radioscintigraphy (technetium 99m)
MRCP or ERCP

Choledochal Cysts

Treatment:

- Excision of cyst with direct anastomosis of proximal normal bile duct to Roux en Y loop of jejunum
- External drainage is temporizing measure reserved for emergency decompression
- Cyst enterostomy should not be done
  - High rate of stricture
  - Possibility of biliary malignancy

Choledochal Cysts

Treatment:

Type I

-Surgical exploration and cholangiography

-Cholecystectomy

-Primary excision with Roux en Y

Type II

-Resection and reanastomosis of CBD

Type III

-Transduodenal approach – excision of cyst and sphincteroplasty

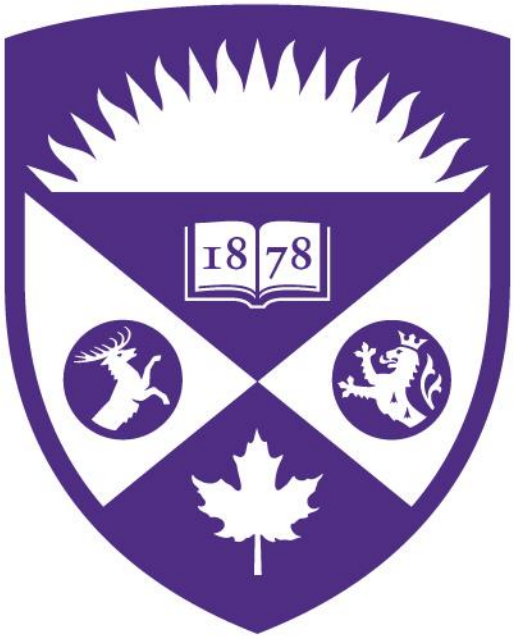
Type IV and V intrahepatic cysts

-Roux en Y

Choledochal Cysts

Complications:

- Cholangitis
- Stricture formation
- Choledocholithiasis
- Cholangiocarcinoma
  - Incidence is 2.5-5% if incomplete excision of choledochal cyst
  - Survival time is <1 year after cancer detected



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References

1) Mulholland, M et al. Greenfield's Surgery: Scientific Principles and Practice 5th edition. 2011.'
2) Ashcraft et al. Pediatric Surgery 3rd edition. 2000.
3) Moss, L. R. et al. Case Studies in Pediatric Surgery. 2000.
4) UpToDate