Pediatric surgery

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Intestinal obstruction

- Incidence: this depends on the site of obstruction; however, it generally ranges between 1:300 to 1:1000 live births
- Types:
    - Atresia: complete obliteration of the lumen of any tube. It is usually congenital; however, it can be acquired. It can happen at any level of the GI tract.
    - Stenosis: narrowing of the lumen. It is common in the duodenum; however, it is rare in other parts of the GI tract.
    - Meconium ileum
    - Meconium plug
    - Hirschprung's disease
    - Volvulus neonatorum: results from malrotation of the bowel. In the case of volvulus neonatorum, the bowels are not fixed properly to the posterior abdominal wall.
- Pathophysiology:
    - Failure of canalization: During development, the intestines develop as a cord. However, during later stages of development, vaculations develop and join to form a lumen. Failure of this process results in atersia. This theory best applies to the duodenum.
    - Vascular accidents: Loss of blood supply to any part of the intestines leads to necorsis of that specific part. The necrosed part becomes atretic. The blood supply can be compromised due to volvulus, intussusception, thrombus, or an embolus in the blood vessel. This theory can be applied to any part of the GI tract.
    - Failure of separation: During development, the esophagus and trachea develop as one tube. Later during their development, a septum separates them. This applies to the rectum and urogenital sinus. Failure of separation will lead to atresia and fistula formation.
- Classification:
    - Simple obstruction: no complications
    - Strangulated obstruction: complicated by ischemia and necrosis
    - Closed loop obstruction: It occurs when both ends of a certain intestinal segment are obstructed. This obstruction leads to a rapid increase in pressure, which ends in rupture of the segment in question. This happens in cases of volvulus and competent ileo-cecal valve.
- Manifestations of obstruction:
    - Esophageal atresia:
        - Upper esophagus:
            - Drooling of saliva
            - Respiratory distress

        - Choking with saliva and food
        - Failure to pass NG tube (diagnostic)
    - Lower esophagus:
        - Accumulation of secretions leads to regurgitation and vomiting.
        - If the pathological insult continues, accumulation of fluids will increase the pressure on the esophageal wall. This will leads to obstruction of the arterial circulation. This, in turn, leads to ischemia and physiological death of the esophageal mucosa. Which will lead to a ruptured esophagus.
  - Other parts of the GI tract: The more distal the obstruction, the worse the distention.
    - Abdominal distention
    - Vomiting
    - Failure to pass meconium
    - Jaundice (if the obstruction involves the hepatic circulation)
- Associated congenital anomalies:
  - Polyhydramnios:
    - Increased amount of amniotic fluid.
    - During fetal development, the amniotic fluid circulates. The fetus drinks the fluid, and passes it in the urine. However, when there is an obstruction, this process will be interrupted leading to a pathological accumulation.
    - The higher the site of the obstruction, the more the accumulated fluid.
    - Esophageal atresia, in some of its forms, is not accompanied with polyhydramnios. This is due to the presence of a tracheo-esophageal fistula.
  - Low birth weight
  - Trisomy 22
  - VATER syndrome: vertebral, ano-rectal, tracheo-esophageal fistula, and renal anomalies
  - VACTERL syndrome: vertebral, ano-rectal, cardiovascular, tracheo-esophageal fistula, renal or radial anomalies, and limb anomalies.
  - Down syndrome
- Investigations:
  - In utero diagnosis: Can be done as early as the 16^{th} week gestation.
    - Amniocentesis
    - Biochemical testing
    - Genetic testing
    - Ultrasound
  - Clinical diagnosis: when symptoms of obstruction are obvious
  - Failure of passage:

        - NG tube: diagnostic of esophageal atresia
        - Rectal thermometer: diagnostic of ano-rectal atresia
    - Plain X-ray:
        - Stepladder sign: appearance of multiple air-fluid levels when the patient is in a sitting position
        - Distention of the lumen
        - Double bubble sign: signifies duodenal stenosis or atresia
    - Contrast studies (rarely used):
        - Barium meal
        - Barium enema
    - CT scan (rare)
    - Ultrasound (rare)
- Staging the disease:
    - Early stage: vomiting and distention
    - Perforation: less vomiting and distention; signs of systemic toxicity
    - The presence of dehydration signifies prolonged vomiting
- Differential diagnosis:
    - Mechanical obstruction
    - Non-mechanical obstruction: Hirschsprung's disease
    - Sepsis:
        - Sepsis can lead to obstruction
        - Obstruction can lead to sepsis
        - Differentiation is based upon history
    - Hypothyroidism: presents with a picture of obstruction without the presence of a true obstruction.
    - CNS pathology:
        - Intra-ventricular hemorrhage: leads to reflex ileus
        - Cerebral palsy
        - Hydrocephalus
        - Raised intracranial pressure
    - Narcotic addiction of the mother: evident two to three days after birth
- Management:
    - Fluid resuscitation: water and electrolytes
    - GI decompression: suction of the fluids will decrease the pressure on the more distal parts of the GI tract
    - Parenteral nutrition
    - Antibiotics: given to all children before the development of sepsis.
    - Surgical correction of the underlying problem
    - NICU admission
- Factors that affect survival:

    - Birth weight
    - Age: premature babies are at a higher risk for death
    - Associated anomalies
    - Early detection and treatment
    - Appropriate surgical procedure
    - Preoperative and postoperative NICU care
- Types of atresia:
    - Intestinal:
        - Stenosis: the lumen is narrowed, but not occluded. The mesentery is normal
        - Type 1:
            - Lumen occluded by a membrane
            - Intact mesentery
            - Continuous posterior abdominal wall
        - Type 2:
            - Complete obstruction
            - Gap of fibrotic segment (cord)
            - Intact mesentery
        - Type 3a:
            - Long gap without a fibrotic cord
            - Mesenteric defect
            - The length of the missing part cannot be estimated
        - Type 3b: (apple peal or Christmas tree deformity)
            - Atresia of the whole intestines
            - The intestines form a loop around the superior mesenteric artery
        - Type 4:
            - Sausage shaped intestines
            - Multiple atresias
    - Esophageal atresia:
        - Type 1:
            - The most common
            - Proximal atresia
            - Distal tracheo-esopheal fistula
            - Presents as drooling of saliva
            - In utero, the amniotic fluid passes through the trachea to the esophagus. No polyhydramnios
        - Type 2:
            - Proximal atresia
            - Distal atresia

        - No amniotic fluid will pass to the stomach resulting in microgastria and a gasless abdomen
    - Type 3:
        - Fistula without atresia (H-shaped)
        - Not discovered at birth
        - Discovered due to repeated episodes of choking
    - Type 4:
        - Proximal fistula
        - Distal atresia
    - Type 5:
        - Proximal fistula
        - Distal fistula

- History of a suspected case of intestinal obstruction:
    - Bilious vomiting:
        - Vomiting esophageal or stomach contents yields a colorless vomitus.
        - Bilious vomiting is greenish-yellowish in appearance
        - It originates from the duodenum or upper jejunum
        - Intestinal obstruction is the most common cause of bilious vomiting
        - Other causes include sepsis and increased intracranial pressure
    - Abdominal pain: Although present, abdominal pain is not a useful sign as neonates cannot express themselves.
    - Abdominal distention: Children below the age of two have a normally distended abdomen. This means that this is not a very useful sign
    - Delayed or scanty passage of meconium:
        - Meconium is the equivalent of feces in an older child or an adult. It is the greenish substance passed within the first 48 hours of life.
        - Delay in the passage of meconium for more than two days is always abnormal
    - Polyhydramnios:
        - It occurs as a result of interruption of the normal circulation of the amniotic fluid
        - In intestinal obstruction, neonates are not able to circulate this fluid properly. The higher the level of the obstruction, the more pronounced the symptoms.
    - Down's syndrome patients have a higher incidence of intestinal obstruction
    - Drug history:
        - Mother's drug history is important due to the contact between the mother's milk and her child during lactation

    - Family history: it is important to rule out certain hereditary disease
        - Hirschprung's disease
        - Jejunal atresia
        - Maternal diabetes: children born to diabetic mothers have a higher tendency for developing functional intestinal obstruction.
- Investigations: keep in mind the aforementioned differential diagnoses. Rule them out using proper investigations. Moreover, perform the following tests to rule in or rule out intestinal obstruction:
    - Plain film (plain reontgenogram):
        - A plain abdominal X-ray depends on the gas in the GI tract as a contrast medium
        - Gas is normally found in the abdomen and the colon. Absence of gas in these areas indicates an abnormality
        - Gas is absent in the small intestines; the presence of gas in this area indicates an abnormality.
        - Gas in the GI tract is mainly swallowed; only a small percentage is formed by gas forming bacteria.
        - In cases of obstruction, gas is found in the segments proximal to the site of the lesion. In fact, the intestines are dilated proximal to the lesion and narrowed distal to the site of lesion
        - Obstruction around the jejunoileal junction will produce a characteristic x-ray appearance called "the accordion sign". This appearance is due to the presemce of valvuli coneventi (pica circularis).
        - Obstruction around the ileocecal valve will not produce a characteristic appearance. However, you will dilated segments proximally and narrowed segments distally.
        - Calcifications can be seen; however, they are not specific for intestinal obstruction
    - Contrast enema:
        - Administration of barium can exacerbate a case of intestinal obstruction, so it is not given in cases of intestinal obstruction
        - Administration of barium is contraindicated in cases of intestinal perforation as it can cause systemic toxicity if it reaches the peritoneal cavity
        - Barium enema is helpful as a diagnostic and therapeutic tool of meconium plug in Hirschsprung's disease. Administration of a barium enema will locate the site of the plug. In addition, it will dilate the distal segments of the bowel relieving the obstruction in the proximal parts of the colon.
        - During development, the bowel receives its innervation from the neural crest in a proximal to distal fashion. In Hirschsprung's disease ganglionic

cells are absent in the distal part of the GI tract. The affected segment will lose its peristaltic ability. In Hirschsprung's disease the rectum is always affected. Between the normal ganglionic segments and the abnormal aganglionic segment a transitional area exists. The part proximal to the transitional zone is dilated due to fecal accumulation. This gives the intestines a characteristic funnel-shaped appearance. Another important sign for the diagnosis of Hirschsprung's disease is the delay of passage of the contrast substance. This is proven by taken a second radiograph 24 hours after the administration of the enema.

