

PEDIATRIC SURGERY

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OVERVIEW

- ❖ Head and Neck
- ❖ Congenital Diaphragmatic Hernia
- ❖ Chest and Mediastinum
- ❖ Congenital Abdominal Wall Defects
- ❖ Congenital Obstructions of the GI Tract
- ❖ GastroEsophageal Reflux Disease (GERD)
- ❖ Hypertrophic Pyloric Stenosis
- ❖ Appendicitis
- ❖ Intussusception
- ❖ Necrotizing Enterocolitis (NEC)
- ❖ Hernias, Hydrocoeles and Undescended Testicles
- ❖ The Biliary Tract
- ❖ Malignancy
- ❖ Trauma

APPENDIX I

APPENDIX II

APPENDIX II

SUMMARY

PEDIATRIC SURGERY – AN OVERVIEW

Dr. G.M. Lees

Why Pediatric Surgery?

You have the ability to give your patient a whole life ahead of them.

The cardiac surgeons only give their patients another 3-5 years of golf.

The trauma surgeons only give their patients another 30 years of stealing cars and doing drug deals.

We will concentrate our attention on a few of the most important topics in surgery as it pertains to children. These notes will be applicable to the “surgery rotation” in Phase III and where appropriate, illustrations, pictures and Xrays accompany the text...

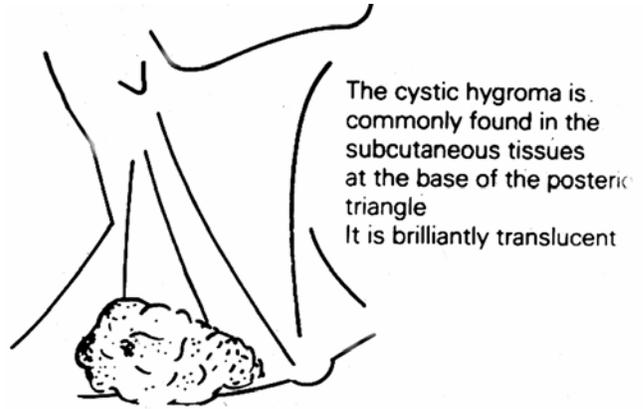
A few basic principles in all cases should be applied as regards fluids and electrolytes, blood volume (Appendices I & II), hypothermia and nutrition. All children (in particular though, the newborn and premature) are at risk for **hypothermia** before, during and after surgery. A large body surface area, minimal amount of insulating fat and reliance on nonshivering thermogenesis all contribute to this increased susceptibility to hypothermia. As glycogen stores are depleted rapidly and brown fat stores are used up, energy needs must be met, primarily with infusions of glucose intravenously. Later on, during the course of the child’s illness, total parenteral nutrition (TPN) may be required.

A very brief discussion of a few of the more important surgical problems in children follows.

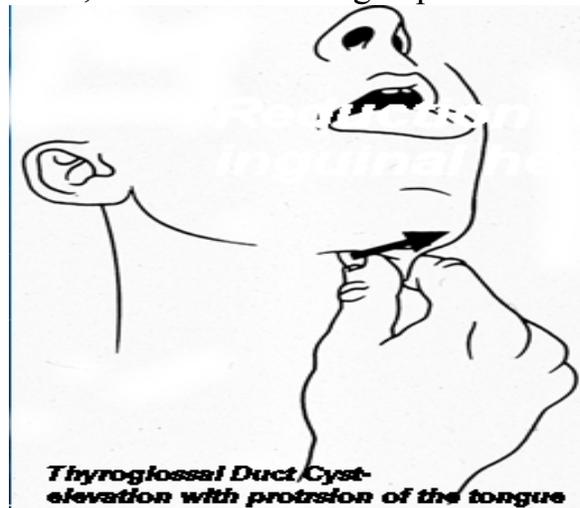
HEAD AND NECK

An assortment of congenital anomalies occurs in this area:

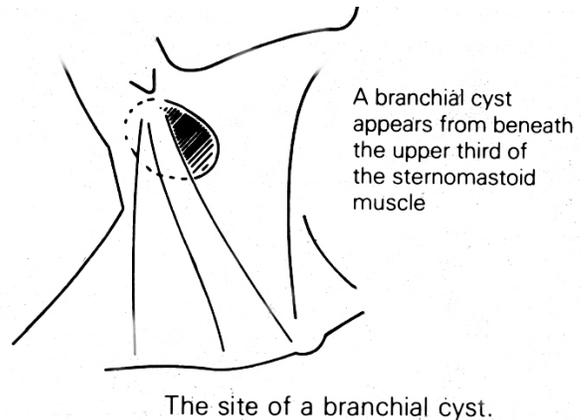
Cystic Hygroma: Found in the base of the posterior triangle. N.B. transillumination.



Thyroglossal Duct Cysts: Found in the midline, elevates with tongue protrusion.



Branchial Cleft Cysts and sinuses: Found under upper 1/3 of sternomastoid muscle.



Lymphadenopathy may be due to infection (viral, pyogenic, tuberculous – typical or atypical) or neoplasia (lymphoma, Hodgkin’s disease, metastases).

Lymphoproliferative Disorders

- **Constitutional Symptoms**

Fever, weight loss, pruritus (“B” – symptoms)

- **Mediastinal Disease**

PFT’s – Flow-Volume Loop

CT:– ratios & tracheal cross sectional area

- **Biopsy (cervical / prescalene lymph node)**

Biggest, lowest, deepest

FRESH – cell marker studies

Atypical Tuberculous – no skin involvement:



Atypical Tuberculous – skin involvement; described as a chronic suppurating wound, typically non-tender



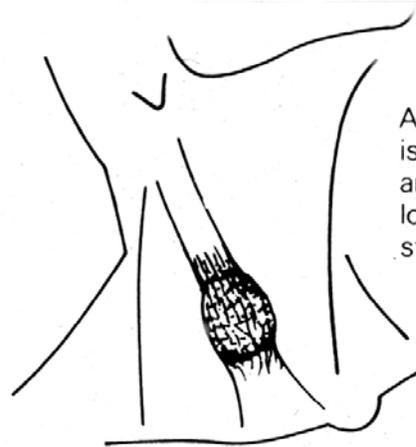
Suppurative Lymphadenitis – *staph*, *strep*, *haemophilus* (“non-specific”). *Specific infections: Actinomyces, scrofula (tuberculosis) and cat-scratch disease.*



Dermoid cysts may occur at the lateral corner of the eyebrow (external angular dermoid) or on the scalp or midline of the neck.



Torticollis – marked shortening of the sternocleidomastoid muscle:



A 'sternomastoid tumour' is an area of oedema and necrosis in the lower third of the sternomastoid muscle

Torticollis The site of a sternomastoid tumour.

Mucocele – typical location is lower lip (left image), but can also be found involving the floor of mouth, termed a “ranula” (right image)

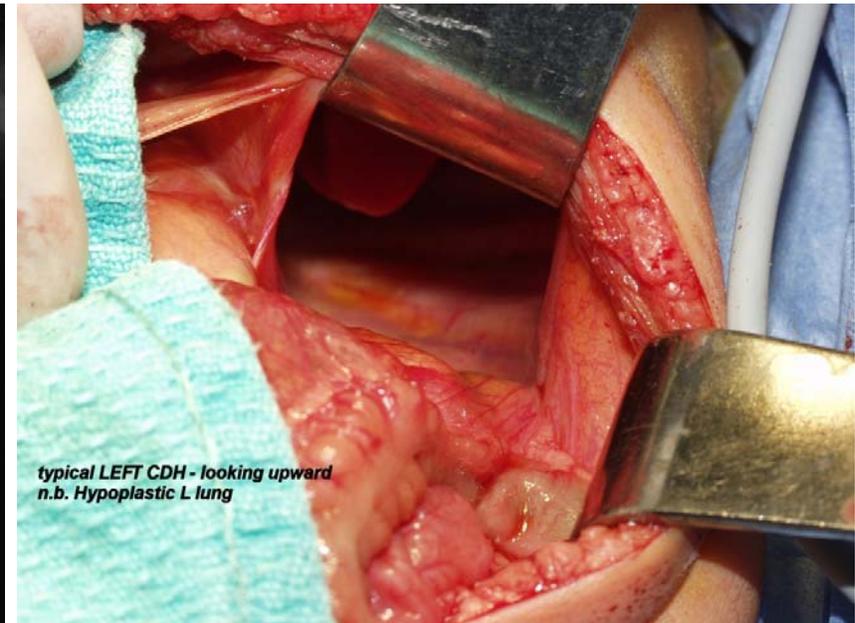


CONGENITAL DIAPHRAGMATIC HERNIA



Defects in the diaphragm (foramen of Bochdalek) result in herniation of the abdominal viscera into the chest (most often the left) in-utero. The diagnosis of C.D.H. can be suspected in the newborn who is dyspneic and has a **scaphoid abdomen**.

The resulting compression of the lung results in ipsilateral hypoplasia, and in severe cases, bilateral **pulmonary hypoplasia**.



The hypoplasia in turn results in **pulmonary hypertension** and a **right-to-left intra and extra cardiac shunt** i.e., the unoxygenated blood from the right side of the heart completely bypasses the lungs – a condition known as **P.F.C. (persistent fetal circulation)**.

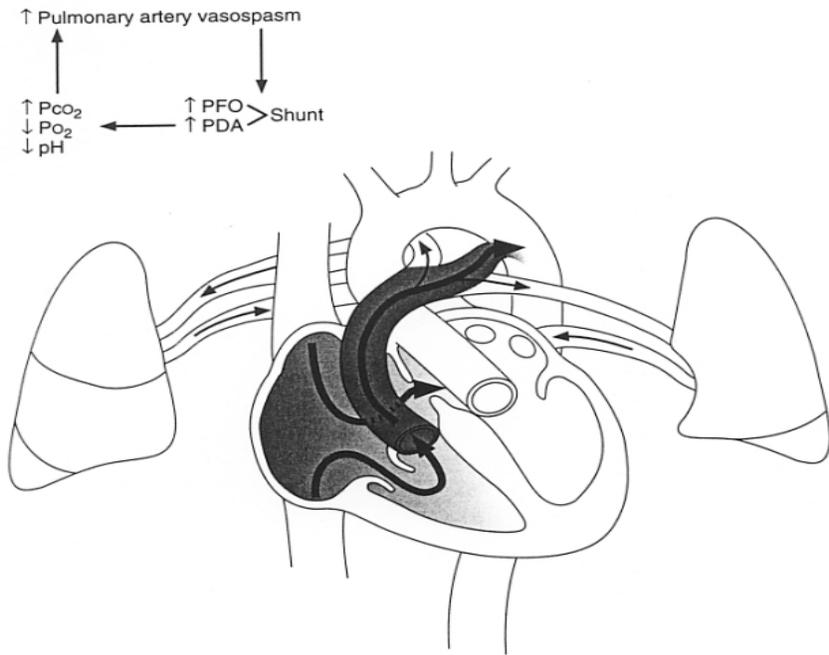
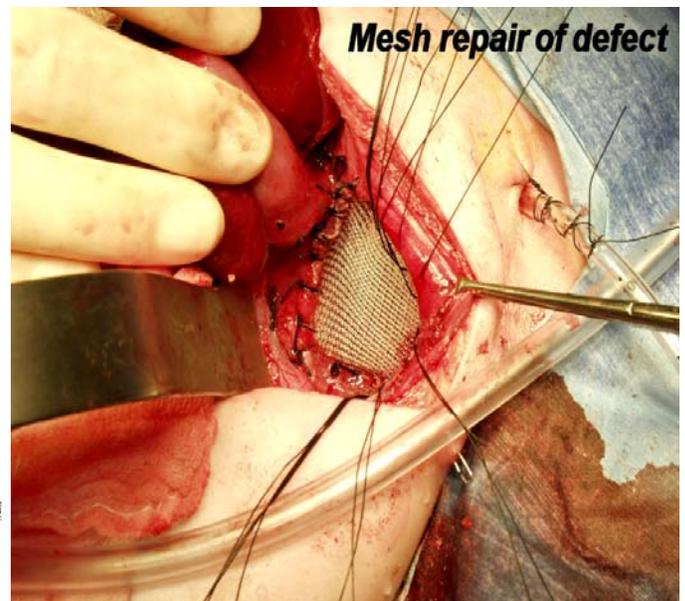
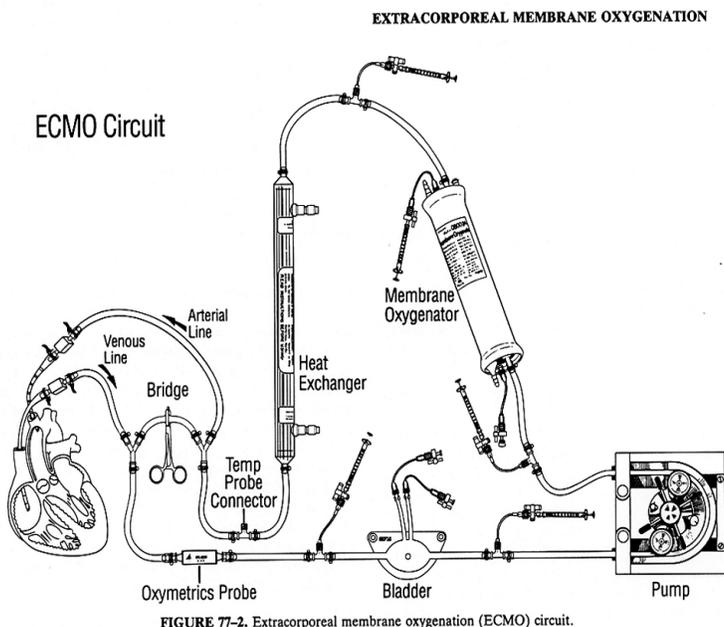


FIG. 7-31. The pathophysiology of persistent fetal circulation. Venous blood returning to the right heart preferentially flows (shunts) through the patent foramen ovale (PFO) or the patent ductus arteriosus (PDA) in the setting of severe pulmonary hypertension. This results in hypoxia, hypercarbia, and acidosis, which induce further increases in pulmonary hypertension and worsen the shunt. A vicious cycle of pulmonary hypertension and deteriorating gas exchange develops and can lead to the demise of the patient if not interrupted.

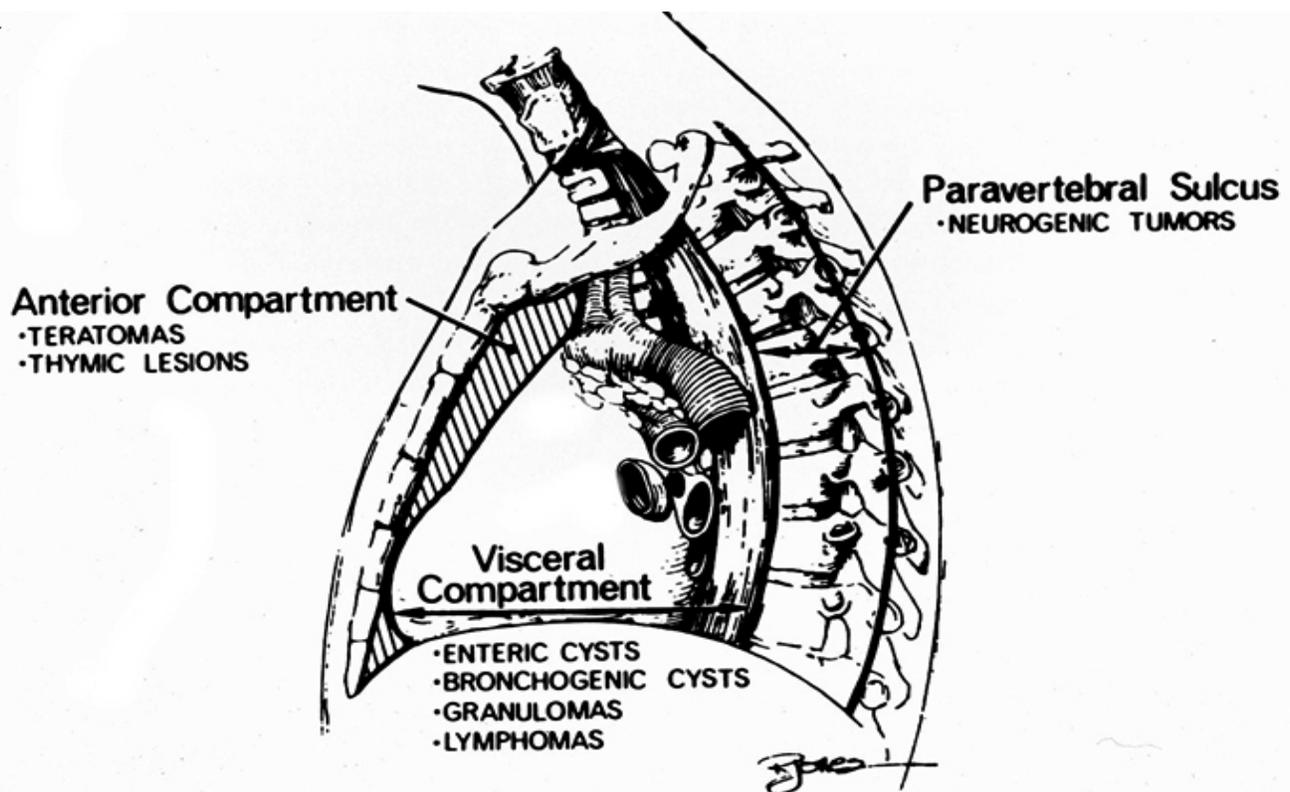
A large number of these children will die without specialized ventilation (“gentle” ventilation and “permissive hypercapnea” to avoid barotrauma), nitric oxide (a selective pulmonary vasodilator) or **E.C.M.O.** (extracorporeal membrane oxygenation). Correction of hypoxaemia, hypercarbia and acidosis are far more important than the anatomic repair and always takes precedence over it.



CHEST AND MEDIASTINUM

There are a variety of congenital and acquired conditions in the chest (pleura and lungs) and mediastinum affecting infants and children. Dyspnea/tachypnea are the usual presenting symptoms – primarily due to the effect of a “space-occupying” lesion.

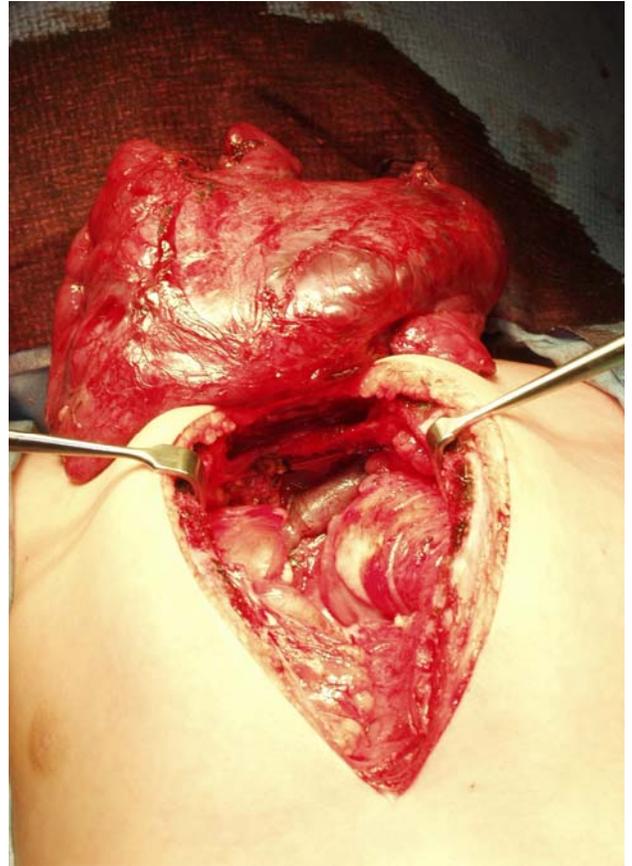
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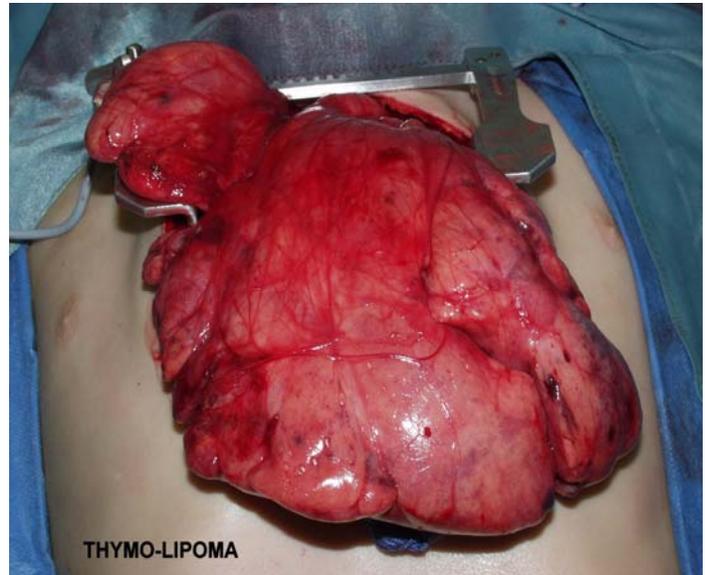
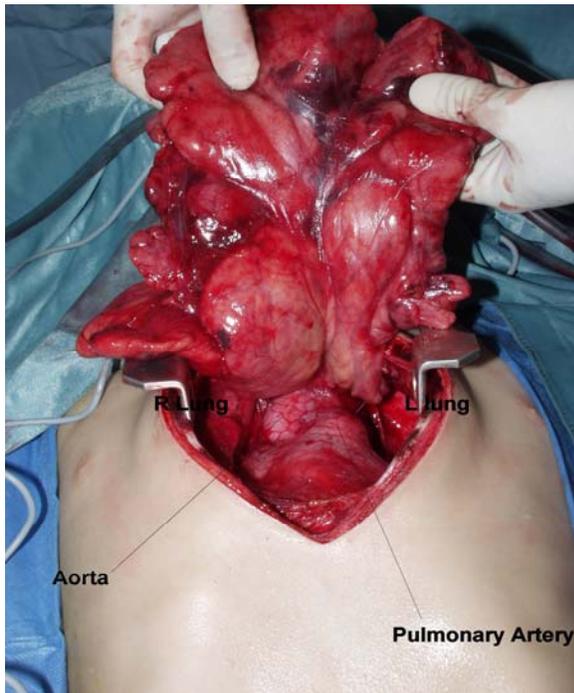
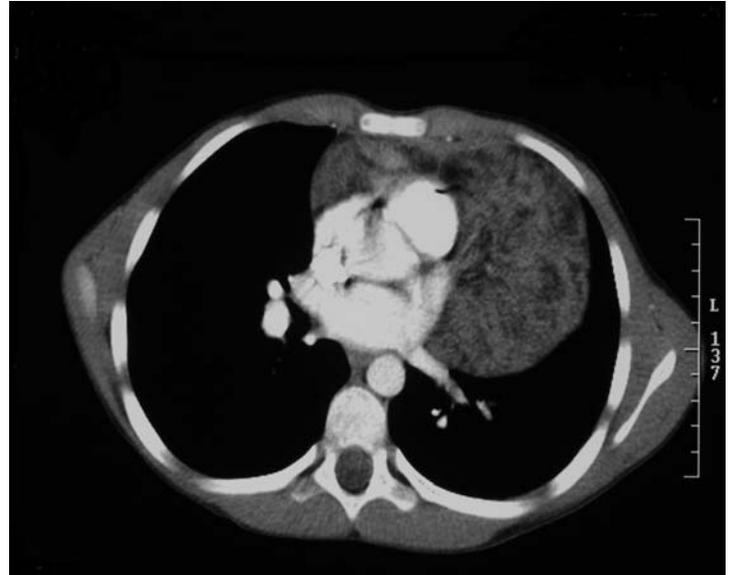
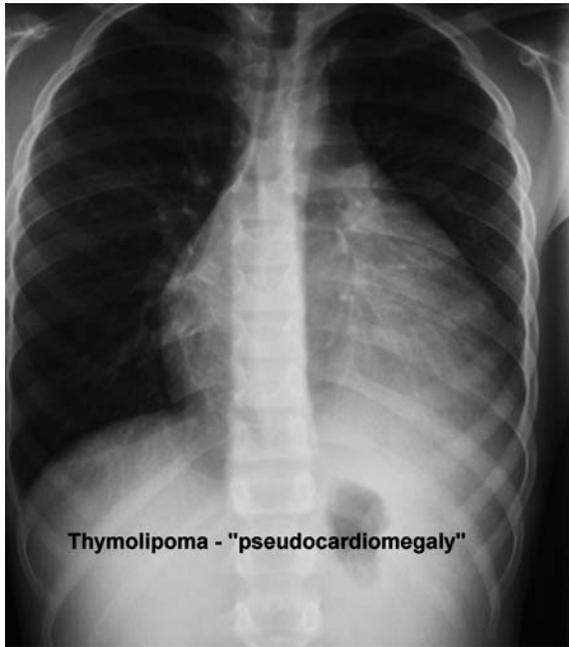
1. **Lymphoma** – is the 3rd most common childhood cancer, with ~60% non-Hodgkin’s lymphomas, and ~40% being Hodgkin’s lymphoma. CXR/CT may show a mediastinal mass or lymphadenopathy.



2. **Teratoma:** is the third most common mediastinal tumor, and is most commonly located in the anterior thorax and contains all three layers of developmental tissue (endoderm, mesoderm, and ectoderm).



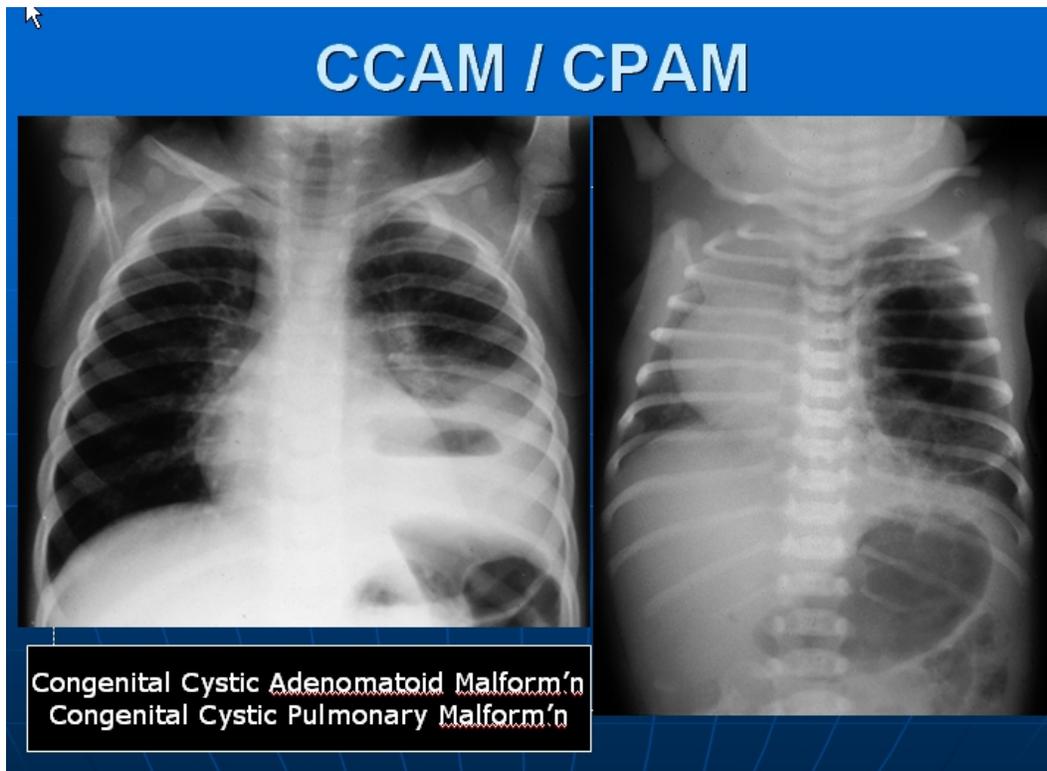
3. **Thymolipoma:** a very rare, benign, anterior mediastinal tumor. May see “pseudocardiomegaly” on CXR (left image), CT scan shows large thymus mass (right image):



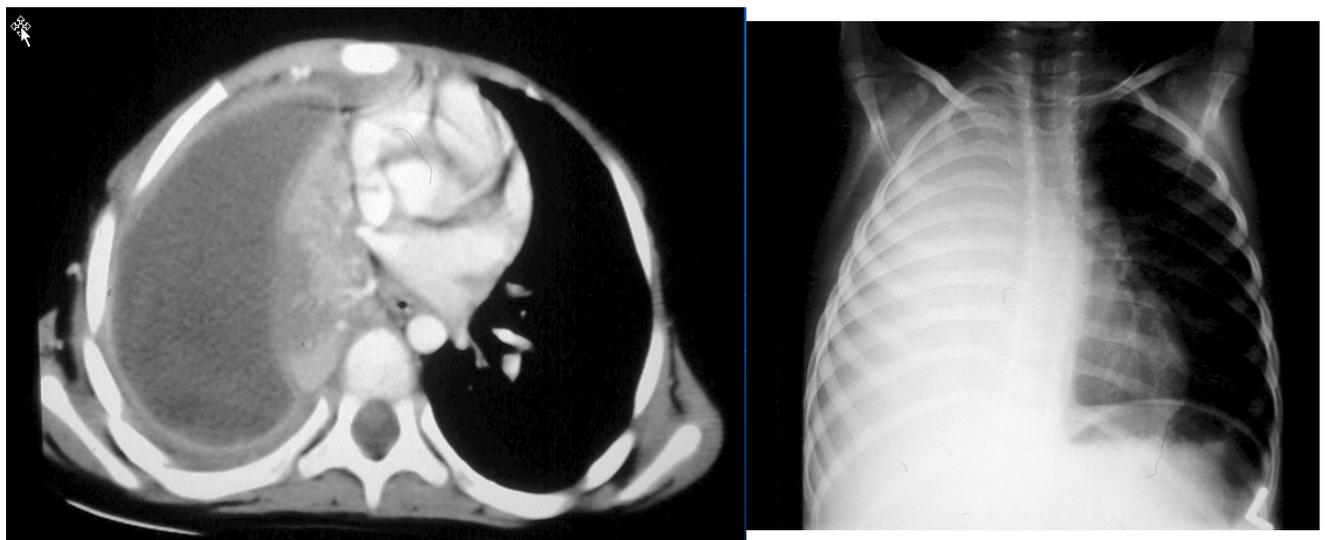
Dyspnea – Childhood

- Diaphragmatic Hernia
- Congenital Cystic Lung Disease
- Pneumonia
- Parapneumonic empyema
- Pneumothorax – spontaneous(secondary to apical bullae)
- Foreign Body
 - Emphysema (due to an *inspiratory check valve*)
 - Pneumonia
 - Lung Abscess / Bronchiectasis (secondary infection)

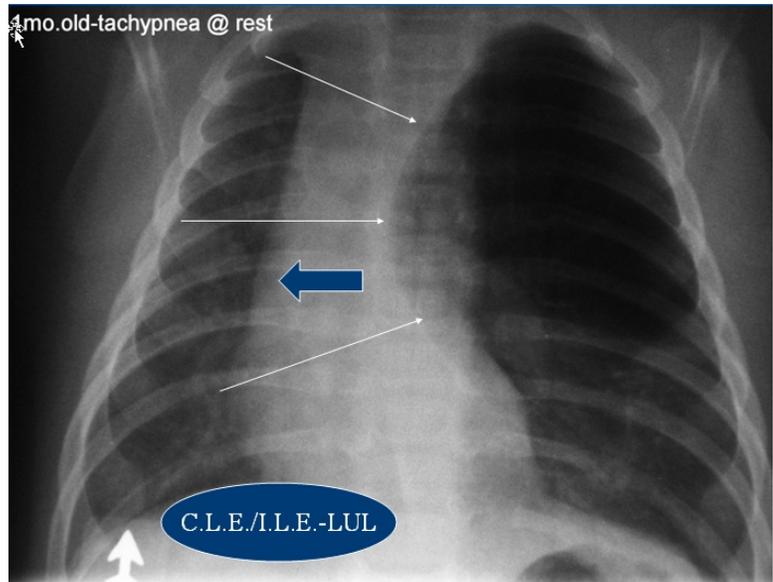
Congenital Cystic Adenomatoid /Pulmonary Malformation: up to an entire lobe of the lung is replaced with non-functional cystic tissue that has an abundance of bronchial structures. There may be an aberrant blood supply.