        - Gastrographin is a hyperosmolar solution that may cause dehydration.
    - Upper GI series (contrast meal):
        - The most important study to exclude malrotation of the bowel.
        - In the case of malrotation, the bowel is floating in the peritoneal cavity. This will lead to a volvulus. Volvulus, in turn, will lead to necrosis of the affected segment. Which, if not treated, will lead to death.
        - In cases of incomplete obstruction, the bowel is partially attached to the posterior abdominal wall via the superior mesenteric artery. This abnormal attachment increases the risk of volvulus and its complications.
        - Volvulus is the most serious cause of intestinal obstruction
        - Normally, the duodeno-jejunal junction is to the left of the spine. In cases of malrotation, an upper GI series will show this junction to the right of the spine. Moreover, it may show a screw-like appearance of the junction.
    - Rectal biopsy:
        - Since Hirscsprung's disease always involves the rectum, a rectal biopsy that shows ganglions excludes the presence of this disease. A rectal biopsy that shows agangliosis is pathognomonic of Hirschsprung's disease.
- The most common cause of intestinal obstruction in children (not neonates) is intussusception.

Pediatric urological anomalies

- Hydronephrosis:
    - Dilation of the pelvi-calyceal system of the kidneys.
    - If this dilation includes the ureters, it is termed hydroyreteronephrosis.
    - An obstruction is defined as an obstacle, whatever its source is, that impeded urine outflow. This will lead to failure of growth of the kidney or damage to the kidney.
    - Although obstruction is the most common cause of hydronephrosis, these two terms are not synonymous.
    - In the case of hydronephrosis without obstruction, the kidney can continue to grow normally. On the other hand, in the presence of an obstruction, the high pressure in the ureter and the pelvi-calyceal systems will exert a back pressure on the kidney, which impedes its growth. In rare cases, damage to the kidney might ensue leading to renal failure.
    - Most cases of hydronephrosis can be detected antenatally using ultrasound.
    - In certain conditions, hydronephrosis resolves spontaneously. These include:
        - Overhydration of the fetus
        - Presence of another abnormality that resolved during embryogenesis
    - It is important to detect obstructive hydronephrosis early, because surgical treatment is recommended as early as possible.
    - Etiology:
        - Pelviureteric junction obstruction: the most common cause of hydronephrosis
        - Vesicoureteric junction obstruction
        - Posterior urethral valve
        - Vesicouretral reflux: the most common cause of renal failure in children
    - Diagnosis:
        - Ultrasound
        - Voiding cysto-urethrogram:
            - This test estimates the capacity of the bladder in addition to the smoothness of the mucosa. Moreover, this test can be used to investigate the urethra for the presence of bladder outlet obstruction.
            - A contrast material is introduced through a urethral catheter to fill the bladder. A picture is taken; if the ureters appear, this confirms the presence of a reflux. After taking the picture, the catheter is removed. The child is asked to urinate; and, a picture is taking

during urination. Any reflux that was not apparent on initial imaging will appear during voiding.

- Diuretic renogram:
  - The most commonly used materials for this test are DTPA and MAG3
  - It is more accurate than IVU in which we use a contrast material that is given iintravenously; this IV material is filtered y the renal system, then is seen on an X-ray. Around 100 photos are taken, then a computer will analyze and plot them in a curve
  - A normal curve would show the material being taken up by the kidneys (within 10 minutes), then the tracer will be washed out with our without the use of diuretics. The whole test takes 20 minutes.
  - If the patient has an obstruction, then the curve will show the uptake phase. However, the tracer will not be washed out. This will show as a plateau on the curve. The patient is given a diuretic. If there is no excretion within 15-20 minutes, this indicates a definitive obstruction.
  - Sometimes, the study can take more than 20 minutes, but the curve descends down in a slower fashion. In these cases, the patients have no definitive obstruction. These patients are followed actively with ultrasounds taken every 6 months. If the patient starts to have symptoms, or the ultrasound shows a deterioration, then you should interfere surgically.
  - In dilated, but not obstructed syste,s, the urine will start to wash out once you give the diuretic.
  - As a general rule, always start with the least invasive method of investigation. This is why an ultrasound is the first investigation in cases of suspected hydronephrosis in infants. In older children, you may use IVU; however, this is not necessary in an infant.
- Retrograde uropathy:
  - Invasive
  - Rarely done

- Treatment:
  - If symptomatic: surgery (pyeloplasty)
    - Excise the dilated area below and reimpant the urter in the normal posistion. It is one of the most successful operation with a success rate reaching up to 95%
  - If asymptomatic (with no PUJ obstruction): conservative.

                - A patient who has no symptoms and is detected antenatally or accidentally is observed to assess the kidney function through a diuretic renogram. The ideal test should show an equal uptake of tracer by both kidneys. A difference of more than 15% in the uptake between the two kidneys is considered a normal variation; however, if the difference is more than 15%, these patients should be treated surgically.
            - A PUV obstruction is worse than a VU obstruction because in the case of a VU obstruction the pressure will be distributed to the urters before reaching the kidney. This will spare the kidneys from any damage.
- Mega ureter: a dilated ureter. Sometimes used interchangeably with hydronephrosis.
    - Any ureter with a diameter > 7mm on the IVU or ultrasound.
    - Types:
        - Obstructive
        - Refluxing
        - Obstructive and refluxing
        - Non-classified
    - Management depends on the cause:
        - If reflux: medical management. Give prophylactic antibiotics. Reflux in the presence of an infection will affect the kidney function. Surgery is used in 10-15% of these cases.
        - Posterior urethral valve obstruction:
            - Most common cause of bladder obstruction
            - Incidence: 1:5000-800 male patients.
            - The obstruction is in the prostatic urethera due to the presence of urethral folds. If the patient presented during the neonatal period, he will be septic, azotemic, with renal failure and electrolyte imbalances.
            - The treatment is to stabilize the condition by correcting the acid-base balance or by removing the valve to remove the obstruction.

Abdominal wall defects

- Umbilical disorders:
    - Congenital
        - Drainage:
            - Vitello-inetestinal duct (patent as a fistula between the ileum and the umbilicus), cyst, sinus, or a fibrous band connecting the bowel to the umbilicus.
            - Allantois-Urachus: patent as a urinary fistula or a cyst
            - Umbilical arteries mass if infected
        - Masses
        - Hernias
    - Acquired:
        - Omphalitis
        - Polonidal sinus
- Umbilical masses:
    - Types:
        - Granuloma
        - Polyps
        - Cysts, urachus or vitello-intestinal duct
        - Dermoid
        - Hemangioma
        - Ectopic tissue, bowel mucosa, liver, etc…
    - treatment:
        - control local and systemic sepsis
        - sinuses are more common than fistulas
        - all sinuses and fistulas can be excised through an infraumbilical incision preserving the umbilicus
        - fistulas require excision of the involved bowel segment
        - urachus excision requires excision of a bladder segment
- umbilical hernia:
    - very common
    - cause: incomplete development of the umbilical ring leading to a failure in its closure. The umbilical cord emerges from this ring and contains 2 umbilical arteries, 1 umbilical vein, Wharthon's jelly, and the urachus.
    - The umbilical ring is reinforced by the round ligament, uracus, lateral umilical ligaments, and an extension of transverse fascia.
    - More common in blacks
    - Familial tendency

    - Associated anomalies include thyroid disease (hypothyroidism), trisomy 18, 13, 21, Beckiwth-Wiedmann syndrome, and Hurler's syndrome.
    - Many close spontaneously, especially ones less than 2 cm.
    - Many remain patent and require surgical repair. The procedure is performed after the age of 2, but before the age of 10.
    - Complications such as incareration and stangulation are rare because they run in a vertical non-oblique course. Moreover, the defect is large when compared to other hernias.
    - These defects are usually repaired because there is no benefit to strapping
- Supraumbilical hernia:
    - Includes paraumbilical and epigastric hernias. These are all defects in the midline fusion of the rectus muscle rather than in the linea alba.
    - Because the defect is in the covering fascia and not in the abdominal wall, the contect of this hernia does not contain bowel. It contains pre-peritoneal fat.
    - Supraumbilical hernias cause discomfort upon movement, so they are treated surgically.
- Urachus:
    - It is aremnant connecting the umbilicus and the urinary bladder. It could present as a fistula especially in those collage diseases or muscular diseases. Patients with Prune-belly syndrome have a patent urachus
    - A blind sinus is another pathology where the urachus is only partially closed
    - A cyst is formed when 2 sides of urachus close and the mid portion remains patent as a cyst. This cyst collectes fluids or mucus inside. Later, the cyst could become infected leading to the formation of an abscess.
    - A diverticulum at the bladder is another pathology. It most commonly presents at the dome of the bladder.
- Vitello-intestinal duct:
    - May present in adults as Meckel's diverticulum, which emerges from the ileum and connects it to the umbilicus resulting in umbilical fecal fistula.
- Omphalitis:
    - Inflammation of the umbilicus that occurs only in the newborns. The most common location is at the site of the attachment of the umbilical cord.
    - A pilonidal sinus is another presentation; however, it only presents in adults.
    - Inflammation of the umbilicus in newborns can be fatal due to portal vein thrombosis.
    - Before the era of antibiotics and intestive care omphalitis, portal vein thrombosis and hypertension were common.
    - In omphaltis, the infection can spread to the abdominal wall.
- Vesicointestinal fissure: herniation of the terminal ileum through the cecum foring the so called elephant trunk deformity