Empyema – a collection of pus within the pleural cavity, often originating from a pneumonia and may present with symptoms of fever, chest pain, cough, and shortness of breath. CT scan below shows a large right sided pleural effusion with atelectasis and mediastinal shift. CXR shows complete opacification of the right hemithorax with tracheal and mediastinal shift left.



Congenital Lobar Emphysema
– is characterized by air retention and overinflation, resulting in decreased respiratory function. Congenital lobar emphysema is commonly identified within the first two weeks of life, and may present with wheeze, shortness of breath, and peripheral cyanosis.



CONGENITAL ABDOMINAL WALL DEFECTS

Umbilical Hernias



Umbilical hernias are extremely common and generally will resolve with time. Very rarely a loop of bowel may **incarcerate** resulting in a bowel obstruction. Repair of the hernia then becomes urgent. Elective repair is left until the child is 2-4 years of age.

Gastroschisis



Gastroschisis is a condition characterized by a full thickness abdominal wall defect occurring to the right of the midline through which most or all of the abdominal viscera herniate. The herniated bowel is not protected by a covering layer/membrane. Thus, **heat and fluid loss and infection** are the major concerns initially; malnutrition from failure of gut function to return occurs late. Shown here are some complications of gastroschisis: intestinal atresia and small bowel perforation in the upper image, and obstruction resulting in ischemia and intestinal necrosis in the lower image.

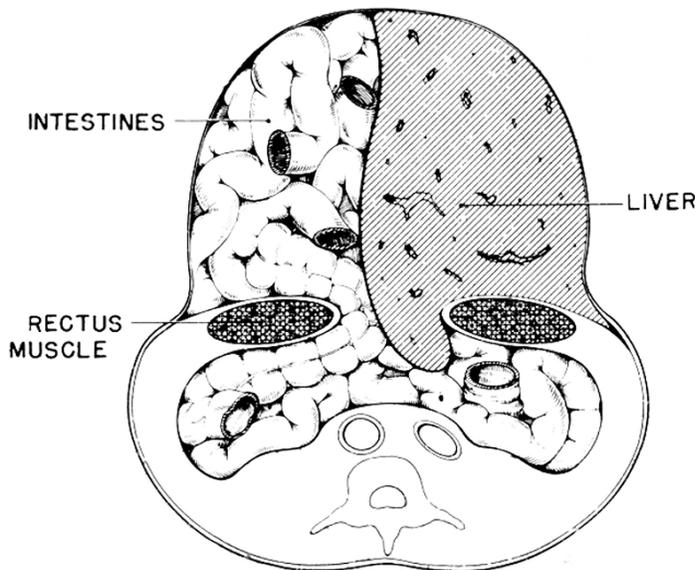


These babies are frequently SGA (small for gestational age) or premature, but rarely have other life-threatening anomalies, and so if they are treated properly, survival is the norm (greater than 95 %). Antenatal ultrasounds are helpful in early diagnosis and management planning (left image). Shown below on the right is a spring-loaded silastic bag, used to gradually reduce the bowel into the peritoneal cavity.

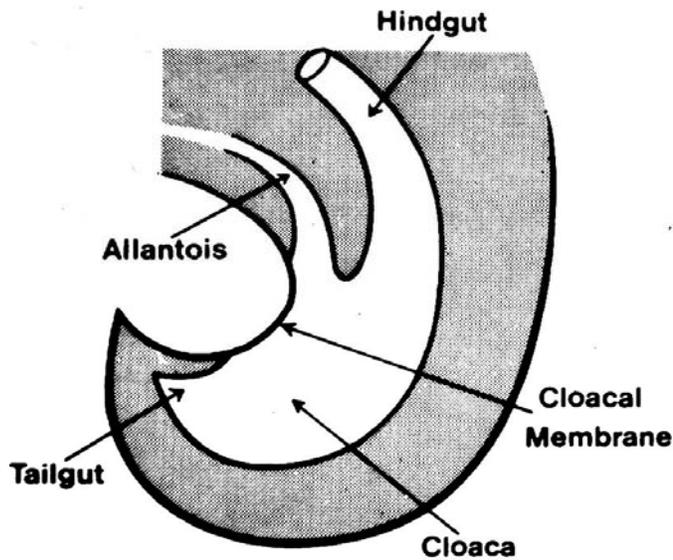
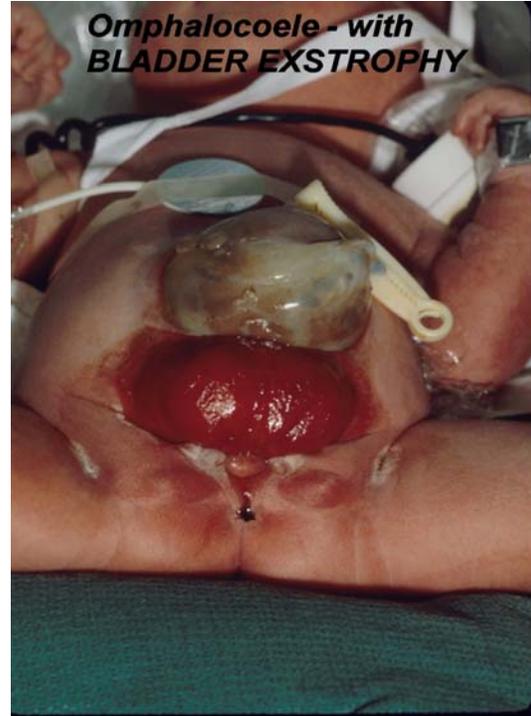


Omphalocele

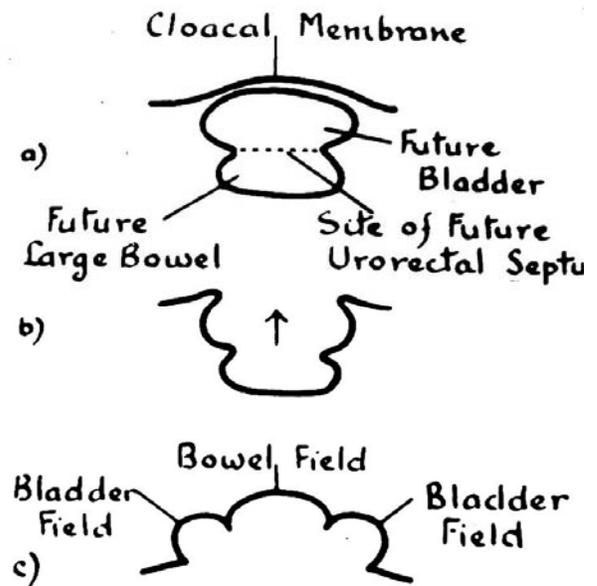
Omphalocele is a condition arising from a central abdominal wall defect, but where the viscera are covered with a membranous sac (peritoneum, chorion, etc.). These babies frequently have **other potentially lethal abnormalities** (including cardiac and chromosomal), which need to be considered prior to, during and after surgery.



The most extreme examples of the abdominal wall defects are the **exstrophic anomalies** (vesiconintestinal fissure): exstrophy of the cloaca (left image) and exstrophy of the bladder (right image).



Cloacal Exstrophy - Embryology

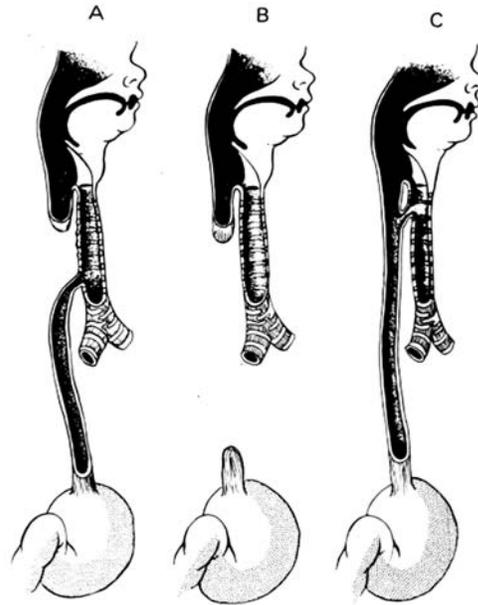
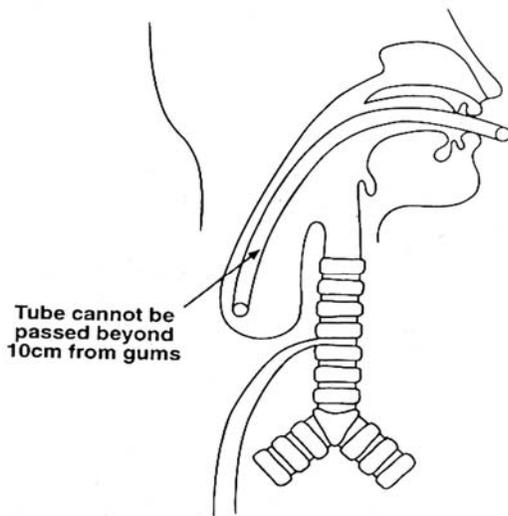


Clinical Findings of Gastroschisis and Omphalocele

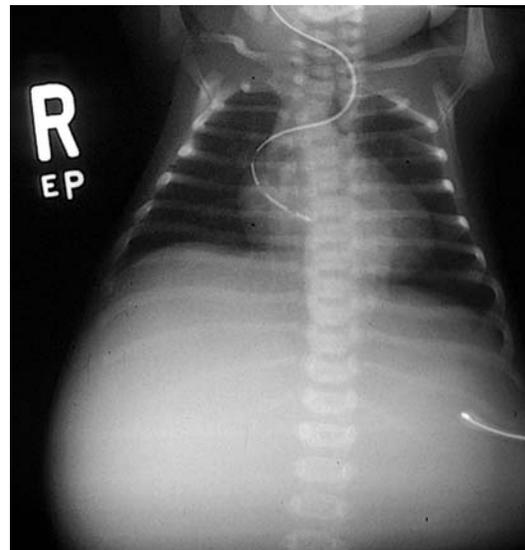
Factor	Gastroschisis	Omphalocele
Location	Lateral to Cord (right)	Umbilical Ring
Defect Size	Small (2 to 4 cm)	Large (2 to > 10 cm)
Cord	Normal Insertion	Inserts in Sac
Sac	None	Present
Contents	Bowel, Stomach	Liver, Bowel, etc.
Malrotation	Matted, Inflamed	Normal
GI Function	Present	Present
	Prolonged Ileus	Normal
Associated Anomalies	Unusual (Intestinal	Common (30 to 70%)
Syndromes	Atresia 15%)	Beckwith
	Not Observed	Wiedemann,
		Trisomy 13 – 15,
		Trisomy 16 – 18,
		Lower Midline
		Syndrome,
		Pentalogy of Cantrell

CONGENITAL OBSTRUCTIONS OF THE G.I. TRACT

Babies are continually swallowing and digesting amniotic fluid in-utero. The fluid is absorbed from the G.I. tract, enters the fetal circulation and is ultimately circulated to the mother by the placenta. If the fetus cannot swallow or digest and absorb the fluid, an excess of amniotic fluid may develop – **polyhydramnios**. This is one of the cardinal features of esophageal atresia and proximal obstructions of the G.I. tract.



Esophageal atresia can occur with (left image) or without (right image) an associated tracheoesophageal fistula: Intra-abdominal air means there *must* be a fistula from the trachea to the distal esophagus/stomach.



Abdominal distention and failure of passage of meconium are two other cardinal features of obstruction. It is important to know that a baby may have an obstruction with one or more of the above features missing! **Bilious vomiting** virtually always occurs with a mechanical obstruction.

GREEN VOMIT MUST BE CONSIDERED A SURGICAL EMERGENCY until proven otherwise. Very proximal obstructions (duodenal or upper jejunal) frequently are seen with little or no abdominal distention.



Pediatric G.I. Obstruction – Neo-Natal

- E.A. and T.E.F.
- Duodenal
 - Volvulus (extrinsic)
 - Atresia (intrinsic)
 - **X-Ray: “double-bubble”**
- Jejuno-Ileal Atresia
- Meconium Ileus
 - (*cystic fibrosis)
- Hirschsprung’s Disease (aganglionosis)
- Meconium Plug Syndrome
- Small Left Colon Syndrome (diabetic mother)
- Imperforate Anus

Polyhydramnios BILIOUS VOMITING Distention Failure to Pass Meconium

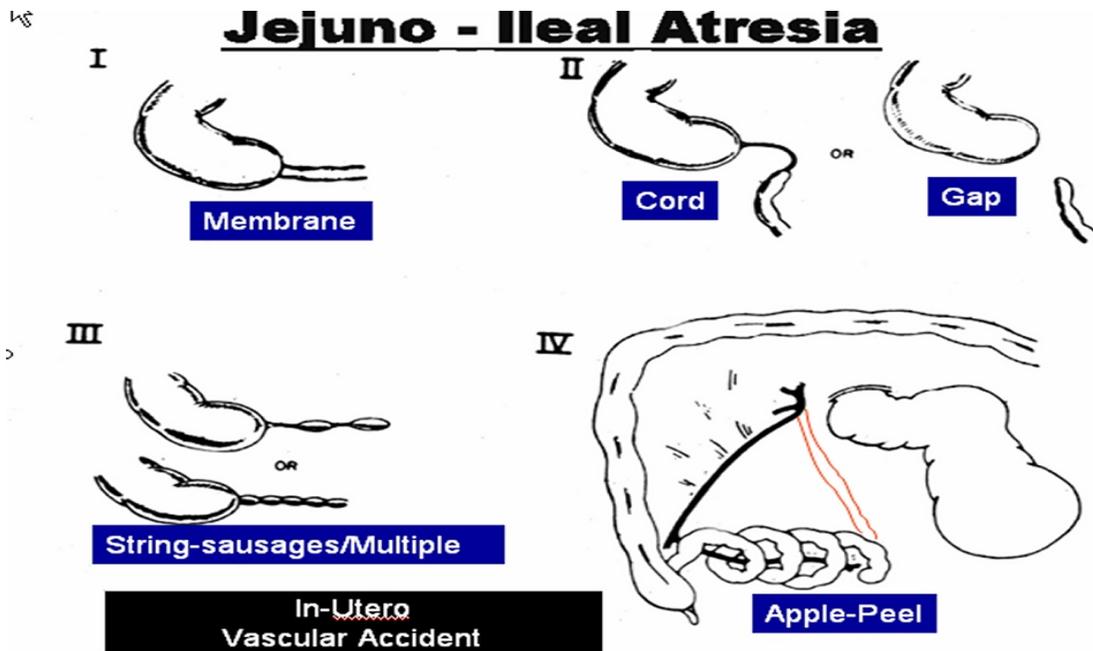
Siblings with cystic fibrosis



Duodenal Stenosis

Duodenal Stenosis

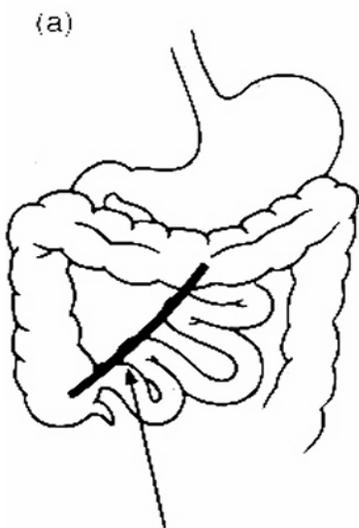
Duodenal Atresia-no Distal gas



It is important to remember that if an anomaly occurs in one area, look for other anomalies: e.g. the **VACTERR** association (vertebral, anus, cardiac, tracheo-esophageal, radius and renal).

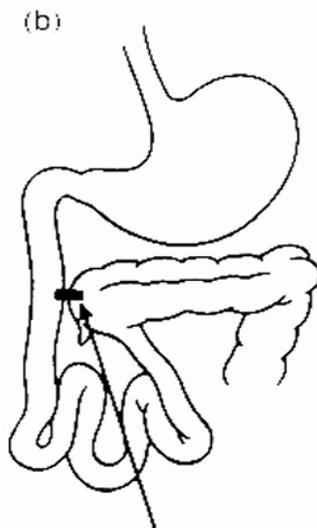


Malrotation with mid-gut volvulus is the most lethal form of bowel obstruction occurring in the newborn period and not infrequently presents much later in childhood. **A newborn infant who passes flatus or transitional stool, then develops signs of a bowel obstruction (i.e., bilious vomiting) has malrotation and a mid-gut volvulus 'til proven otherwise!** Obstruction of the superior mesenteric artery and vein occurs as the bowel twists, and ischaemic necrosis and gangrene occurs rapidly – within a few hours.



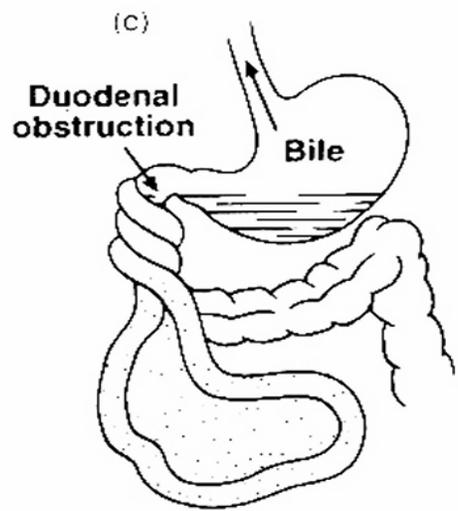
Wide base of mesentery

Normal

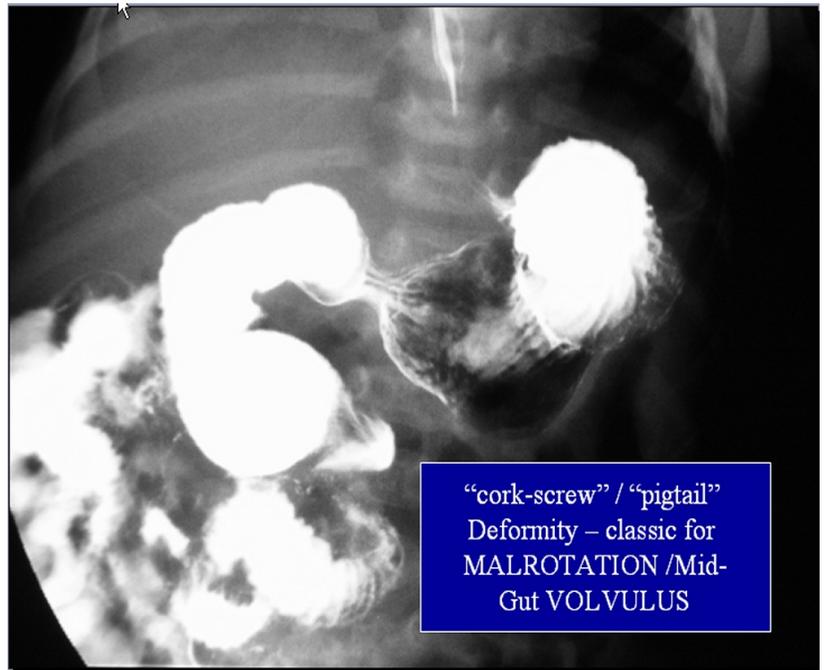
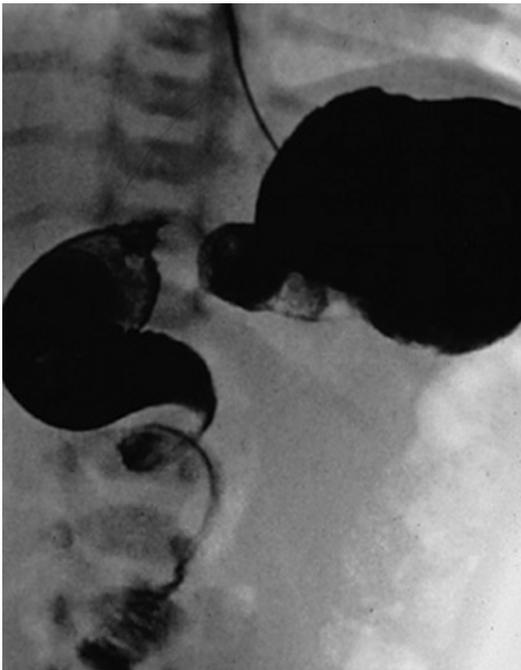


Narrow mesenteric attachment

Malrotation



Malrotation with volvulus



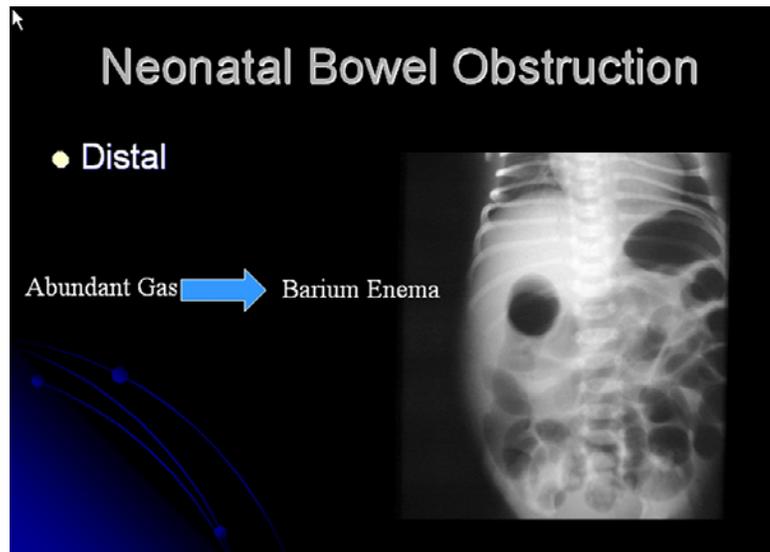
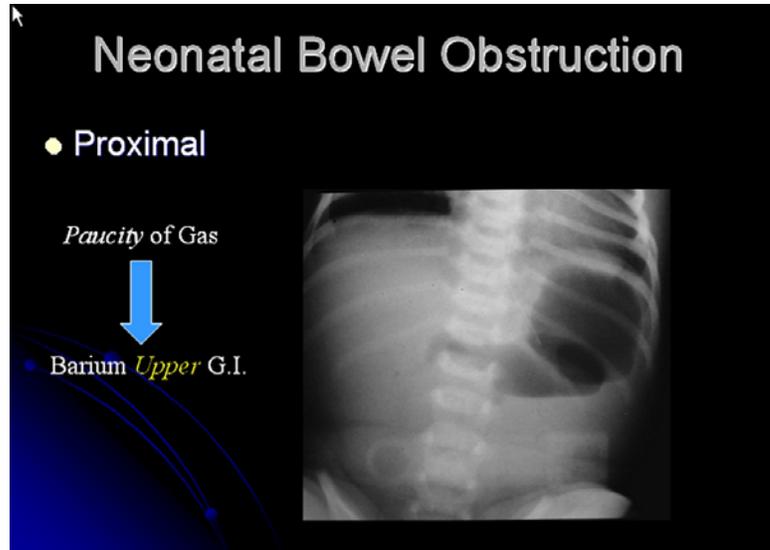
"cork-screw" / "pigtail"
Deformity – classic for
MALROTATION /Mid-
Gut VOLVULUS

Most of the above conditions are diagnosed by perinatal ultrasound, history, thorough physical examination and plain x-rays.

Contrast studies (barium upper G.I. or barium enema) are used **selectively** to diagnose the level of obstruction: **upper G.I.**, if the suspected obstruction is **proximal**; **barium enema** if the obstruction is likely colonic or terminal ileum. i.e **distal**

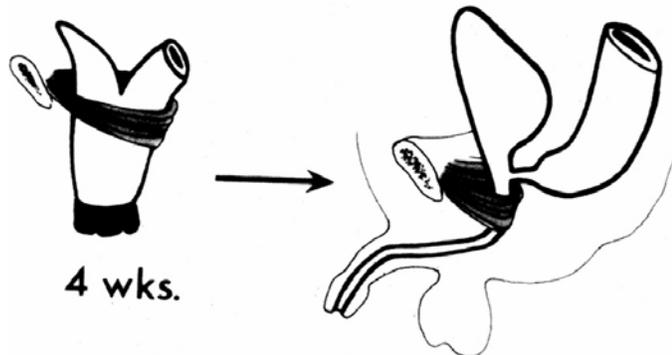
Gastrograffin, because of its hyperosmolarity (1600mosm/L) is **NEVER** given by mouth or N/G tube because of the risk of aspiration and subsequent pulmonary edema.

Meconium peritonitis refers to in-utero perforation of the bowel with scattered **calcification** in the peritoneal cavity seen on plain x-ray. The cause of the perforation may be an isolated vascular accident or meconium ileus – therefore **cystic fibrosis** must be excluded.

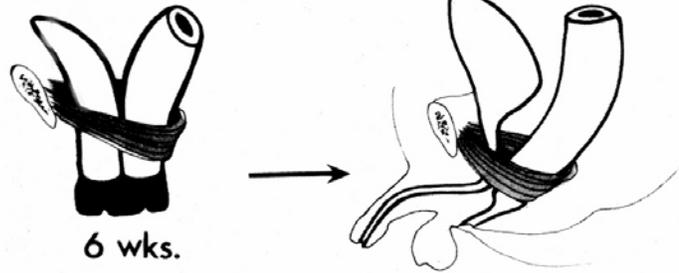


An **imperforate anus** or **anal atresia** is a birth defect in which the rectum is malformed. There are several forms of imperforate anus:

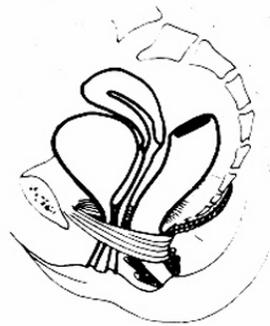
MALE: A **high imperforate anus**, in which the colon is much higher up in the pelvis and there is a fistula connecting the rectum and the bladder or urethra..



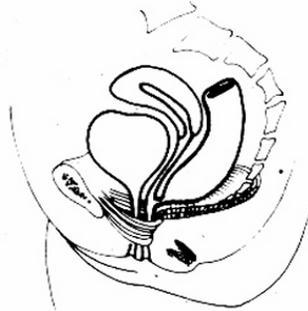
Male: A **low imperforate anus**, in which the colon remains close to the skin, and the anus may be stenosed or missing altogether.



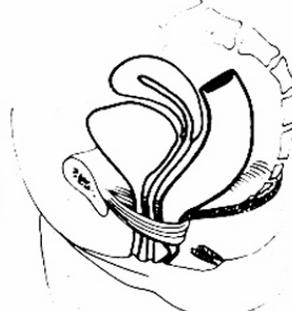
Female: images are shown below for high and low imperforate anus:



normal



high I.A.

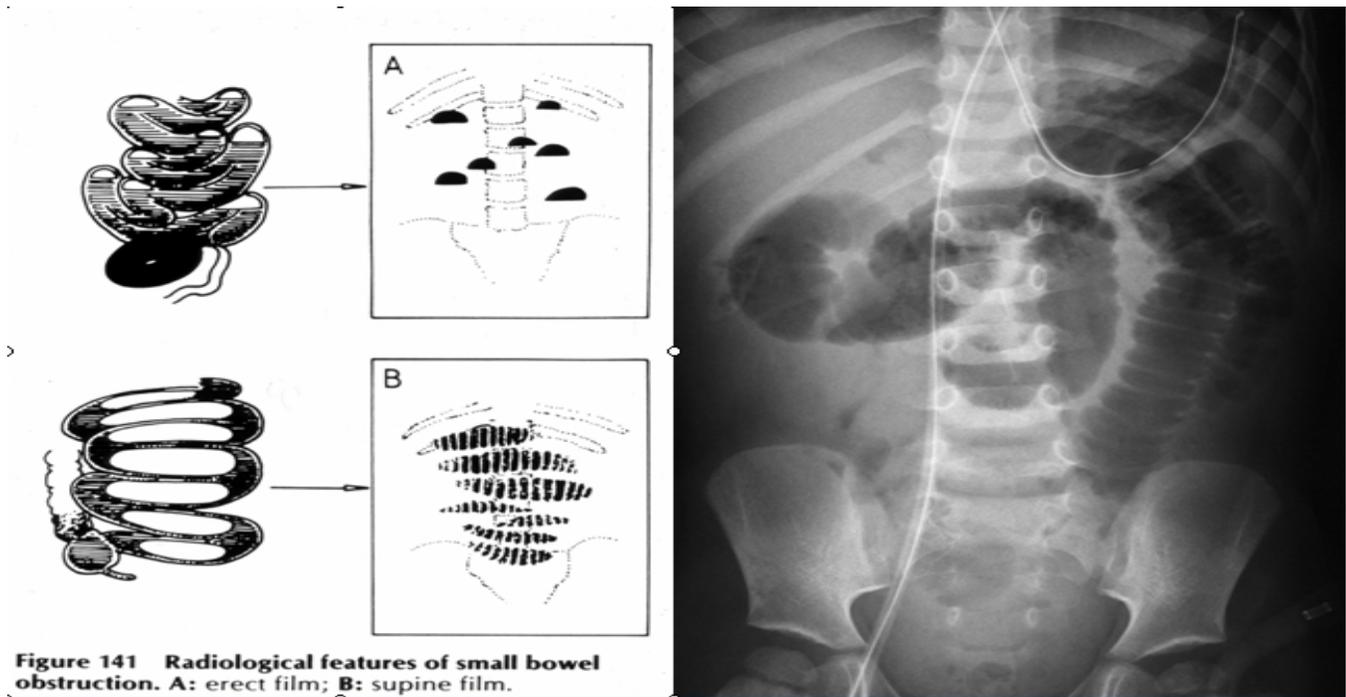


low I.A.