- Omphalocele:
    - A congenital defect of the anterior abdominal wall charecterized by the protrusion of the intestines covered by peritoneum and the amniotic membrane.
    - Incidence: 1:4000-6000
    - Pathogenesis:
        - An abnormal developlement of the gut prior to the 3rd week of embryonic life, which somehow precents the later return of the midgut into the abdominal cavity (physiological herniation during development that ffails to return to its normal position)
    - Types:
        - Epigastric: defect in the cephalic fold
        - Classic: defect in the lateral fold
        - Hypogastric: defect in the caudal fold
    - Surgical management:
        - Primary closure:
            - Hey: 1803
            - Indicated for infants with small to moderate isolated intestinal evistration
            - Primary closure without removal of the inflammatory peel
            - Limits: increase in intraabdominal pressures compromising lungs, circulation, and/or gastrointestinal contents
        - Staged reduction:
            - Schuster: 1967
            - Indicated for large defects and in infants who can't tolerate a primary closure.
            - Prosthetic silo of mesh-reinforced silastic material with progressive daily reductions over a week
            - Limits: mesenteric vascular compromise and infections after 7-10 days along suture lines.
    - Other types of omphaloceles:
        - Epigastric omphalocele: as a part of Canterlls pentalogy.
            - The pentalogy includes:
                - Lower thoracic wall malformations: cleft sternum
                - Cardiac defects: pericardial or ectopia cordis
                - Anterior midlie diaphragmatic defects
                - Epigastric omphalocele.
        - Hypogastric omphalocele:
            - Colonic agenesis
            - Exstrophy of the bladder
            - Impeforate anus

                - Vesicointestinal fistula
        - Clinical features:
            - Classic omphalocele:
                - Umbilical cord inserted into the apex of the sac
                - Small intestine contained within the sac with liver and spleen.
                - Bowel loops are normal
                - Abdominal cavity is less developed than in gastroschisis
                - Covering sac: avascular sac composed of fused layers of amnion and peritoneum
                - Associated anomalies:
                    - 33% Tetralogy of Fallot
                    - 19% ASD secondum
                    - Rare: VSD, PS, coarctation of the aorta, PDA, A-V canal
                    - Trisomy 21
                    - Beckwith-Wiedman syndrome
- Gastroschisis
    - Almost always presents to the right of the umbilivus
    - Here, the umbilical ring closes normally, but there is an accidental thrombosis of the right omphalomesenteric artery. This leads to loss of part of the right rectus muscle leading to a defect in the abdominal wall. The herniating intestine may shrink and peel off outside the body. However, they will return to normal once installed back in place.
    - Such patients can be covered with a plastic covering to prevent loss of fluids.
    - The size of the protruding bowel is larger than available space in the abdominal cavity. If they were installed back in place, they might compromise respiratory function. This necessitates the use of muscle relaxants and artificial ventilation for a few days. In addition, the lower limb's blood supply is monitored for dear of DVT or edema.
    - These patients require TPN

	Gastroscisis	Omphalocele
Location	Lateral to umbilicus	Umbilical ring
Defect size	Large usually >10cm	Small 2-4 cm
Cord	Inserted into the sac	Inserted into umbilicus
Sac	Present	Absent
Malrotation	Present	Present
GIT function	Prolonged ileus	Normal
Associated anomalies	Common	Infrequent, 10% atresia
Syndromes	Review text	Rare
Contents	Review text	Bowel only

- Management of omphalocele and gastroschisis (first aid)
    - Carefully wrap in saline soaked pads

    - Support without tension
    - NG tube for intestinal decompression
    - Abdominal ultrasound for nature of herniated viscera
- Investigations:
    - Serum: routine blood work
    - Imaging studies:
        - Abdominal X-ray
        - Abdominal ultrasound
        - 2D echo
    - Work up for associated anomalies
- Management (Definitive):
    - Supportive
    - ABC's (airway, breathing and circulation)
    - Temperature control:
        - Wramp infant in Salan wrap or a plastic drape
        - Warm saline bathing
    - Infection control: antibiotics
- Complications:
    - IUGR
    - Premature (10-50%)
    - SGA (35%)
    - Associated anomalies (review table)
    - Cardiac anomalies (21-40%)

Circumcision

- The removal of the prepuce (foreskin) and exposing the glans anteriorly. The prepuce is the skin of the penis that folds anteriorly and covers the glans, then meets the penis at the coronal sulcus. The inner surface of the prepuce is mucosa that normally secretes white material called smegma; this is a potential place for collection of dirty materials due to lack of hygiene
- Foreskin is retractable in 5% of newborns, 15% at 6 months, 50% at 1 year, and 90% at 3 years.
- Potential benefits of circumcision:
    - Protection against penile cancer
    - Protection against UTI
    - Protection against STD's
    - Protection against carcinoma of the cervix
- The aforementioned complications are usually due to lack of hygiene not to lack of circumcision. Those who oppose this procedure explain how to keep that area clean in order to prevent the transmission of these diseases. However, the hygiene of this area is difficult to maintain especially in newborns (especially that only 5% have retractable skin). Failure to clean this area will predispose the subject to the development of these diseases.
- Contraindications to circumcision:
    - Hypospadias and associated anomalies
    - Unstable or sick infant
    - Family history of bleeding tendency
- Potential complications of circumcision:
    - Serious:
        - Life threatening: bleeding and infection
        - Non-life threatening: fistula and amputation
    - Not serious:
        - Inadequate skin removal
        - Excessive skin removal
        - Inclusion cyst
        - Skin bridges
- Surgical technique:
    - Steps:
        - Free the foreskin
        - Identify the meatus
        - Retract the foreskin
        - Identify the coronal sulcus
        - Cut

    - Anesthesia: local, general, or none
    - Equipment: Glomco clamp and a plasty bell
- Ideal circumcision:
    - Neonatal age: smaller child, smaller surgery, and fewer complications
    - Local anesthesia
    - Selective delay for those who need it
    - Instruction for parents about risks, infection, and bleeding
    - Follow up
- Notes:
    - Phimosis: stenosis of the preputal ring with an inability to retract the foreskin. It is the most common medical indication FOR circumcision
    - Paraphimosis: retention of the retracted foreskin in an area proximal to the coronal sulcus
    - Balanitis: inflammation of the foreskin
    - Posthitis: inflammation of the glans
- Hypospadias:
    - Definition:
        - A congenital defect of the penis resulting in an abnormal urethral opening on the undersurface of the penis associated with incomplete development of the urethra, corpora cavernosa, and prepuce.
        - Incidence: 1:150-1:300
        - Clinically, results in deflection of the urinary stream and an abnormal appearance of the penis
        - Severe hypospadias is associated with Chordae (20%), which may result in infertility secondary to a difficulty in insemination. Here it represents a cosmetic and a function defect.
    - Embryology:
        - At one month of gestation male and female genitalia are indistinguishable
        - Masculinization of the male genitalia occurs under the influence of testosterone, so cases of hypospadias can be looked at as under masculinization of a male.
        - Male urethra and prepuce are completely developed by the end of the first trimester
        - Abnormal development leads to hypospadias, which results in dystopia, chordae, bifid scrotum, penoscrotal malposition, a hooded penis, and a ventral skin defect.
    - Classification:
        - Anterior (50%): the meatus is on the glans in a coronal or subcoronal position. The more anterior the milder.

        - Middle (30%): the meatus is on the shaft of the penis. Anterior and middle hypospadias represent cosmetic defects
        - Posterior (20%): the meatus is between the perineum and the penoscrotal junction. It represents a cosmetic and a functional defect.
    - Specific terms:
        - Subcoronal: opening on the coronal sulcus
        - On the shaft: proximal, mid or distal shaft
        - Penoscrotal and scrotal: abnormal scrotum opened exteriorly. It is a severe form
        - Perineal: similar to female urethra. The most severe form.
    - Associated anomalies:
        - Undescended testes: 10-30%. Depends on the severity of the hypospadias
        - Inguinal hernia: 10%
        - Utricle: remnants of the Mullarian duct system
        - Urinary tract anomalies are infrequent in isolated hypospadias.
        - Hypospadias alone or when associated with undescended testes or a hernia do not require further investigations.
        - Severe hypospadias, especially when associated with undescended testes, should be investigated for possible intersex with karyotyping and endocrine workup.
    - Treatment:
        - Surgery is best performed at 6-18 months of age.
        - Single stage or multistage (depending on severity)
        - Outpatient or inpatient
        - Goal of repair: normal urethra, normal glans
        - Final goals: straight shaft, normal skin, normal appearing meatus, normal skin coverage, and normal penoscrotal position.
    - Complications of surgery:
        - Formation of a fistula
        - Failure of the whole procedure. It is a difficult procedure' it has more than 250 ways for performing it. It is personal dependent.
    - Operation types:
        - Utilization of local tissue, skin tabularization, flaps, grafts, and urethral advancement
        - Utilization of adjacent tissue, skin, prepuce, penile skin, and scrotal skin in the form of flaps or grafts.
        - Utilization of remote tissue, skin mucosa, buccal mucosa, or bladder mucosa
        - Staged repair
        - Operation for chordae correction

    - Results and complications: success rate is 70-100%. This depends on the severity of the hypospadias, the surgical technique, and the surgeon
- Epispadius:
    - Extremely rare condition (1:20,000)
    - Urethral opening in on the dorsal surface of the penis.
    - Associated with an abnormal penis. Usually associated with a syndrome that includes extrophy of the urinary bladder.

Inguinoscrotal disorders in children

- Hernias, hydroceles, and undescended testes form the majority of inguinoscrotal disorders in children.
- Acute scrotum is a rare condition that usually presents to the ER, not to the clinic
- Hernias and hydrocele:
    - In children, both have the same basic pathology, a patent processus vaginalis.
    - Important structures in the spermatic cord include the vas deferens, testicular artery, veins (that start as the pampiniform plexus and continue as the testicular vein), and a processus vaginalis.
    - Processus vaginalis is the obliterated part of the peritoneal pouch that was pulled down from the peritoneal cavity during testicular descent. The remaining part surrounds the testicles, and is called tunica vaginalis.
    - In adults, a secondary cause (tumor or infection) leads to the accumulation of fluids in tunica vaginalis However, in children, the fluid accumulates due to a patent processus vaginalis.
    - Management in adults is different than that in children.
    - To differentiate between a communicating hydrocele and a hernia, look at the neck of the mass. Hydroceles have narrower necks, thus allowing peritoneal fluid rather than abdominal viscera into the patent processus vaginalis.
    - 90% of children have a patent processus vaginalis at birth. However, the incidence of hernia is 1-2% of the general pediatric population.
    - These lesions are more common on the right side. Bilateral hernias occur in 5-15% of children with hernias.
- Hernias:
    - Conditions associated with a higher incidence of hernias:
        - Premature babies
        - Family history
        - Ascites and VP shunt
        - Abdominal wall defects
        - Ambiguous genitalia and hypospadias
        - Extrophy of the bladder
        - Undescended testicle (almost always associated with inguinal hernia)
        - Mucopolysaccharidosis: a connective tissue disorder that causes weakening of the fascia of the abdominal wall. It is associated with a high recurrence rate.
    - Presentation of a hernia:

        - Present at 1-2 years
        - Can present as a complicated or an uncomplicated hernia.
        - When complicated, the presentation is either an irreducible, strangulated (signs of obstruction including sepsis), or obstructed hernias (signs of obstruction).
        - In the case of an uncomplicated hernia, the mother will usually pinpoint the site of the bulge. Most of the time, the bulge is reduced at the clinic. The most common causes for a bulge in that area include a hernia or a retractile testicle where cremasteric muscles are hyperactive.
        - Diagnosis depends, in the majority of cases, on the history taken from the mother. A hernia will bulge when the baby cries, and reduces when the baby is relaxed.
        - During physical examination, the affected side may have a thicker cord as the cord has extra layers (hernia's sac)
    - Management of a hernia:
        - Uncomplicated hernias must be operated. They will not resolve spontaneously.
        - The procedure of choice is herniotomy.
        - In children, the procedure must be performed as soon as possible; on the next available operative list. In the case of NICU babies who have problems other than hernias, the policy is to delay the hernial repair until their discharge from the NICU. It is important to treat the hernias of the immature as they have a higher rate of complications than those of odler children.
    - Exploration of the other side (ETOS):
        - 10-15% of children with a hernia on side will develop a hernia on the other side.
        - It is controversial whether exploration of the other side should be done routinely in children younger than 1 year of age.
        - ETOS is done when the hernia Is on the left side.
        - In females, ETOS can be done with little or no compications; however, in males, we are afraid of damaging the testicular artery or the vas. Moreover, the incidence of a contralateral hernia is more in females.
        - ETOS is done routinely in cases of: female patient less than 1 year of age, VP shunt, ascites, and periotneal dialysis
        - A laporoscopic exploration is not considered as part of ETOS. However, exploration by a laporoscope is one of the means by which one can screen the geenral population for a patent processus vaginalis. Other means include post-mortum studies.

    - Complicated hernias usually present in the ER with pain and signs of sepsis. Management of these patients has three steps:
        - Resuscitate: if the patient is in shock
        - Reduce: if there are no signs and symptoms of strangulation. The majority will reduce; however, if the baby is agitated, it is hard to deal with the case. Thus, these babies are usually sedated. In many cases, once the baby is put to sleep, hernias will reduce spontaneously.
        - Repair:
            - If reduced: repair within 24-48 hours. This will give the echymoses and edema of the cord structures a chance to decrease, which will make the herniotomy easier.
            - If not reduced: to avoid intestinal obstruction and testicular atrophy, it is operated within hours.
- Hydrocele:
    - Excess fluid in tunica vaginalias.
    - Classification:
        - Communicating: communicating with the peritoneal cavity. Fluid accumulation happens at the end of the day; absent upon waking up.
        - Encysted hydrocele: here, the fluid is sequestered between two obliterations. In some cases, the cyst is large enough to mimic the communicating hydrocele. If you can reach above the swelling, it is an encysted hydrocele. In the case of an encysted hydrocele, the swelling is separate from the testes. However, in the case of a communicating hydrocele, the fluid surrounds the testicles.
    - Management:
        - In most infants, the hydrocele will resolve within the first year. These patients should be observed during this period. If the hydrocele increased in size within this first year, it will not resolve spontaneously. However, if it got smaller, leave it for one more year.
        - Any hydrocele appearing after the first year must be operated. It will not resolve spontaneously.
- Undescended testicles:
    - A testicle which is not in the scrotum
    - Incidence:
        - Newborn: 4%
        - 9 months: 1%
        - 18 years: 1%
    - If the testicles do not descend by the age of 9 months, they will not descend later on spontaneously.
    - Undescended testicles should be managed at the age of 1 year.

    - 20% of undescended testicles are impalpable; of them 25% are absent because of neonatal or ante-neonatal torsion. The rest are found in the abdomen or the inguinal canal.
    - Classification:
        - Retractile: a testicle with a normal cord length. The only problem here is the presence of a hyperactive cremasteric muscle. They can be considered as normal testes as long as they size is equal on both sides, they persist in the scrotum after pulling them, and present in the scrotum most of the time.
        - Ectopic: a testicle found in a site other than the normal tract of the descent.
        - True: a testicle that stopped its descent along the normal tract of descent.
    - Why is this important?
        - Infertility: parents should be reassured that the patient would be a normal individual. However, it should be noted that patients with undescended testicles are less fertile than the general population
        - Trauma: as the testicle is found in an abnormal place, it is prone to trauma
        - Torsion: torsion of the testicles at the level of rete testis.
        - Hernia
        - Cancer: in general, the incidence of testicular cancer is rare. In those children, there will be a 20-40 fold increase in their chances of getting testicular cancer. However, it is still a rare entity
    - Treatment:
        - Orchidopexy by the age of 1 year (6-12 months).
        - After two years, the testicles will be abnormal and dysfunctional.
        - Biochemical changes start by 6 months. Histological changes start at the end of the first year
- Acute scrotum:
    - Scrotal pain and swelling
    - Differential diagnosis:
        - Testicular torsion
        - Torsion of the testicular appendages, which has a similar presentation to testicular torsion
        - Epididymoorchitis
    - Acute scrotum is testicular torsion until proven otherwise. Usual torsion of the adults occurs at the level of rete testes. However, in the neonatal and anteneonatal period, torsion occurs at the cord.
    - By the time they are examined, the testicles are usually necrotic.
    - If the patient presents with an acute scrotum, you must make sure that they reach the operation room immediately. Here time is a very important factor. If the

testicles were operated in the first 6 hours, the salvage rate is about 90-95%. However, if operated after 24 hours, the salvage rate is 10%.

Hirschsprung's disease

- Absence of the ganglionic cells distal in the bowel as a result of failure of the cephalocaudal migration of neural crest cells. The result is a functional intestinal obstruction with a contracted non-peristaltic affected segment and a dilated hypertrophied proximal segment. The disease is also known as congenital mega colon and aganglionic mega colon.
- History:
    - Hirschsprung first described the disease in 1886; however, he knew nothing about the underlying pathology. Later on, many years after his death, pathologists discovered the absence of ganglionic cells. The disease was name after him to honor him
- Incidence:
    - 1:4000 live births
    - Male: female 4:1
    - Familial tendency in 5-10% of cases. It is usually associated with agangliosis of longer segments.
    - Females tend to have longer affected segments.
    - Other anomalies such Down's syndrome are present in 10-15% of cases.
- Classification:
    - Ultra short: the aganglionic segment is distal to the peritoneal reflection
    - Short: affects the rectosegmoid area
    - Long: affects the rectosegmoid area and the descending colon
    - Total colonic agangliosis: affects the whole colon and the terminal ileum
    - Total agangliosis: the most severe form; ganglionic cells are absent along the total length of the GIT. It is a fatal disease.
- Presentation:
    - Failure to pass meconium in the first 24-48 hours of life
    - Chronic or intermittent constipation
    - Intestinal obstruction, abdominal distention, vomiting, constipation, and abdominal colic
    - The neonate may develop enterocolitis, so the patient presents with explosive watery diarrhea and fever. Enterocolitis is caused by Clostridium difficile in 33-50% of cases. It is of high mortality rate and the neonate, once diagnosed, should be admitted immediately. Management is aggressive to prevent the fatal complications such as sepsis and perforation.
    - In older children, chronic constipation with persistent abdominal distention in the absence of soiling.
    - Malnutrition with poor growth and development.
- Differential diagnosis:

- Rectal or colonic atresia
- Meconium ileus
- Meconium plug syndrome
- Abdominal hernias causing intestinal obstruction
- Intestinal volvulus
- Systemic diseases that may mimic Hirschsprung's disease:
  - Sepsis due to encephilitis or pneumonia
  - Uremia
  - Hypothyroidism, but not in neonates.
- Habitual constipation in older children

Hirschsprung's disease	**Habitual constipation**
Starts early in life. The diagnosis can be delayed in mild cases	Usually seen in older children
It is a serious disease that causes malnutrition, failure to thrive, and anemia	The patient is only constipated; otherwise healthy.
Alternation of constipation with diarrhea	Patient is constipated the whole time; they never have diarrhea
PR examination may be followed by a gush or expulsion of watery stool and gas. This results from dilation of the sphincter by the examining finger.	The stool is hard and never gushes.
No anal fissures because the constipation is high up. When the stool passes, it is not hard.	Patients almost always complain of an anal fissure
No soiling	Soiling is frequent