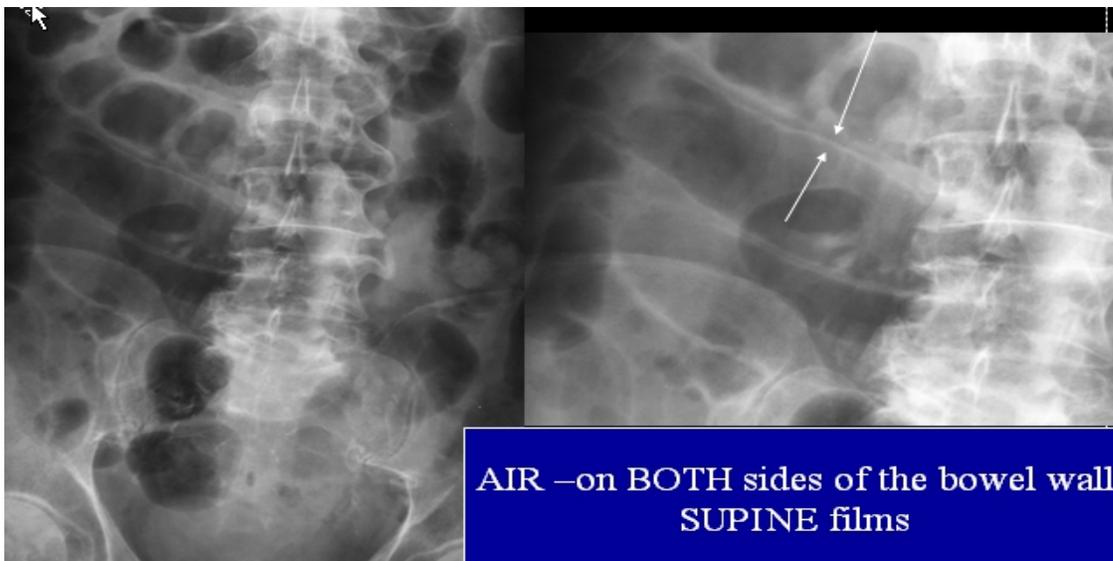
A **CLOACA** (Latin: "sewer") refers to a complex anomaly where the genitourinary and gastrointestinal tracts empty into one common channel.

POST-NATAL BOWEL OBSTRUCTION

A differential diagnosis for bowel obstruction in infancy and childhood would include: volvulus, hypertrophic pyloric stenosis, inguinal hernia incarceration, adhesions, intussusception, abscess(e.g. **appendiceal**), and tumor. The initial goal in diagnosing a bowel obstruction is to differentiate between small and large bowel:

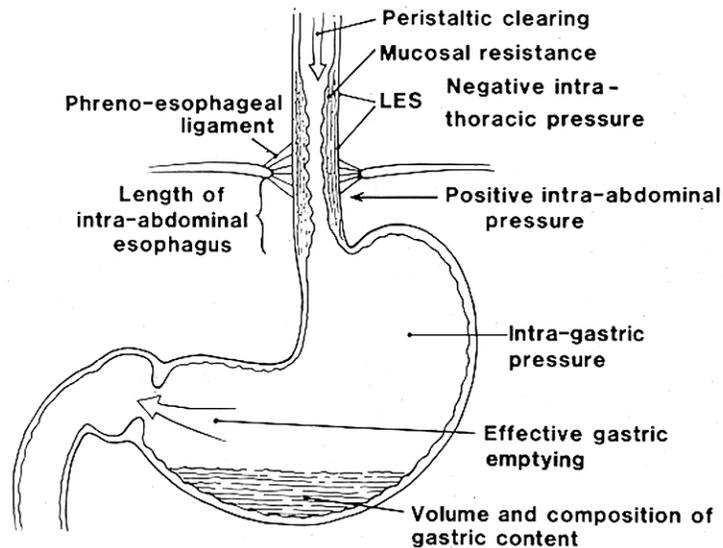


Large bowel typically has a greater degree of distention than small bowel, and is characterized by haustra. Large bowel obstruction is characterized by large peripheral loops of bowel, whereas small bowel obstructions demonstrate central loops of bowel distention on imaging. Untreated obstruction can result in intestinal ischaemia and necrosis with bowel perforation-diagnosed clinically or by the presence of free air.



GASTROESOPHAGEAL REFLUX DISEASE (G.E.R.D.)

G.E.R.D. is a common condition in infants and older children. One may see **respiratory problems** secondary to aspiration, esophageal injury from prolonged exposure to gastric contents resulting in **esophagitis and failure to thrive**. **Stricture** and more rarely, **Barrett's esophagus** ("columnar-lined esophagus") are relatively infrequently seen as consequences of esophagitis.



Diagnosis is made on the basis of a clinical history along with imaging studies – barium swallows, radionuclide gastroesophageal reflux scan, endoscopy and **24-hour pH monitor** (the "gold standard" in cases difficult to diagnosis). Bronchoscopy with B.A.L. (bronchoalveolar lavage) may demonstrate the presence of lipid-laden macrophages.

Treatment is primarily medical with positional therapy, thickening of feeds, promotility agents and either H₂ receptor antagonists or Omeprazole. Surgery is reserved for those infants who fail medical therapy, which would include virtually all infants with respiratory symptoms on the basis of G.E.R.D. The standard operative procedure is a **Nissen Fundoplication**.

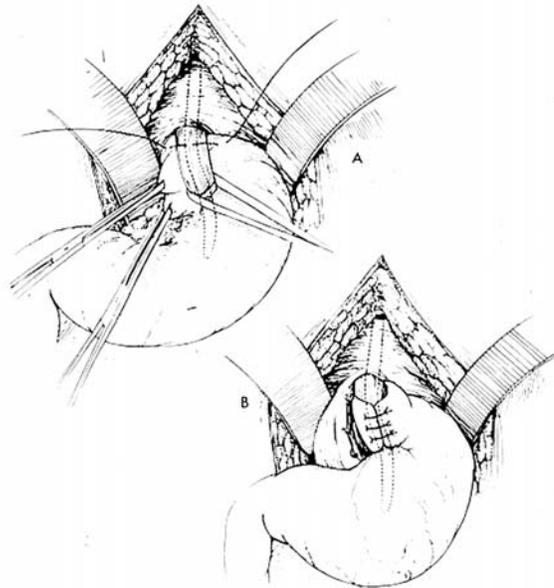


Fig 1. Nissen fundoplication. (A) First suture as fundus is wrapped around the abdominal esophagus. The tape passes around the GE junction. (B) Completed procedure shows construction of fundic wrap. Gastroesophageal tube, of size 18 or 20, ensures adequate lumen.

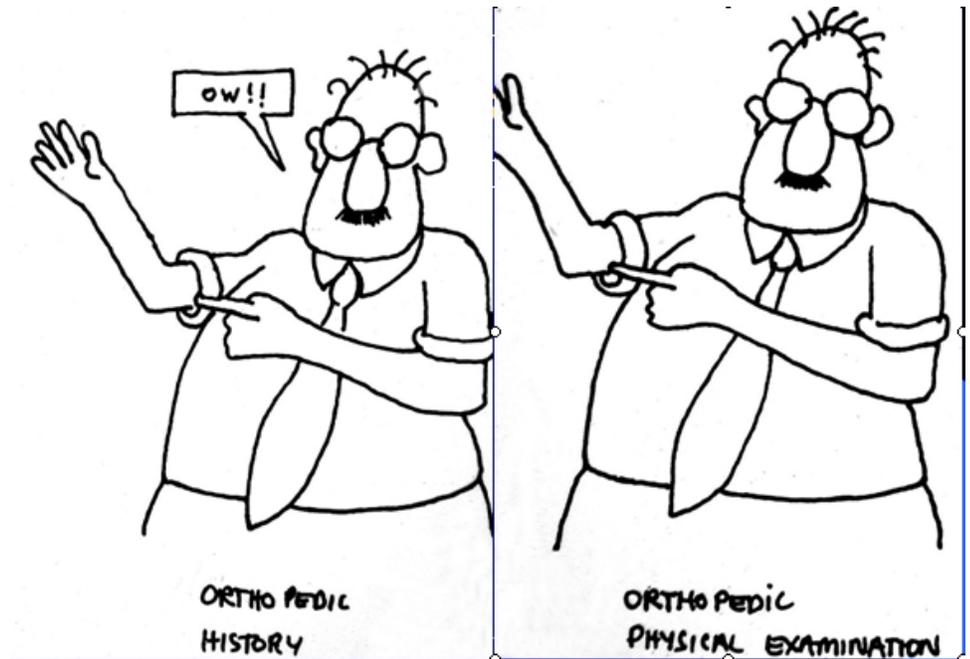
HYPERTROPHIC PLYORIC STENOSIS

H.P.S. is a relatively common condition occurring between 2 and 8 weeks of life. It is **NOT** congenital. **Projectile, NON-Bilious vomiting** in an otherwise apparently healthy infant is the classic presentation. The condition is more common in first-born males. A family history of H.P.S. may be present (particular in the mother). Dehydration with a **hypochloremic, hypokalemic, metabolic alkalosis** will be seen in varying degrees dependant upon the severity and duration of the vomiting. Treatment is rehydration and correction of the electrolyte abnormalities – followed by surgery (**Ramstedt Pyloromyotomy**) when the baby is stable.



APPENDICITIS

Appendicitis in the older child is usually a fairly straight-forward diagnosis if a thorough history and physical exam are obtained.). It is important to take a thorough history and characterize the pain (onset, severity, location, radiation, relieving and aggravating factors), and a rectal exam should always be considered part of the routine physical exam in children and adults with abdominal pain.



Patients classically will localize the point of maximal pain the “**McBurney’s point**”, located $\frac{1}{3}$ the distance from the ASIS (anterior superior iliac spine) to the umbilicus.



McBurney’s point

However, diagnosis of appendicitis can present many traps for the unwary in the younger child – typically the **toddler** (18 mos. to 3 yrs of age). However, in the younger child, the history may be atypical and the symptoms are frequently attributed to gastroenteritis. Abdominal pain preceding the onset of vomiting should raise one's suspicion. The incidence of **rupture** is higher in this group. Shown below are the abdominal x-rays for two different children with perforated appendicitis and bowel obstructions:



NEVER omit the **rectal examination** in a child/infant with abdominal pain.

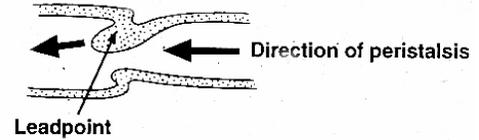


The bacteriology of appendicitis includes any/all gram negative and positive aerobes and anaerobes. The most common causative species is *Bacteroides fragilis*, but *Enterococci* (ex: *Streptococci*) and gram negative aerobes (ex: *Pseudomonas*, *E. coli*, *Proteus*, *Klebsiella*, etc.) are also common.

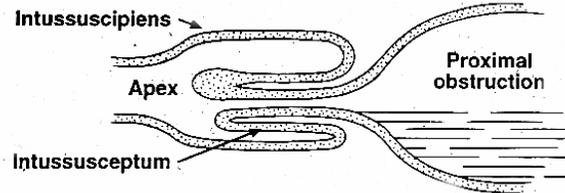
INTUSSUSCEPTION

Intussusception may be idiopathic (due to hypertrophied Peyer’s patches in the wall of the ileum) or secondary to a pathological “lead point” (e.g., Meckel’s diverticulum, polyp). The bowel essentially inverts itself into the lumen and tries to propel this segment distally.

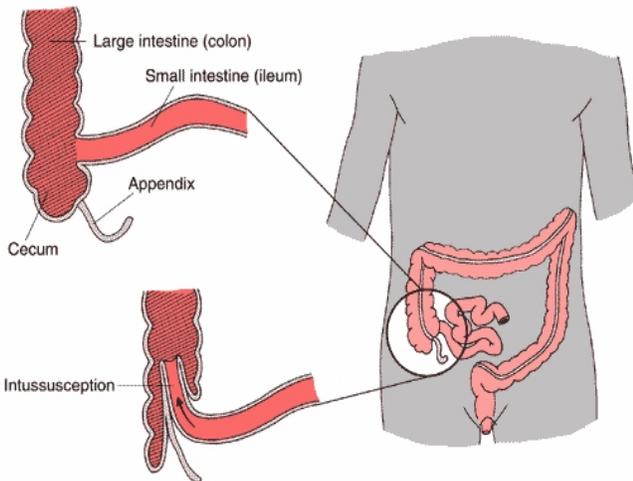
(a) Commencement of intussusception



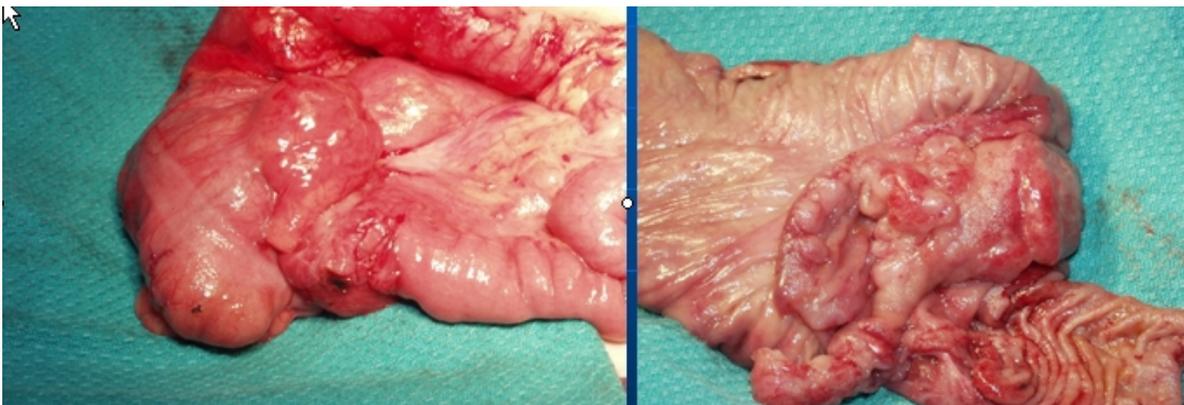
(b) Progression of intussusception



The most common site of occurrence is in the ileocecal region. Clinically, **PAIN** is the most frequent symptom/sign followed by **vomiting, lethargy** and finally, passage of a stool with mucus and blood admixed (“**currant jelly**”).