- Diagnosis:
  - History
  - Physical examination:
    - The majority of patients are full term (weight at birth 3-3.5 kg)
    - During PR, the rectum is empty and the sphincter is tight. A gush of watery stool might take place.
    - On abdominal examination, distention is found and the impacted stool in the greatly dilated and distended sigmoid colon can be palpated across the lower abdomen
  - Investigations:
    - Plain abdominal X-ray: you can see dilated loops of bowel. Air fluid level indicates a distal obstruction
    - Barium enema: you can recognize the affected narrowed segment, a transitional zone, and a dilated segment (funnel appearance). Irregular, bizarre contractions of the aganglionic segment may also be recognized

giving a saw-tooth appearance. Moreover, the passage of the contrast can be delayed.
        - Anorectal manometry: in normal subjects, when the intrarectal pressure increases due to accumulation of stool, the anal sphincter relaxes allowing the stool to pass and defecation takes place. This is what is known as the anorectal reflex. In Hirschsprung's disease, this reflex is absent. The manometric study is done by inserting a balloon in the rectum and inflating it to increase the rectal pressure. In the case of Hirschsprung's disease, the sphincter's pressure will increase instead of decreasing.
        - Rectal biopsy: definitive diagnostic tool. There are 2 types of a biopsy: the suction rectal biopsy done in children, and full thickness biopsy done in adults.
        - Immunohistochemistry: cholinesterase enzyme in the mucosa. High levels are due to hypertrophy in the nerves.
- Treatment:
    - Surgery is the only treatment. The idea is to remove the aganglionic segment, then perform a pull through procedure. However, it is important to preserve the anal sphincter as much as we can in order to prevent incontinence.
    - The operation can be done in 3 stages or as a 1 stage operation:
        - The multi-staged procedure: since surgeons cannot work on a dilated bowel, a colostomy is done first. It is left until the bowel returns to normal status; then, the affected segment is resected. This is followed by closure of the colostomy. An anastamosis is made in a third stage.
        - The one-staged procedure: the colostomy stage is replaced by conservative procedures that cause the dilated segment to return to its normal status. These include irrigation of the bowel, rectal washout, and saline enemas. The surgery is done to resect the affected segment
    - Surgical techniques:
        - Swenson operation: Removal of the whole rectum except for 2 cm interiorly to protect the prostate area
        - Duhamel: modification of the pull through procedure with the establishment of a longitudinal anastomosis between the proximal ganglionated segment of the colon and the rectum leaving the rectum in situ.
        - Soave operation: anorectal pull through operation with the normal colon connected to the anus through a rectum denuded of mucosa.
    - Complications:
        - Anastomotic leak
        - Necrosis of the pulled through section
        - Incontinence and soiling

- Episodic constipation and abdominal distention if the internal sphincter is kept completely intact.

GI bleeding in infancy and childhood

- GI bleeding in the pediatric group differs from that of the adults in the sense that the disease causing the bleeding in each group is different. Furthermore, adults usually have dependent bleeding (there is a concurrent pathology such as atherosclerosis) and, therefore, have higher mortality rate. However, a better prognosis is expected in pediatrics after treating the cause of bleeding as their bleeding is usually a single problem.
- Regardless of the site, bleeding in a child is very alarming to the family and the child is often promptly taken to the hospital. This is very fortunate for the child as this decreases the blood loss and allows for rapid treatment. This is especially critical in pediatrics considering the small blood volume in patients of this group. It is estimated to be 80-85 ml/kg. For example, a 1 year old weighing 10 kg will have nearly 850 L of blood. To this patient, losing 500 mL of blood is fatal; which is definitely not the case in adults. In other words, even minimal amounts of blood loss in pediatrics is serious. Likewise, replacement of blood loss in pediatrics is very rewarding. Therefore, it is imperative to estimate the blood volume whenever a child presents with bleeding (and in cases of dehydration, as well). In addition, because the amount of blood is small, the amount of blood taken for cross matching or for any other purpose should be small, as well. In summation, mortality in children is low because of the rareness of comorbid conditions and attentive care provided to them. GI bleeding an uncommon occurrence in pediatrics and is usually of a benign etiology and spontaneous resolution.
- Upper GI bleeding:
    - Neonates:
        - Maternal blood: during delivery, the newborn might swallow some of the maternal blood. The blood remains in the stomach temporarily and is then evacuated as hematememsis or passed as melena or hematochezia
        - PUD: more common in older age groups, but it is very a very common cause of GI bleeding in the pediatric group as PUD accounts for nearly half the cases of upper GI bleeding. It increases in incidence with increased age. It might occurs in cases of stress in children (asphyxia during birth)
        - Trauma: from an NG tube; during resuscitation of newborns after delivery an Ambu bag is used. Air from the bag will be propelled into lungs and stomach. Other purposes of NG tube mainly involve decompression of the GIT. Injury might involve the esophagus or stomach and is usually related rough handling of the NG tube.
        - Necrotizing enterocolitis: the cause of which is mostly ischemic, complicated by infection and sepsis. It first involves mucosa and may reach full thickness after the mucosal barrier is lost. Peritoneum may be

involved afterwards. These patients mainly present with intestinal obstruction; however, they may present with GI bleeding. It is a disease of prematurity. With the progress achieved in neonatal care, it has been possible to save the lives of more premature newborns.
        - Coagulation disorders: a rarity nowadays in pediatrics as all newborns are given a vitamin K injection soon after birth
    - Infants:
        - Duplication cyst: duplication of a segment of the bowel. It presents on its mesenteric border resulting in two adjunct tubes. It may involve any part of the GIT and may be a cause of intestinal obstruction, as well. The lining of these structures is usually abnormal, and may be gastric in nature. Thus, it secretes acid, which leads to peptic ulceration and bleeding similar to that in the stomach. These cysts can be tubular or round, communicating or non-communicating. They may present with a picture of intestinal obstruction of Hi bleeding. Technetium scan helps in localizing the lesion.
        - Reflux esophagitis: reflux during the first two years of life is very common that some consider it as a normal condition. It may result in esophagitis that leads to bleeding. Reflux esophagitis is one of the indications of surgery in children with GERD
        - Medications: NSAIDs. Aspirin is rarely used in children
        - Caustic ingestion: such as “hypex”. Usually presents with an esophageal stricture. Acids usually injure the stomach, while alkali injure the esophagus
    - Preschool age:
        - Varices: appear at the age 3-4. They appear after the KAsai operation. AFter this operations patients will ultiately develop cirrhosis responsible for varices
        - Epistaxis: patient may swallow (especially while sleeping) blood from the nose. This is similar to swallowing maternal blood in neonates. This is common in patients who have undergone tonsillectomy
        - Mallory-Weiss tear: seen more often in adults (usually alcoholics) where frequent vomiting leads to tears in the lower esophagus and gastro-esophageal junction. In children, repeated vomiting, whatever the cause is, results in the same outcome
    - School age:
        - Vascular anomalies: Hemangiomas and vascular malformations. They may arise anywhere, when external, they are easily detected.
        - Diffuse lesions
        - Vasculitis: Henoch Schnollen purpura
- Diagnosis:

- Start with history and physical examination
- In the history:
  - You should be familiar with the following terms: hematemesis, melena, hematochezia, maroon stool (has characteristics between melena and hematochezia)
  - The closer the source of bleeding to the anus, the more fresh the blood per rectum is. However, a patient with gastric ulcer may present with fresh blood per rectum. The most common cause of massive bleeding per rectum is massive upper GI bleeding, both in adults and children.
- In the physical examination:
  - Hypotension (the patient must lost 1/3-1/2 of blood volume for hypotension to occur)
  - Skin findings: Henoch Schonlein purpura (purpuric rash in dependant areas)
  - Abdominal findings :ascites, hepatomegaly, splenomegaly, and other signs of portal hypertension
- Insert NG tube:
  - Perform a gastric wash .when saline is injected in NG tube and a clear fluid is sucked out, it indicated cessation of bleeding from that level of the GIT
- Gastroscopy: better and less time consuming. Can be used diagnostically and therapeutically

- Investigations:
  - CBC
  - Coagulation profile
  - LFT
  - Apt test: performed in neonates to find out if blood is swallowed or from the neonate. Take some vomitus or melena, dilute with saline, centrifuge, and then add NaOH. Fetal Hb willl not react with NOH, so there won't be a change in color of the pink supernatent. In contrast, maternal Hb will change into a brown-yellow color.
  - Radiology and nuclear imaging: help in certain situations. They are not diagnostic in most cases. C-rays may reveal foreign bodies or gas bubbles in the wall of the bowel (in necrotizing enterocolitis). Technitieum scan is used in cases of suspected Meckel's diverticulum or duplication cyst as the technitium is withheld in gastric mucosa anywhere in the body.
  - Endoscopy: the mainstay for diagnosis. Upper GI endoscopy is both diagnostic and therapeutic (as it may be used to treat a bleeding ulcer or esophageal varices). Endoscopy is of a special importance in cases of serious unexplained bleeding. Findings:

        - In 40% the cause of bleeding is an uncler in the stomach or duodenum
        - 15% esophagtis
        - 10% varices
- Treatment:
    - Resuscitation is extremely important and should be done even before investigating of the cause. It includes O_2 administration and insertion of 2 canulas (minimum) to restore blood volume.
    - In trauma patients, it is preferable to insert a canula in the upper limb and one in the lower limn, but in cases of GI bleeding, this doesn't matter.
    - Treat underlying diseases including coagulation disorders, hemophilia, or cardiomypathies.
    - Certain medical regimens may be used in cases of PUD.
    - Endoscopic therapy includes electrocoagulation, photocoagulation, and injection of sclerosing agents in or around esophageal varices.
    - Surgery is the last resort for therapy if bleeding continues and cannot be controlled by medical or endoscopic means. Each disease has its own surgical method for treatment.
- Lower GI bleeding:
    - It is usually more obvious than upper GI bleeding. The most common causes include polyps or anorectal fissures.
    - Anorectal blood: presents as drops of fresh blood
    - Allergic proctocolitis: A sort of gastroenteritis. Allergy to certain proteins in milk that disappears once the formula of milk is changed
    - Intussusception: seen between 6-18 months (mainly between 9-12 months). The presentation is that of intestinal obstruction. In 60% of cases, blood and mucus are passed per rectum (described as jelly-like)
    - Juvenile polyps: characteristically presents as fresh blood per rectum and mass prolapsing through the anus. It is a hamartamatous polyp. Treatment is excision and the excised polyp is sent for pathological analysis to make sure that it is not neoplastic
    - Meckel's diverticulum: arises on the anti0mesenteric border of the small intestine; the opposite of a duplication cyst. Bleeding occurs because the extopic gastric mucosa secretes acid and injures adjacent mucosa of ileum and causes ulceration and then bleeding. Rule of 2:
        - 2% of the population has it
        - Presents 2 feet from ileocecal junction
        - Bleeding in children of less than 2 years of age
        - 20% of cases have ectopic mucosa.