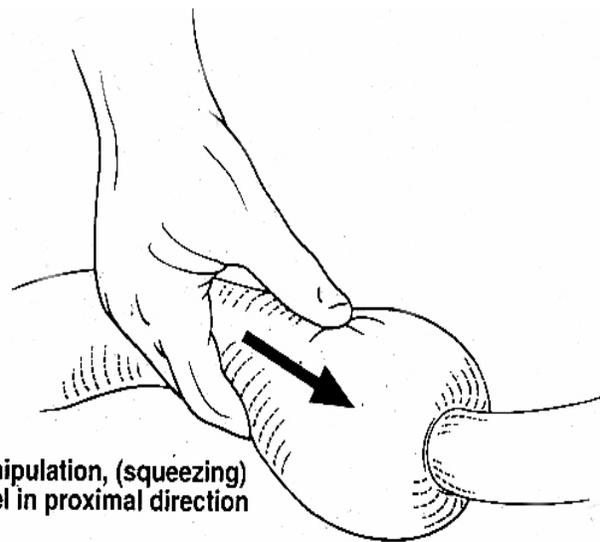


It occurs in otherwise healthy children (although it may be associated with Burkitt’s lymphoma, see images below) and seems to be triggered by an antecedent viral illness (i.e., in the idiopathic form).





Confirmation can be made with an ultrasound and a contrast enema and hydrostatic reduction with barium (or preferentially, **AIR**) can be therapeutic/curative. If hydrostatic reduction is unsuccessful, then manual reduction or resection at **surgery** is required – before ischaemic necrosis, infarction/perforation occurs. In general, pathologic lead points are seen in children younger than 4-6 months and older than 2 years. **Recurrence** (of the idiopathic form) may be as high as 10%.



Gentle manipulation, (squeezing) of the bowel in proximal direction

Avoid traction of ileum proximal to intussusception

NECROTIZING ENTEROCOLITIS (N.E.C.)

This is an acquired condition primarily affecting **premature** infants, a list of risk factors is listed in the table below. It is due to a combination of factors including poor blood flow to the bowel (e.g., the “diving reflex”), infection and exposure of the gastrointestinal tract to feeds, which may or may not be hyperosmolar. There is also an association with umbilical artery catheters (emboli).

Prematurity
 Respiratory distress syndrome
 Jaundice
 Umbilical catheters
 Congenital heart disease
 Asphyxia neonatorum
 Neonatal surgery for other congenital abnormalities
 Obstetric problems:
 caesarean section
 antipartum haemorrhage
 multiple births, e.g. twins
 forceps delivery
 prolonged rupture of membranes
 pre-eclampsia
 maternal hypertension

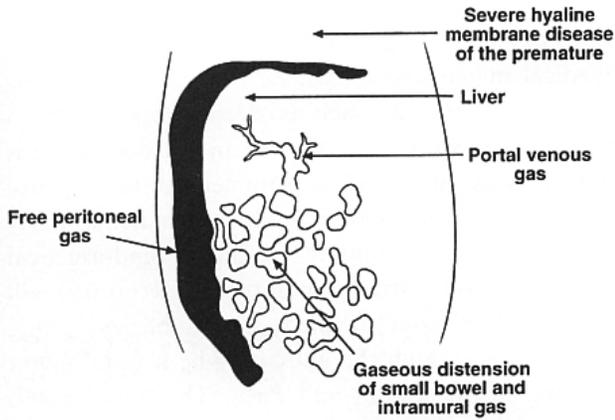
Clinical features of neonatal necrotizing enterocolitis

General features	Lethargy Refusal of feeds Apnoeic episodes Vomiting
Abdominal features	Abdominal distension Oedema and redness of abdominal wall Abdominal tenderness Rectal bleeding

The children usually present with generalized signs of sepsis along with intolerance to feeds, abdominal distention, varying degrees of tenderness associated with abdominal wall discoloration/cellulitis, and passage of blood per rectum.

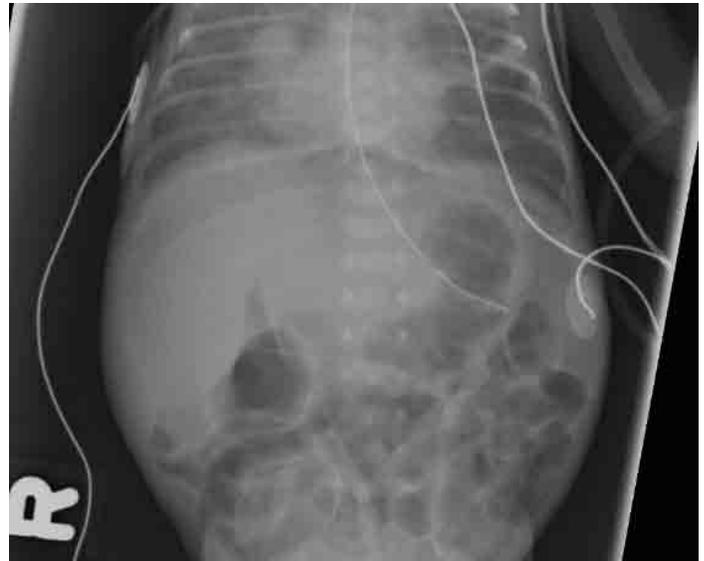


Abdominal x-rays characteristically show **pneumatosis intestinalis** (air within the wall of the bowel). Pneumoperitoneum (i.e., free air) indicating perforation of the intestine is considered an absolute indication for surgery.



(b)

Portal venous gas is indicative of severe disease: many babies develop full-thickness bowel necrosis and have extensive disease. Free intraperitoneal gas ('pneumoperitoneum') occurs when there is full-thickness bowel necrosis with perforation: (a) radiograph; (b) schematic view.



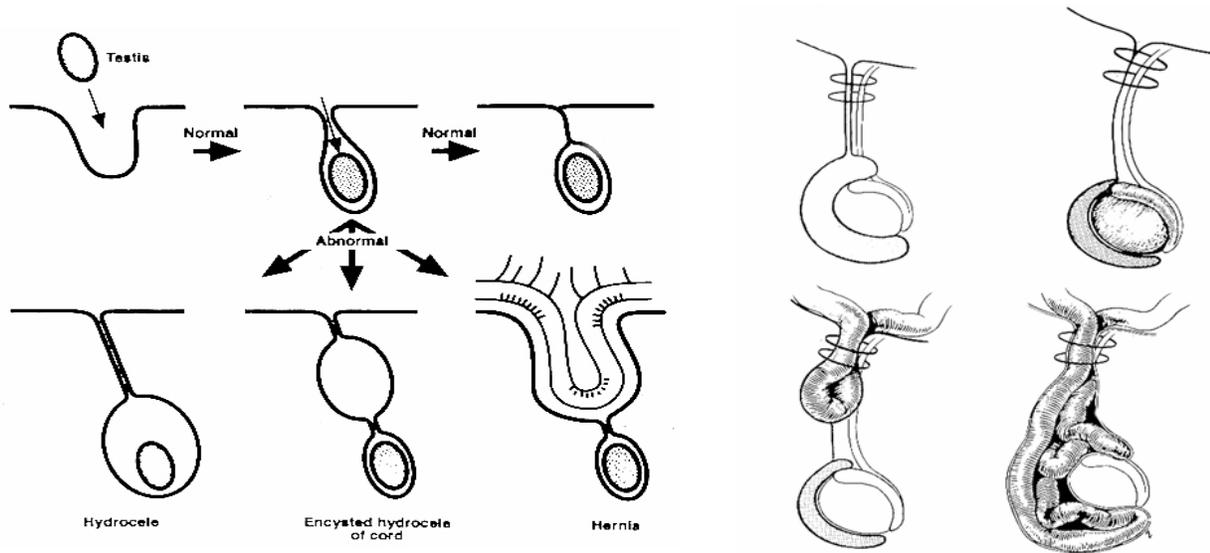
Radiological features of neonatal necrotizing enterocolitis

1. Intramural gas ('pneumatosis intestinalis').
2. Portal venous gas
3. Free gas ('pneumoperitoneum')

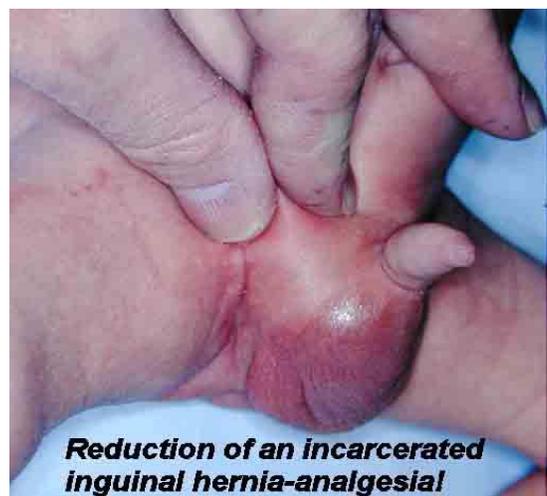
Treatment is generally nonoperative except for instances where there is clear cut or suspected perforation of the bowel. Mortality rates may be exceedingly high in these sometimes very premature and/or low birth weight infants.

HERNIAS, HYDROCOELES, AND UNDESCENDED TESTICLES

Descent of the testes from the retroperitoneum through the abdominal wall to the scrotum (at 30-32 weeks gestation) creates some of the conditions for the development of a hernia or hydrocoele. Failure of the **processus vaginalis** to obliterate preserves communication with the peritoneal cavity allowing either abdominal fluid (hydrocoele) or contents (i.e., bowel – hernia) to enter the groin and/or scrotum. Childhood hernias are **NOT** due to muscular defects or weaknesses (i.e., they are **INDIRECT HERNIAS**).



Hernias should be repaired at any age – as soon as the diagnosis is made, before **incarceration** can occur. Hydrocoeles can be left until one year of age – by which time a large portion will have resolved spontaneously. Hernias occur less frequently in girls (male: female ratio 10:1). The ovary may be palpable within the inguinal canal. Very rarely one may see the testicular feminization syndrome – phenotypic female, genotypic male. **Undescended testicles** are generally repaired around one year of age for the following reasons: cosmesis, psychologic, presence of an associated hernia, avoidance of trauma, spermatogenesis, production of testosterone and finally – earlier diagnosis of subsequent malignancy (3rd to 4th decade usually).



THE BILIARY TRACT

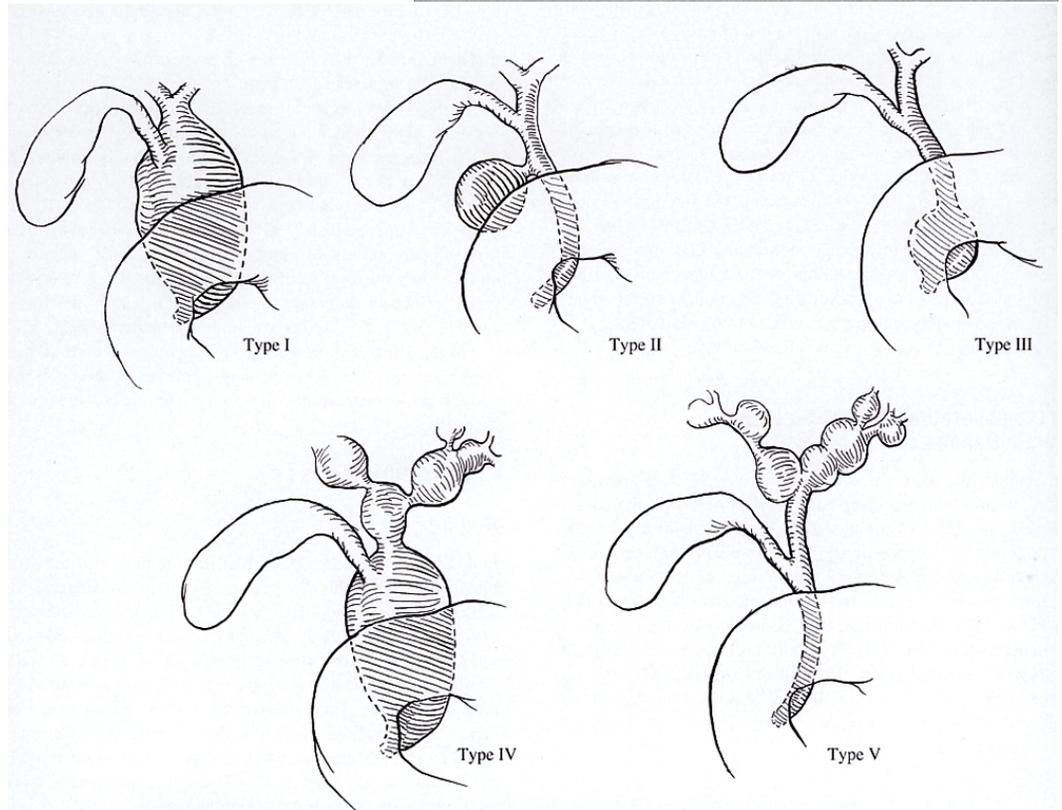
Biliary atresia and choledochal cysts are the two most common conditions affecting the extra-hepatic biliary tract in neonates. Both are idiopathic, resulting in varying degrees of jaundice but there the similarity ends.

Biliary atresia may be correctable with early surgery but frequently requires liver transplantation for end-stage liver disease.

Choledochal cyst, on the other hand (congenital dilation of the Biliary tract) is more easily corrected with surgery, with uniformly good, long-term results.

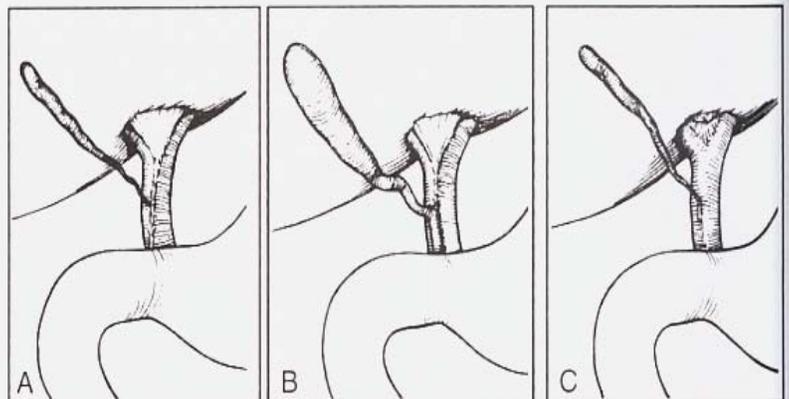
TABLE 103-1 Diagnosis of Biliary Atresia

- Physical examination with an emphasis on
 - Stool color
 - Liver consistency
- Liver function tests
- Ultrasonography
- Hepatobiliary scintigraphy (diisopropyliminodiacetic acid [DISIDA]) with phenobarbital preconditioning
- Percutaneous liver biopsy



The Todani modification of the Alonzo-Lej classification of cystic malformations of the biliary ductal system.

BILIARY ATRESIA The pattern of fibrous obliteration of the extrahepatic ducts varies at exploration. *A*, The most common pattern with fibrotic extrahepatic ducts in continuity. *B*, The gallbladder, cystic, and common ducts are patent with fibrous obliteration at the porta. *C*, Fibrous cord representing the gallbladder; the remaining biliary tree is scanty to absent. (From Altman RP, Lilly JR, Greenfield J, et al: A multivariable risk factor analysis of the portoenterostomy [Kasai] procedure for biliary atresia: Twenty-five years of experience from two centers. *Ann Surg* 1997;226:348, discussion 353. Courtesy of Lippincott Williams & Wilkins.)



MALIGNANCY

The most common malignancy in childhood is leukaemia. The most common solid tumor is a malignant brain tumor (astrocytoma, medullablastoma). Hodgkin's disease, non-Hodgkin's lymphoma, Wilms' tumor, neuroblastoma, hepatoblastoma, rhabdomyosarcoma and teratomas are the more common solid tumors occurring in the head and neck, mediastinum, abdomen, retroperitoneum, pelvis and extremities of children. Surgical removal of solid tumors is the mainstay of treatment. Radiation and chemotherapy are usually reserved for advanced or metastatic tumors and adjuvant therapy..

Distribution of Cancers in Children Under 15 years of Age

Leukemia	30%
CNS	19%
Lymphoma	13%
Neuroblastoma	8%
Soft Tissue Sarcoma	7%
Wilms' Tumor	6%
Bone	5%
Retinoblastoma	3%
Liver	1%

Improvement in Survival for Children with Cancer 1960-63 versus 1986-91

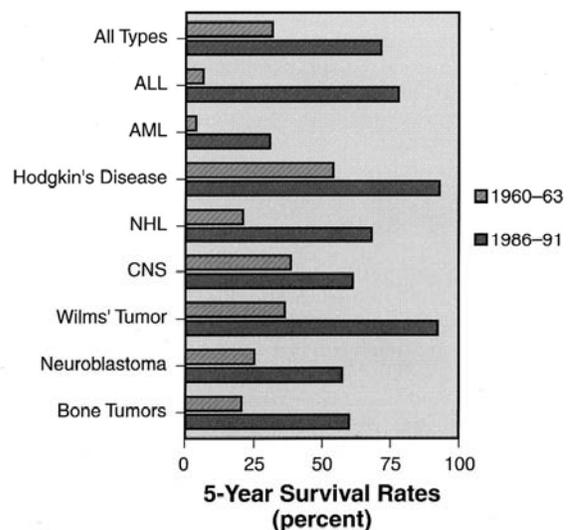


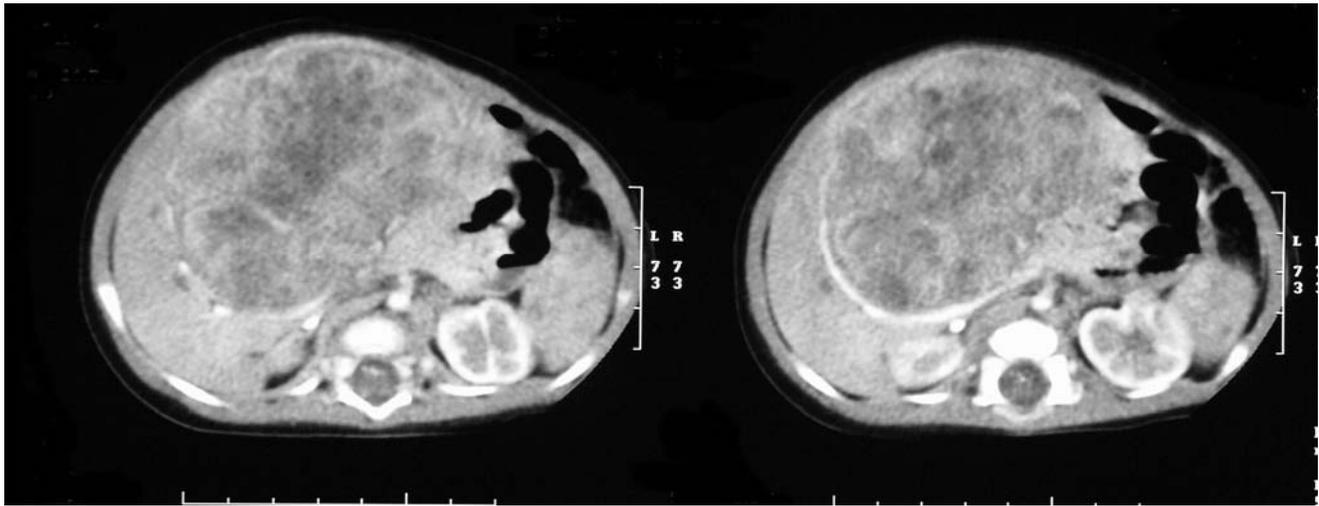
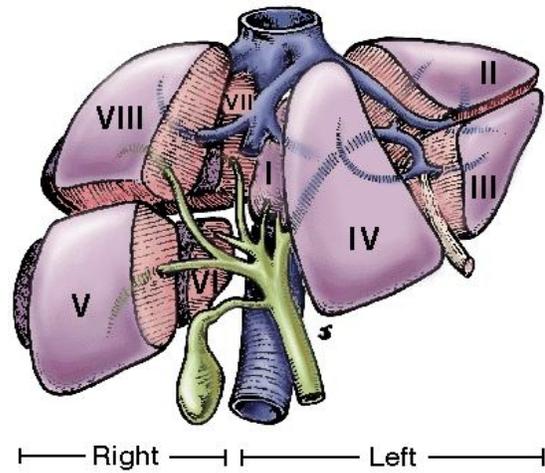
Figure 63-1. Comparison of patient survival during the period 1960-1963 versus 1986-1991 for the common childhood malignancies. (From Parker SL, Tong T, Bolden S, et al: Cancer statistics 1996. CA Cancer J Clin 65:5-27, 1996.)

Changing Patterns of Childhood Malignancies with Age

Malignancy	0-4 yrs	5-9 yrs	10-14 yrs
Leukemia	56%	22%	22%
CNS	42%	35%	23%
Neuroblastoma	87%	12%	1%
Wilms' Tumor	84%	15%	1%
Retinoblastoma	100%	0%	0%
Hepatoblastoma	80%	15%	5%
Non-Hodgkin's Lymphoma	17%	36%	47%
Hodgkin's Lymphoma	1%	30%	69%
Soft Tissue Sarcoma	51%	25%	24%
Bone Sarcoma	6%	24%	70%
Gonadal + Germ Cell	54%	11%	35%

Hepatoblastoma

Hepatoblastoma is an uncommon malignant liver neoplasm occurring in infants and children and composed of tissue resembling fetal or mature liver cells or bile ducts. Patients usually present with an abdominal mass, and are treated with surgical resection, adjuvant chemotherapy, or liver transplantation. A large liver mass is demonstrated in the CT scans below:



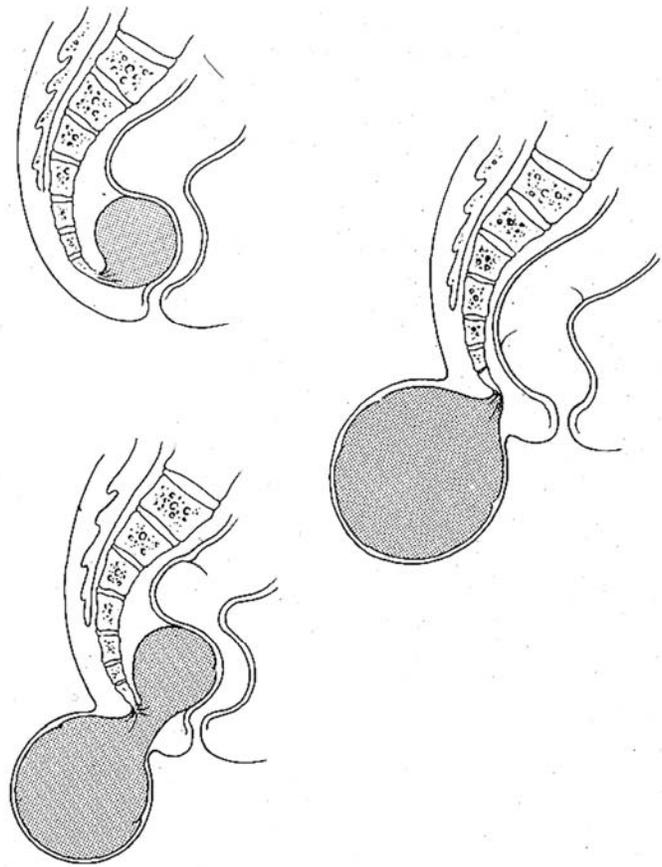
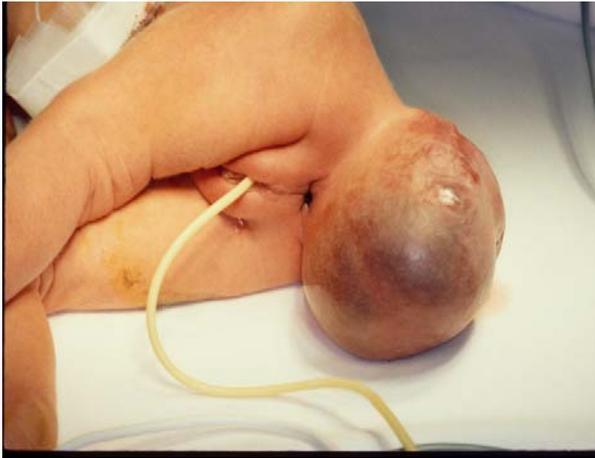
Neuroblastoma

Neuroblastoma is the most common cancer in infancy, and the most common extracranial solid cancer in childhood. It is a neuroendocrine tumor, arising from any neural crest element of the sympathetic nervous system, commonly in the abdomen, thorax, spinal cord, or intracranially.



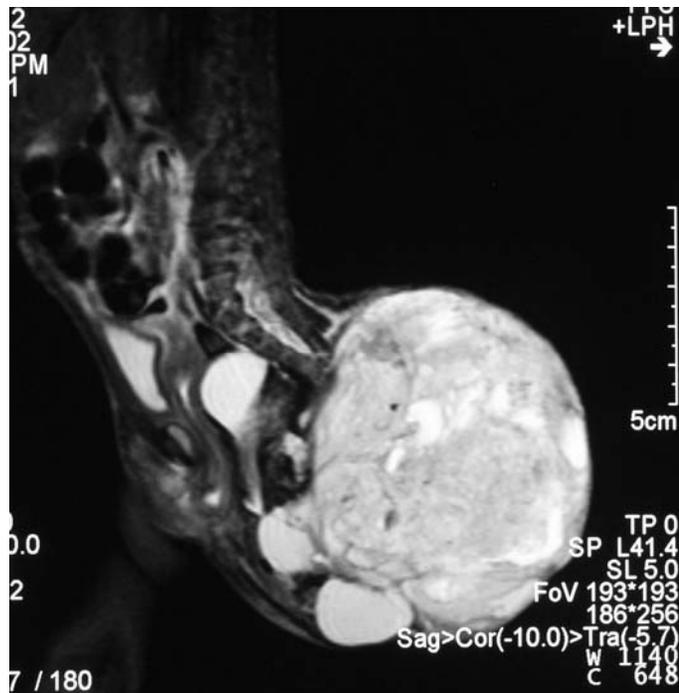
SacroCoccygeal Teratoma

SacroCoccygeal teratoma is a teratoma (neoplasm containing all three germ layers) located at the base of the coccyx, and thought to be a derivative of the primitive streak. It is the most common tumor presenting in newborns, and requires surgical resection.



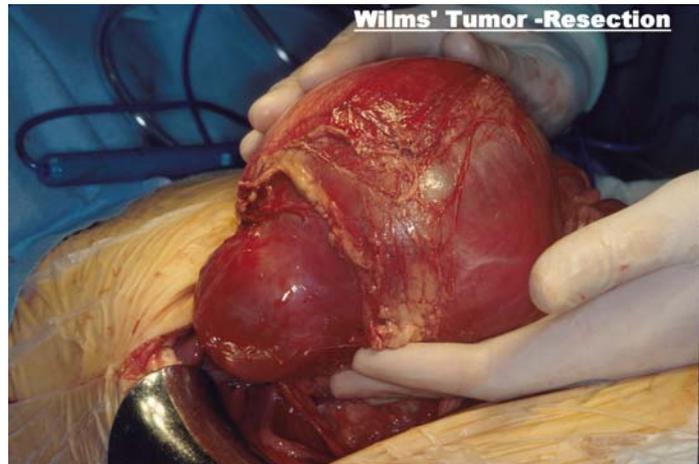
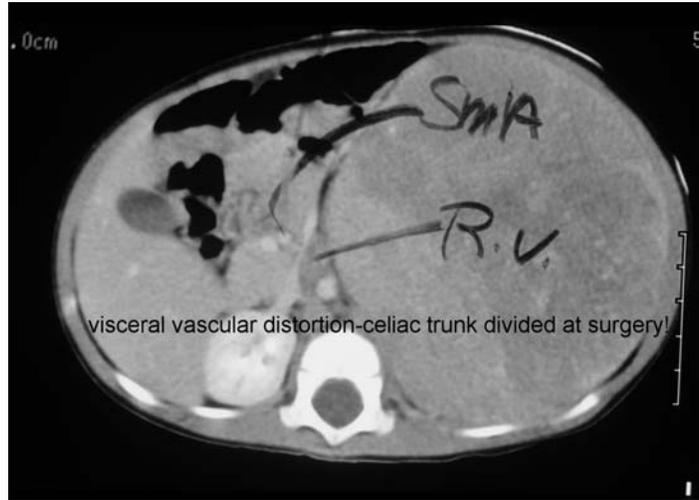
18-31. SacroCoccygeal teratomas may be chiefly anterior or posterior to the coccyx or they may be dumbbell-shaped.

MRI in the sagittal plane shows a SacroCoccygeal teratoma at the base of the spine:



Wilms' Tumor

Wilms' Tumor, also called nephroblastoma, is a tumor of the kidneys that typically occurs in children and may be associated with other developmental abnormalities in up to 25% of patients. Imaging and pathology are used to stage the tumor and determine the patient's treatment, which may involve a combination of nephrectomy, chemotherapy, and radiation,.



TRAUMA

In any disease state, **prevention** is the key to reduction in mortality and morbidity. Unfortunately, a major reduction in death and disability rates in childhood cannot be dealt with as in the adult population by eliminating one single, readily identifiable factor such as cigarette smoking. Epidemiological studies have documented repeatedly that **trauma** is the single largest killer in all childhood age groups except for the first year of life. Children who die in the first year of life from injury, do so as the result of child abuse.

The treatment of the severely injured child is often shared by the general practitioner, emergency physician, pediatrician, surgeon, and intensivist – and they need to work closely together to provide optimum care. To reduce morbidity and mortality rates in the critical early hours (“**golden period**”), improvements in transport and early resuscitation are necessary. To this end, the **ATLS** and **PALS** courses are invaluable. Injury awareness programs direct their energies to the lay population, ie: those individuals at risk.

Public health concerns to reduce morbidity and mortality are directed towards: 1) child “farm implement control”, 2) seatbelt use, 3) helmets, 4) avoidance of ATV (all terrain vehicle) use by children, and 5) stricter gun control. The initial evaluation and management of the injured child follows the ATLS guidelines – primary survey and resuscitation followed by secondary survey, definitive care phase and transport. The primary survey and resuscitation are done simultaneously: a patent airway is established while maintaining C-spine control. Adequate ventilation, treatment of shock, and identifying life-threatening injuries are the next priorities.

As in the adult trauma patient, every seriously injured child must be considered to have a **cervical spine injury** until proven otherwise, and even at that a spinal cord injury without radiological abnormality may occur (“**SCIWORA**”). Maintenance of a **patent airway** is obviously the most critical factor and once this is established one can continue with the primary survey and resuscitation.