        - Treated by excision. Preoperative diagnosis is rare as Meckel's diverticulus is usually an operative finding. Technetium scan is helpful in diagnosis.
    - Diagnosis:
        - History and physical examination
        - Colonoscopy is the preferred diagnostic modality for all cases of rectal bleeding. It may be used therapeutically as in excision of polyp
        - Radiology and nuclear medicine

Pyloric stenosis and intussucecption

- Pyloric stenosis:
    - General features:
        - It is a disease of early infancy
        - It has many other names including: hypertrophic pyloric stenosis, congenital hypertrophic pyloric stenosis (a misnomer because it s not congenital). Infantile hypertrophic pyloric stenosis. All the names indicate that tehre is hypertropy of the pyloric muscles and this hypertrophy causes pyloric obstruction.
        - It is one of the most common surgical conditions in the west, so there is a strong racial variation
        - There are genetic factors' if the mother and/or father were affected, then their children have a very strong tendency to be affected, but specific genes haven't been located, yet. The tendency increases if the mother is affected more than if the father Is affected
        - It affects males more than females' male: female ratio is 4-6:1
        - Overall incidence: 3/1000 live births.
        - Affects a very limited age group 3-6 weeks. It rarely affects a child more than 2 months of younger than 3 weeks of age. We can find atypical cases under the age of 3 weeks or more than 6 weeks.
    - Signs and symptoms:
        - Progressive, persistent, projectile, non-bilious vomiting.
        - Associated with a good appetite. This is not a sickening or systemic disease. There is obstruction of the pylorus and so there is coming caused by the peristalsis of the stomach. The baby is still hungry and eats more. This is what leads to chronic dehydration not acute as in cases of acute diarrhea or vomiting due to infectious diseases which are associated with fever. The obstruction is partial, so some food will pass through. The baby will drink milk more than usual, so it is a deceiving disease.
        - Constipation is typical
        - Gastritis due to prolonged vomiting and hematemesis, which will lead to anemia due to chronic loss of blood.
        - Olive sign: the enlarged pyloric canal is felt during physical examination like an olive to the right of the vertebral column just below the liver.
        - Succusion splash: pyloric stenosis in children has a mirror image in adults, which is pyloric stenosis due to PUD. This causes narrowing of the pyloric region due to strictures, not hypertrophy. It has the same clinical picture: chronic obstruction, chronic vomiting, and a distended stomach. After

ingestion of fluids, the patient's stomach will produce audible sounds when moving the patient
        - Visible peristalsis: you may see peristaltic waves of the stomach from left to right. This indicates that the stomach is hypertrophic and trying to overcome the obstruction.
- Pathophysiology:
    - Hypertrophy of the smooth muscles of the antrum due to unknown reasons. This causes narrowing of the pyloric canal, which becomes easily obstructed.
    - Vigorous peristalsis
    - Hypertrophy of the stomach
    - Gastritis, hematemesis, dehydration
    - The end result is hypokalemic hypochloremic metabolic alkalosis. Due to excessive vomiting, there will be a massive loss of acids, potassium and chloride. The kidneys are responsible for correcting the pH and electrolytes; if there is alkalosis, they will excrete alkaline urine. In this case, there will be excretion of Na and K to retain H ions. However, if vomiting increased to cause dehydration, the kidneys will prefer life over quality of life. Under the influence of aldosterone, there will be retention of sodium accompanied with water, loss of potassium, and paradoxical acidurea.
- How did the pathophysiology come into being?
    - Failure of relaxation, which results in spasms
    - Abnormal ganglia were found in some cases, but not all of them
    - Deregulation of the VIP, nitric oxides, and polypeptides of the GIT.
- Diagnosis:
    - Typical clinical picture in 90% of the patients.
    - Ultrasound: it has become the first choice for a diagnostic tool. Through ultrasound, we can see the thickening of the pyloric wall along with a long pyloric canal.
    - Barium swallow: used to see the elongation of the pyloric canal
    - Serum electrolytes, CBC, and pH
- Differential diagnosis:
    - Overfeeding: the commonest cause of vomiting in the neonal and posneonatal periods
    - Gastroesophageal reflux
    - Pylorospasm
    - Delayed gastric emptying
    - Duodenal stenosis
    - Malrotation

        - Any partial intestinal obstruction

    - Treatment:
        - Medical treatment to correct dehydration and acid base imbalances. You don't treat with normal fluid maintenance. These patients are given normal saline or Ringer's lactate. They are given 1.5-2 times their maintenance needs because of suspected dehydration.
        - Surgical treatment: pyloromyotomy to open the muscle of the pyloric canal without incising the mucosa

- Intussuception:
    - Invagination of a proximal segment of the bowel into an adjacent distal segment. It is a cause of partial intestinal obstruction (in later stages it might cause complete obstruction)
    - Epidemiology:
        - Incidence: 2-4/1000 live births.
        - There are racial and geographical variations.
        - Slightly affects males more than females.
    - Age incidence:
        - Specific narrow age group: 3-9 months
        - Idiopathic up to 3 years of age. After that, it is very rare and becomes a different entity)secondary causes)
    - Clinical picture:
        - Prodromal febrile illness related to the GIT. Starts with lethargy, malaise, and fever. Then the patient starts to complain from other symptoms.
        - A previously healthy child, who suddenly comes with acute abdominal pain (colicky in nature). The pain lasts for seconds then disappears, then appears again. The baby may become pale and cyanosed. In between the attacks, the child is healthy looking.
        - Vomiting
        - Blood in the stool (Redcurrant jelly stool). Intussuception is the commonest cause for intestinal obstruction in children between 5 months and 3 years. Like any other cause of intestinal obstruction, there is a mechanical obstruction that causes lymphatic obstruction, venous obstruction, arterial obstruction, then and infarction. This will lead to the formation of a gangrene, sepsis, or shock. When there is ischemia, the first part to get infarcted is the mucosa because it is the farthest from the blood supply; this causes the redcurrant jelly stool.
    - Physical examination:
        - A mass can be palpable in 50% of the patients

        - Abdominal distention if neglected due to intestinal obstruction
        - Sepsis due to intestinal obstruction
        - Shock
    - Etiology:
        - Idiopathic:
            - 5 months- 3 years
            - At the ileocecal junction
            - Hypertrophy of the lymphoid tissue in the ileocecal junction, which leads to thickening of the wall leading to intussuception
        - Due to an underlying cause (tumor, polyp, Mickel's diverticulum, surgical causes)
            - Above 3 years of age or in adults
            - Either at the jejuno-jejunal junction, jejuno-ileal junction, or the ileo-ileal junction, but not at the ileocecal junction.
    - Diagnosis:
        - Clinical features: the typical presentation is a previously healthy child with acute attack of colicky abdominal pain associated with vomiting and blood in the stool
        - Plain X-ray: first investigation to exclude intestinal obstruction
        - Ultrasound: to show two lumens inside each other
        - Barium enema: diagnostic as well as therapeutic
    - Differential diagnosis:
        - Acute abdomen
        - Appendecitis
        - Gastroenteritis (very common, but does not mimic intussuception)
        - Encarcerated hernia
        - Volvolus
    - Management:
        - Correct fluid and electrolyte imbalance
        - Nasogastric decompression
        - Conservative or operative treatment:
            - Conservative:
                - Hydrostatic reduction
                - Retrograde gas reduction: gas is introduced through a catheter under pressure control. Pressure does not exceed 90-110 mmHg. The pressure introduced distally to proximally will reduce the intussuception
                - Barium enema: in early stages, the success rate reaches up to 80%
            - Operative:

- Indications:
  - Peritonitis
  - Sepsis
  - failure of the conservative treatment
  - Recurrence (5%)
  - When there is a leading point
- Procedure:
  - Manual reduction of Intussuception.
  - Look for a leading point
  - Resection if:
    - unable to reduce the lntussuception
    - there is a gangrenous bowel segment
    - presence of a leading point.

- Prognosis
  - In infants: Excellent
  - Older age: depends on the underlying pathology.
- NOTE:
  - Intussuceptum: the proximal segment
  - Intussucepient: the distal segment

Pediatric trauma

- Epidemiology:
  - The most common cause of death 1- 14yrs of age
  - Very common, high mortality, high morbidity, very costly.
  - Fatality, MVA, bicycle, Drowning, House fires (burn), Homicide.
  - Multisystem injury is the rule.
- Death due to trauma
  - 20% Head injury alone
  - 30% Disability
  - 70% Head injury plus other associated organs
  - 80% are blunt injuries
  - Birth trauma
  - Child abuse
- Death in children
  - 50% Accidents
  - 10% Malignancy
  - 5% Congenital anomalies
  - 35% Miscellaneous, infections, metabolic etc.
- Death due to trauma
  - 20% Head injury alone
  - 30% Disability
  - 70% Head injury plus other associated organs
  - 80% are blunt injuries
  - Birth trauma
  - Child abuse
- Children are
  - Neonates
  - Infants
  - Toddlers
  - Preschool age
  - School age
  - adolescents
- Children are different:
  - Larger surface area to mas
  - Less subcutaneous fat
  - More body water
  - Higher metabolic rate
  - Size and shape
  - Skeleton