The **infant’s trachea** is short, and the larynx is smaller with a more anterior and caudal angle. The epiglottis is relatively large and attached to the vocal cords at an acute angle. These things may make visualization for direct cannulation more difficult. The risk of bronchial intubation is far greater in the child. Apart from the very rare occasion where needle cricothyroidotomy may be required, “surgical establishment” of an airway (open cricothyroidotomy or tracheostomy) is virtually never required.

Due to the excellent physiological adaptation of children, **shock** may go unrecognized in its early stages. The child’s **blood volume** can be estimated at **80cc/kg**, and it is generally assumed that any child who is hypotensive secondary to hypovolemia has lost at least 25% of their blood volume. A “normal” systolic blood pressure can be estimated by adding 80 to two times the child’s age in years. A normal diastolic blood pressure is roughly 2/3 of the systolic pressure. Fluid resuscitation is generally done with Ringer’s Lactate using the “3-to-1” rule. A fluid bolus of 20cc/kg of **Ringer’s Lactate** is given over a short period of time. If

normovolemia is not restored with three bolus infusions of 20cc/kg then a blood transfusion is required (60cc/kg of crystalloid will roughly achieve a 20% blood volume replacement). A very limited amount of time (60 to 90 seconds) should be spent establishing a peripheral venous line in the hemodynamically unstable child. Attempts at percutaneous subclavian and internal jugular catheterization should **NOT** be attempted. **Intraosseous infusion** and **venous cutdown** (long saphenous, median antecubital, external jugular) provide rapid access to the circulation and are far safer when routine access on the back of the hand and at the ankle(saphenous) or wrist(cephalic) are not successful.

Adequate urinary output (1-2 cc/kg/hr) is important in the assessment of ongoing fluid replacement, but in the immediate timeframe of resuscitation, vital signs are more important.

Normal “adult” renal function is reached at approximately six years of age. The kidney of the young child has a limited ability to excrete a free water load and it may take longer for the obligatory post-resuscitation diuresis to occur. For this reason there may be a need for diuretic agents in the post-resuscitative phase (re: pulmonary and cerebral edema).

It is important that all intravenous fluids, including blood, be warmed because of the risk of **hypothermia**. Children have a **large body surface area** in relationship to their weight, relatively **thin skin**, and **lack of insulating fat**. These lead to an increased loss of water and heat, and appropriate measures must be undertaken to ensure they do not become hypothermic (overhead heaters and thermal blankets, raised room temperature, heated IV fluids and blood).

The role of the PASG (pneumatic antishock garments) is controversial. The amount of “autotransfusion” is probably insignificant. Primary effects are through increasing systemic vascular resistance and tamponade of external and internal bleeding sites and stabilization of lower extremity and pelvic fractures. Absolute contraindications to the use of the pneumatic antishock garments are pulmonary edema and known myocardial contusion or other cardiac injury.

The **secondary survey** will identify those potentially life threatening cardiopulmonary injuries that were not immediately evident in the primary survey.

The rib cage of the child, because of its inherent elasticity and less rigid nature, provides little protection to the solid organs in the upper abdomen or the heart and lungs within the thoracic cavity itself. Major compressive injuries of the chest wall may result in severe underlying injuries with no rib fractures and very little evidence of external trauma. Children, because of this inherent elasticity of the chest wall, tolerate flail chest and tension pneumothorax poorly. Myocardial and pulmonary contusion are relatively common, but injuries to the great vessels (i.e.: aorta) are quite rare except with penetrating injuries. **Physical examination** and x-ray are the physician’s primary means of diagnosing these injuries. The principles of treatment of specific entities such as tension pneumothorax, hemothorax, open pneumothorax, pericardial tamponade, and flail chest are the same as in the adult patient.

Specific patterns of injury should be looked for. There is a well-known association of lap-belt injuries and lumbar spine fracture with small bowel/mesentery injury. A very high index of suspicion must be maintained, as the bowel/mesentery injury may be very difficult to diagnose.

The Lap Belt Complex: Intestinal and Lumbar Spine Injury in Children

KURT D. NEWMAN, M.D., LEON M. BOWMAN, B.S., MARTIN R. EICHELBERGER, M.D., CATHERINE S. GOTSCHALL, M.A., GEORGE A. TAYLOR, M.D., DENNIS L. JOHNSON, M.D., AND MICHAEL THOMAS, M.D.

The "seatbelt syndrome" describes intestinal and spinal injury caused by lap-style automotive restraints. More than 2,600 children were admitted to Children's National Medical Center with blunt injury in 3 years; 395 were involved in a motor vehicle crash. Ninety-five of the crash occupants (24%) were known to be wearing safety belts.

Ten children sustained a "lap belt injury": five with lumbar spine injury, four with combined lumbar spine and intestinal injuries, and one child with intestinal injury. All ten children presented with a characteristic transverse abdominal ecchymosis.

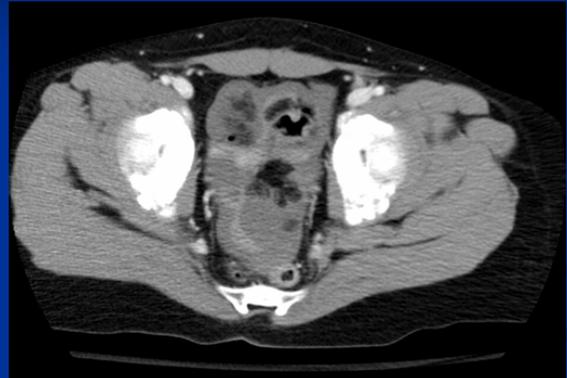
The CT scan was unreliable in evaluation of both spinal and intestinal injury. Lateral radiographs were required for definitive diagnosis in eight of nine children with lumbar spinal injury. CT scan was clearly diagnostic in only one of five children with intestinal injury.

Children wearing lap belts are at risk of a "lap belt complex." Lateral spine X-rays, peritoneal lavage, and early laparotomy are recommended to establish an accurate diagnosis and to decrease morbidity.



Examination of the abdomen during the secondary survey may reveal significant injuries, but it is important to appreciate that, as in the adult patient, a large number of injuries of a life-threatening nature may be missed! Blood, within the peritoneal cavity can be surprisingly nonirritating with virtually no peritoneal signs, and conversely, the frightened child with a simple abdominal wall contusion may have physical signs suggesting frank peritonitis. Virtually every child with significant torso trauma will have a **gastric dilatation**, and a nasogastric tube of adequate size should be placed.

CT findings: free intraperitoneal fluid, no free air



Frequent **re-examination** is extremely important and the examination can be supplemented with diagnostic peritoneal lavage, ultrasound, CT scan, intravenous pyelography, and radionucleotide scans (where available). CT scan is invaluable in the assessment of abdominal/truncal trauma. Identification of solid organ injuries is usually straightforward. Demonstration of fluid (i.e. blood) without solid organ injury, usually means a significant mesenteric or small bowel injury. The role of FAST (Focused Abdominal Ultrasound in Trauma) is controversial and of limited usefulness. It is advisable that DPL be done by the surgeon who will be reassessing the child and providing definitive care – surgery, should this become necessary. Laparoscopy should

NOT be considered as a useful diagnostic test. Obviously, hypovolemia with abdominal distention requires no further investigations except perhaps a chest x-ray before immediate laparotomy. Valuable time should not be wasted in diagnostic maneuvers that are not necessary and may delay the child's further management and adversely affect subsequent survival.

The intra-abdominal position of the bladder in the younger child and the fact that the bladder is often full (particularly when riding in a car) means that the urinary bladder is more prone to injury and traumatic rupture (either intra or extra peritoneal) should be kept in mind with or without concomitant pelvic injuries.

The usual signs of urethral injuries should alert the physician to the need for cystourethrography. Suspected renal injuries are evaluated in the usual way (IVP, ultrasonography, contrast enhanced CT scan).

The **neurological examination** concentrates on evaluating the level on consciousness, pupillary size, shape and reactivity, movement of the extremities, plantar responses, and evidence of a basilar skull fracture – CSF leak from the nose or ear, Battle’s sign, and blood behind the tympanic membrane. The **Glasgow Coma Scale** quantitates the patient’s **eye opening, best motor response, and best verbal response** (Appendix III). The verbal response in a child under the age of two is altered so that a full score is given if the child cries appropriately. To prevent secondary damage, **avoidance of hypotension, hypoxemia, and hypercarbia** are critical in the management of the child with increased intracranial pressure. Diuretics such as Mannitol and Lasix may be used while awaiting definitive care.

Spinal cord injuries are relatively uncommon, but do occur, and one has to be aware of the entity of “**SCIWORA**” – **Spinal Cord Injury Without Radiological Abnormality**. Up to 2/3 of children with spinal cord injury have no demonstrable x-ray abnormality, but will usually have signs or symptoms pointing to the spinal cord (paresthesiae, numbness, paresis). X-rays of the child’s C-spine can be very difficult and neurosurgical consultation and assessment along with CT scan and MRI all may be required to complete the evaluation.

Evaluation of **extremity trauma** concentrates on skeletal and soft tissue injuries. In 25% of children admitted to major trauma centers there is at least one missed injury on the initial evaluation. The majority of these are orthopedic injuries and although the risk to life is negligible, the consequences for the child may be severe (nerve damage, deformity, limb length discrepancy). The “head-to-toe” evaluation in the **secondary survey** should identify virtually all areas of concern. These can then be further evaluated with x-rays. Soft tissue injuries require debridement, irrigation, antibiotic coverage, and the usual precautions regarding tetanus.

The ABCs of trauma in children are identical to those in the adult:

- A) Airway with c-spine control
- B) Breathing
- C) Circulation, etc., etc.

Fluids and blood requirements are different and the pediatric airway presents specific problems. Avoidance of **hypothermia** is critical and specific injury patterns including **SCIWORA** (spinal cord injury without radiographic abnormality) and the **lap belt** complex must be remembered. The possibility of child abuse (N.A.I. – nonaccidental injury) should be kept in mind particularly with unusual injury patterns including repeated trauma.

SUMMARY

Basic principles of the care and treatment of the injured child do not differ from those of the adult. Anatomical and physiological differences require some alteration in one's priorities and strategy for treatment. Greater technical expertise may be required with such things as intubation and establishment of vascular access.

APPENDIX II

Perioperative Pediatric Care

Fluids & Electrolytes

	NS	RL	2/3 1/3	D512
• 4 ml/kg/hr for 0-10 kg (100 ml/kg/24 hr)	Na	154	130	52
• 2 ml/kg/hr for 10-20 kg (50 ml/kg/24 hr)	K		4	
• 1 ml/kg/hr for > 20 kg (20 ml/kg/24 hr)	Ca		109	52
• Na+ 2-5 meq/kg/24 hr	Mg			
• K+ 1-3 meq/kg/24 hr	Cl	154		
• 2/3 – 1/3 is fluid of choice for maintenance	Lactate			
• add 50 ml D50W to each 500 ml of	Dextrose			33.3 g/l
2/3 – 1/3 for age < 3 mo	HCO ₃		28	5 g/l
	Osmolality	208	280	

Blood Transfusion

- Blood volume
 - 85 ml/kg for infants
 - 80 ml/kg for toddlers and young children
 - 75 ml/kg for older children
- Transfusion
 - 10 ml/kg packed RBC ~ 1 unit in adults
 - >30 ml transfusion requires clotting factors also
- Shock
 - bolus 20ml/kg (NS or RL)

Bowel Preparation

- Golytely – 100 cc/kg @ 25-35 cc/kg/hr PO/NG

Analgesia

- Acetaminaphen
 - 15 mg/kg/dose q4 – 6 hr PO
 - 20 mg/kg/dose q4 – 6 hr PR suppository
- Codeine
 - 0.5 – 1 mg/kg/dose q4 – 6 hr IM/PO (4 mg /kg/24 hr max in divided doses)
- Meperidine (Demerol)
 - 1 – 2 mg/kg/dose q3 – 4 hr IM

Perioperative Pediatric Care (continued)

Analgesia (continued)

Morphine

- – 0.2 mg/kg/dose q2 – 4 hr IM
- – 0.05 mg/kg/hr IV maintenance (i.e., 1.25 mg/kg in 500 ml D5W at 5 – 10 ml/hr)

Naloxone (Narcan)

- mg/kg/dose IV or IM
- for narcotic overdose

Diazepam (Valium)

- 0.05 mg/kg IV or PO

Lidocaine

- 3 – 4 mg/kg max. without epinephrine
- 7 mg/kg max. with epinephrine

Antibiotics

Cefazolin (Ancef)

- 30 mg/kg q8h

Metronidazole (Flagyl)

- 10 mg/kg q8h

Penicillin G

- 100,000 IU/kg/24 hr q6h IV for moderate infections
- 250,000 IU/kg/24 hr q2 – 6 hr IV for severe infections

Penicillin V

- 25 – 50 mg/kg/24 hr q6 – 8 h IV

Cloxacillin

- 50 – 100 mg/kg/24 hr q6h IV or PO for moderate infections
- 200 mg/kg/24 hr q6h IV for severe infections

Amoxicillin

- 25 – 50 mg/kg/24 hr q8h PO

Ampicillin

- 100-200 mg/kg/24 hr q6h IV

Gentamicin

- 7.5 mg/kg/24 hr q8h IV (measure serum levels pre and post third dose)

Sepra (Trimethoprim-Sulfamethoxazole)

- 8 mg/kg/24 hr TMP and 40 mg/kg/ 24 hr SMX q12h (i.e., 0.5 ml/kg bid of suspension)

APPENDIX III

Glasgow Coma Scale - Pediatrics

Best eye response: (E-Score)

4. Eyes opening spontaneously
3. Eye opening to speech
2. Eye opening to pain
1. No eye opening

Best verbal response: (V-Score)

5. Infant coos or babbles (normal activity), appropriate words, social smile
4. Infant is irritable and cries, but consolable
3. Infant is persistently irritable
2. Infant is restless and agitated
1. No verbal response

Best motor responses: (M-Score)

6. Infant moves spontaneously or purposefully
5. Infant withdraws from touch
4. Infant withdraws from pain
3. Abnormal flexion to pain for an infant (decorticate response)
2. Extension to pain (decerebrate response)
1. No motor response

10 key points you need to remember about Pediatric Surgery:

1. **Green vomit** is a surgical emergency until proven otherwise.
2. **Midgut volvulus** = dead or dying bowel
3. **Polyhydramnios** demands exclusion of **esophageal atresia** (by passage of a radiopaque NG tube).
4. **Abdominal wall defects:** ***Gastroschisis** = “good”
***Omphalocele** = “awful”
5. **Inguinal herniae** are due to a congenital processus vaginalis (ie: peritoneum), **NOT** muscular defects of the abdominal wall.
6. **Constipation** (or failure to pass meconium) may be the only clue to the diagnosis of **Hirschsprung’s** disease.
7. Scaphoid abdomen + respiratory distress (in the newborn) = **diaphragmatic hernia**.
8. **Intussusception** – colicky abdominal pain, vomiting ± blood/mucous per rectum.
9. **Hypertrophic pyloric stenosis** is acquired (not congenital) and characterized by projectile, non-bilious vomiting ± the “classic” electrolyte disturbance (hypokalemic, hypochloremic metabolic alkalosis).
10. **VACTERR** association of anomalies: **V**ertebral, **A**nus, **C**ardiac, **T**racheo-**E**sophageal, **R**enal, **R**adial **R**ay.