    - Body surface area
    - Psychological status
    - Equipment related to size
- Fluid balance:
    - Maintenance fluid requirement:
        - 0-10 kg: 4ml/kg/hour
        - 10-20 kg 2 ml/kg/hour + 40 mL
        - 20+ kg 1 mL/kg/ hour + 60 mL
    - Adequate urine output: 1-2 mL/kg/hour
    - Weight estimate: 8 + (Age x 2)
    - Circulating blood volume: 85 mL/kg
- Venous Access
    - Percutaneous peripheral
    - Intraosseous
    - Percutaneous femoral
    - Venous cut down
    - Percutaneous external jugular (neck collar or airway compromise)
- Priority of resuscitation: A, B, C, D, E
    - Airway, Breathing, Circulation, Disability, Exposure
    - Anatomical and physiological trauma score systems
    - Pediatric Trauma Score Systems.
    - (PTS), (Glasgow coma score), ISS
- Priority by injury
    - Injuries that interfere with vital functions and immediately: life threatening; airway obstruction, massive bleeding
    - Injuries that are not life threatening: cut wounds, bruises, stab wounds, and blunt head trauma (Majority)
    - Injuries that cause occult damage. closed head and abdominal injuries. potentially life threatening. (latent hemorrhage
- Fluid for resuscitation
    - 20cc/Kg =25% of Blood Volume(80cc/Kg).
    - can be repeated once.
    - CRYSTALLOID solution
    - If no response:
        - 10cc/Kg Type specific, Onegative blood, PRBCs.
    - If no response to RBC's: Surgical intervention is required,
    - If good response
        - TRIAGE
        - TRANSPORT,
        - THE GOLDEN HOUR

- What is a good response?
    - Decrease in pulse rate <130 bpm; pulse pressure <20
    - Decrease in skin mottling
    - Improving sensorium
    - Increase in urine output >1 mL/kg/hour
    - increase in systolic blood pressure >80
- Battered child (child abuse)
    - Discrepancy between history and physical injury
    - Prolonged interval between injury and seeking medical advice
    - History of repeated trauma, different E/R
    - Parents response inappropriately or don't comply with medical advice
    - Different history between parents
    - Multiple subdural hematoma, especially without skull Fracture
    - Retinal hemorrhage
    - Perioral injuries
    - Ruptured internal viscera without antecedent major blunt trauma
    - Trauma to genitalia or perianal region
    - Evidence of frequent injuries typified by healed scars or healed fractures
    - Fracture of long boned in younger than 3 years
    - Bizarre injuries such as bites, cigarettes marks or rope marks
    - Sharp demarcated second and third degree burns in head or unusual areas

Foreign Body Aspiration

- It's one of the real life threatening emergencies which require immediate intervention.
- Age incidence:
  - Children between 9-30 months are more affected, because of their mobility and their oral orientation at this age.
  - Older children and adults add to the problem by giving them things to eat
- Types of Foreign Bodies
  - Organic materials: Peanuts, Sunflower seeds ,Almond , Popcorn, Apple or orange seeds
  - Non-Organic: Buttons, Toy's parts
- Location:
  - In adults:
    - 69% (the majority) of F.B. go to the right side because the trachea and right main bronchus are more in line.
    - 31% located in the left' bronchial tree.
    - 3.6% located in both sides.
  - In children
    - 52.2% located in the right bronchial tree
    - 47.5% located in the left bronchial tree
- Symptoms:
  - The most important symptoms is penetration syndrome defined as sudden onset of choking and intractable cough with or without vomiting
  - Cough
  - Fever
  - Breathlessness
  - Wheezing
  - Cyanosis
  - No symptoms
- Progression of the disease:
  - Symptomatic phase: penetration syndrome
  - Asymptomatic phase
  - Phase of complications: due to foreign body obstruction of certain parts of the lung for a long time (one or two weeks), these complications include:
    - Pneumonia
    - Lung abscess
    - Bronchiectasis
    - Hemoptysis

        - Erosion and perforation: it may perforate the tracheobronchial tree and go to the esophagus or aorta causing severe bleeding, or it might penetrate the chest wall
    - The only way to exclude foreign body aspiration is to do a bronchoscopy. If you find a foreign body upon bronchoscopy, remove it. If you don't find anything, you lose nothing!
- Signs:
    - Respiratory distress (tachypnea, cyanosis)
    - Localized wheezing; when there is significant narrowing or partial obstruction, it is heard over one side of the chest
    - Poor air entry
- X-ray findings:
    - Atelectasis: 14%
        - Collapse of a lobe or segment of the lung depending on the location of the foreign body
    - Air trapping 64%:
        - Emphysematous change
        - Sometimes, the foreign body causes partial obstruction and works as a valve allowing for air entry. However, upon exhalation, the diameter of the airways will decrease, which prevents air from going out trapping it inside. This change is the most common radiological finding shown on X-ray. It may be called hyperlucency of the lung of hyperinflation of the lung. The change is visible on the X-ray as:
            - Radiolucency of the affected side, which appears hyperinflated
            - The space between the ribs)intercostals) is wider
            - Diaphragm is pushed down
    - Pneumonia: 13%: if the foreign body stays for a long time
    - Visible foreign body: 4%
    - Normal radiograph: 12%: usually, there is a residual abnormality after removing the foreign body
- Treatment: bronchosopic removal of the foreign body; any child that is suspected to have a foreign body in the airways should undergo bronchoscopy for diagnosis and treatment.

Esophageal foreign bodies:

- Management of the foreign body in the esophagus is totally different from that below the diaphragm
- Narrowest parts of the esophagus:
    - The cricopharngeous muscle or superior constrictor of the esophagus
    - At the level of the aortic arch
    - At the level where the left main bronchus crosses the esophagus

    - Esophageal anomalies (make the esophagus narrower)
        - Esophageal atresia or fistula or those with a corrosive injury damaging the esophagus leading to fibrosis, narrowing, and an abnormality in the peristaltic activity
        - Vascular rings: anomalies in the innominate artery or aortic arch
        - Cartilaginous rests
        - Webs in the esophagus
        - Duplication cysts
        - Achalasia and peptic strictures
- Symptoms:
    - The most common: drooling of saliva, dysphagia, and pain
    - Bleeding is a late phenomenon that may be a lethal complication of erosion or perforation into the aorta or pulmonary artery
    - An esophageal foreign body may erode into the lumen of the airway producing a broncho-esophageal fistula
- Complications:
    - Aspiration
    - Perforation: either to the trachea (tracheo-esophageal fistula) or to the aorta (aorto-esophageal fistula)
- Radiological findings:
    - Radio-opaque foreign bodies: here, we see radio-opaque foreign bodies more than tracheobroncheal foreign bodies
    - Barium swallow: rarely used, but if the history and physical examination are atypical it is done
- Treatment:
    - Esophagoscopic removal
    - A foreign body in the esophagus is unlikely to pass down, so remove it

GIT foreign bodies:

- Approximately, 95% of foreign bodies that are infested and reach the stomach pass the remainder of the GIT without any problems (even sharp objects)
- Exceptions include long objects such as a toothbrush, an elongated key, or a pencil. These objects are likely to become stuck within the duodenum or at the ligament of Trietz
- Pins and nails may perforate the wall, but usually dislodge and move on with time
- Types:
    - Sharp objects:
        - The majority pass without problems
        - If asymptomatic: need careful observation; it is better to admit them and observe the course of the foreign body within 24-48 hours in order to deal with any complications

        - Symptomatic: need surgical intervention in cases of:
            - Peritonitis
            - Bleeding
            - Obstruction: mainly at the ileo-cecal valve or ligament of Trietiz
            - Failure to progress: mainly stay s in the stomach. If it was a sharp object, we can wait 1-2 weeks, if it didn't pass on its own we remove it either by endoscopy or through open surgery.
            - Main sites of problems:
                - Pylorus
                - Duodenal loop
                - Duodeno-jejunal junction
                - Ileocecal valve
    - Blunt objects:
        - The majority pass without problems
        - If asymptomatic: can be observed at home for any changes (vomiting, abdominal pain, etc…) and stool must observed. If it didn't pass, the child should have an X-ray
        - If symptomatic: need surgical intervention if:
            - Peritonitis
            - Bleeding: hematemesis, melena or fresh blood in stool
            - Perforation: peritonela irritation, guarding, rigidity, and tenderness
            - Obstruction: colicky abdominal pain, vomiting, and distention
    - Disk batteries
        - Cause serious complications due to:
            - Corrosive injury due to alkali, especially if localized
            - Electrical current, if they are new
        - Should be removed if they do not progress within 6 hours (they are usually removed right away):
            - If it was in the esophagus, removed immediately
            - If it was below the diaphragm, X-ray should be done every 6 hours. If it is still in place, it should be removed.
- Laxatives are contraindicated especially in case of sharp objects, because peristaltic activity will increase. This will increase the risk of complications. In case of disk batteries, sometimes, we give them in order to pass the battery as soon as possible

Obstructive jaundice

- Biliary atersia
    - It is an obliterative process of the extra hepatic biliary tree. It is rarely seen in premature babies, so the insult comes late during gestation or early on during the neonatal period. If we take a biopsy of the biliary tree after birth, there will be obliteration to a certain degree. However, if we leave it for a month or two, the obliteration will continue. This means that the insult is there and the disease is progressive
    - Biliary atersia should be diagnosed very early. Time is very previous in those babies, and they should be treated as soon as possible.
    - Incidence: 1/10,000 live births. This disease is more common in females
    - Etiology: there is no proof that there is a single causative factor. Infectious agents, especially rhinovirus, are the most accepted etiological agents. The theory states that this virus insults the extra hepatic biliary ducts and later on it will continue to intra hepatic biliary tree. This will cause damage to these ducts leading to progressive obliteration.
    - Types:
        - Surgically correctable biliary atresia: the problem in biliary atresia is the absence of the extra hepatic biliary tree because it will turn into a fibrous cord. Hepatic ducts, cystic duct, common hepatic and common bile duct are all fibrous cords. In this type, the ducts are still patent inside the liver, so the goal here is to reconnect the duct to the intestine. This will allow for the bile synthesized in the liver to be drained into the intestines. In surgically correctable biliary atresia, the intra hepatic biliary tree is normal or dilated. Cysts replace the whle extra hepatic biliary tree
        - Surgically non-correctable biliary atresia: the ducts in the liver are atretic, but there are small ductules that can be rejoined to the small intestines.
    - Presentation: in general, those babies are well at birth. They have normal birth weight. They are healthy with a picture of obstructive jaundice. Obstructive jaundice means a yellow discoloration of the skin and sclera, dark urine, and clay colored stool. If the presentation was late, the baby might have hepatomegaly and splenomegaly due to biliary cirrhosis and portal hypertension. In this case, the patient will have all signs and symptoms of obstructive jaundice in to signs and symptoms of portal hypertension. There are two types of jaundice:
        - Embryonal type of biliary atresia appears very early
        - Neonatal type of biliary ateresia that appears after the second or third month of age
    - Diagnosis:

        - Direct hyper bilirubinemia: we depend on direct bilirubin. If it is >20-50% of total bilirubin, it is positive for hyperbilirubinemia
        - Alkaline phosphatase: elevated
        - Trans-aminases: early in the disease, their level is normal. This helps us to differentiate from neonatal hepatitis in which the trans-aminases are always high.
    - Histological findings: if we take a biopsy early in the disease, the intra hepatic biliary tree will be patent. However, if we take serial biopsies, or if the biopsy was taken during a later stage, there will be obliteration in the intra hepatic biliary tree. This means that this disease is progressive. It is mostly likely acquired, and if we interrupt the process using surgery, these patients would survive longer. Previously, most of these patients died before the age of 2, but nowadays, they can live up to 10 years and there are some reported cases that lived for 30 years.
    - Investigations: biliary atresia is one of many differential diagnoses of obstructive jaundice. Other diseases include metabolic diseases, nutritional deficiencies, sepsis, and hypothyroidism. Liver function tests will be elevated (elevated bilirubin, normal trans-aminases, elevated alkaline phosphatase, elevated GGT. CBC will show the general condition of the patient rather than diagnosing biliary atresia.
    - Diagnostic investigations:
        - Ultrasound: we look for dilated extra or intra hepatic biliary tree or look at the gallbladder. If we see the gallbladder using ultrasound, we feed the baby and look for a contraction. If there is a contraction, this means that the gallbladder is functioning and excludes biliary atresia. If we see a dilated biliary tree, we have to think of something other than biliary atresia because we do not expect to see any bile ducts. They are replaced by fibrous cords. Ultrasounds can exclude the diagnosis of other conditions like coledochal cysts; here the dilation of the biliary tree is apparent on ultrasound. Ultrasound is very accurate and non-invasive
        - Nuclear scan: it depends on certain materials that are taken up by the liver and secreted with bile to the intestines. It is called a hepato-biliary scan. A technicium labeled material is taken up by the liver and excreted with bile ot the intestine. In patients with biliary atresia, there is a total obstruction. When we give them this material, will be concentrated in the liver. It is seen by the gamma camera.
        - MRCP: a similar modality to ERCP. Through ERCP, once can see the opening of the common bile duct and canulate it; then, inject contrast material to take a picture. ERCP is difficult to perform in patients 2-3 weeks of age, so the alternative is MRCP> the concept in MRCP is similar to hepato biliray scan. If the biliary pathways are patent, we can see the

biliary tree. In biliary atresia, the pathways are absent. Its accuracy is 50-60%.

- Treatment:
  - Medical treatment: there is no effective medical treatment. In the past, they used to give a carbohydrate rich diet in addition to drugs that stimulate liver enzyme like phenobarbital. This type of treatment is no longer accepted as it did not show any benefit
  - Surgical treatment: removing all the extrahepatic biliary tree up to porta heptis. Then, a loop of small intestines is connected with porta hepatis. This is called "porto enterostomy"; it is also known as the Kasai procedure. After the Kasai procedure, 50% of patients live 5 years and 25% live for 10 years. There are some reported cases for patients who lived up to the age of 33. In the past, 90% of patients died before the age of 1, and all were dead by the age of two. The most common complication of this procedure is cholangitis. More than 50% of survivors will develop cholangitis. Those who survive cholangitis will develop cirrhosis and portal hypertension. Moreover, there is a risk for hepatobiliary CA.
  - Liver transplant: theoretically, it should be a cure for the disease. However, we don't start with this treatment because the number of centers specialized in neonatal transplants is very small. Moreover, it is major proceure involving two people. The donor can be a live donor or cadaveric donor. The most common sort of transplants are cadaveric transplants. The problems with transplant are: the availability of livers, availability of centers, and matching criteria. To perform a liver transplant, there must be certain indications:
    - Recurrent attacks of cholangitis after Kasai operation
    - Nutritional failure after the operation
    - Progressive liver damage: this procedure should be done at the age of 2-3 months. If the patient is more than 120 days old, many surgeons would not even attempt doing this procedure.

- Acute acalculus cholecystitis:
  - Like adults, it is seen in critically ill patient. It is part of sepsis, MOFS, and is seen in NICU neonates.
  - Treatment: conservative management by draining the gallbladder. The gallbladder is drained to avoid the risk of gallbladder empyema. If patients survive, cholecystectomy is performed when they are stable.
- Calculus cholecystitis:
  - Although it is not common in children, some children can develop gallbladder stones. The most common type of stones in pediatric patients are cholesterol stones. Most cholesterol stones are related to secondary disease process. This

process can include TPN, resection of terminal ileum in cases of enterocolitis or volvulus causing interruption of the enterohepatic recirculation of bile.
    - Pre and around adolescence, with the use of contraceptive pills and increased obesity, they can get the same type of gallbladder stones as adults.
    - Children between 1-5 years of age have more pigmented stones secondary to hemolysis
    - Treatment is cholecystectomy.
    - Clinical case: a patient with a picture of obstructive jaundice, thw ehole work-up was going with biliary atresia. The last step was doing laporotomy with an intrahepatic cholangiogram. During the laporotomy, the bile sample that was taken was greenish in color. This means that the hepatic ducts are patent. By cholgiography, the biliary tree was seen. However, it was not normal. The caliber of the ducts was very small, and the radicals were very few.
        - This is called hypoplasia of the biliary tree. The ducts are smaller in size and the radicals are less. Some patients present as part of Algile syndrome, while other are non-syndromatic.
- Choledochal cyst:
    - They consist of a cystic dilation of the extra-hepatic biliary tree, intra-hepatic biliary tree or both depending on the type.
    - Types:
        - Type I: it consists of uniform dilation of the common bile duct and the gallbladder is hanging on it. It is the commonest
        - Type II: it consists of diverticulum originating from the common bile duct.
        - Type III: it is also called choledochocele. Dilation of the terminal part of the common bile duct as it enters the duodenum.
        - Type IV: the entire extra hepatic biliary tree is dilated.
        - Type V: cystic dilation of the intra-hepatic biliary tree. It is called Caroli's disease, whi is one of the very difficult diseases to treat.
    - Clinical presentation: obstructive jaundice or cholangitis. In adults, the presentation ist he triad of obstructive jaundice, abdominal mass, and biliary colic.. this triad is not diagnostic to choledochal cyst, but it will help us to think about it.
    - Investigations: obstructive jaundice work up: ultrasound, MRCP, MRI, nuclear scan, etc…
    - Treatment: it depends on the type. In the majority of cases the cyst must be excised; however, there are some exceptions. These cysts are abnormal csts; if they were left, they may predispose to malignancy:
        - Type I: the whole common bile duct is abnormal. Roux-en-Y hepato-jejunostomy
        - Type II: simple to treat; excision of the cyst

- Type III: if the diameter is less than 3 cm, we leave them. This also applies to adults, because we can see this type in adults. If the diameter is more than 3 cm, the cysts must be excised
- Type IV: excision of the entire extrahepatic biliary tree and anastamosis of the bowel with the porta hepatis.
- Type V: if the dilation was localized to one lobe or segment, then we remove it. Otherwise, the treatment is liver transplant

Imperforate anus

- Anorectal malformation
- Incidence:
    - 1/1500-5000 live births.
    - Males are slightly more affected than females.
    - There is no racial or familial predilection
- Embryology:
    - Malformation in the development of the anorectal region between the 4th week and 6th month of development
    - Formation of the rectum and upper anal canal:
        - Cloaca(endoderm): it is divided by the urorectal septum into the anterior urogenital sinus (US &. upper urethra) and the posterior intestinal
        - The lateral ingrowth (folds of Rathke) complete the cleavage
        - In the females, the Mullerian ducts (forming the fallopian tubes, uterus, and upper vagina) interpose between the UG sinus and the rectum.
        - Therefore, the recto-urinary fistula rarely occurs in females except in cases of cloacal malformation or abnormal Mullerian duplications.
    - Lower anal development:
        - Anal development begins with the formation of eminences, the anal tubercles, in front of the tail fold
        - The central depression in the tubercles forms the proctoderm
        - Therefore, the lower anal canal develops from the anal tubercles and the proctoderm.
    - Perineal development
        - Develops anterior to the anal tubercles
        - In the male, the inner genital folds form the bulbus urethra and perineal body. However, outer genital folds form the scrotum and median raphe.
        - In the female, the inner genital folds form the labia minora and forchette. However, the outer genital folds form labia majora.
        - Thus, the posterior and lateral walls of the rectum originate from the inner Cloaca above. The anal pit forms from the external genital folds and tubercles
        - The anterior wall of the rectum comes from five components in the female and six in the male. These include the hindgut, urorectal septum, lateral infolds of Rathke, inner and outer genital folds, and the perineal mound (body).
- Associated anomalies:
    - Present is 25-75% of cases
    - They are present more with the high imperforate anus subtype

    - Genitourinary: renal agenesis, hypospadias, epispadias, renal dysplasia, bladder extrophy, cryptorchidism, urethral valves, reflux, ureteral obstruction, uterine and vaginal duplications.
    - Gastrointestinal: esophageal atresia, tracheo-esophageal fistula, bowel atresia, malrotation, omphalocele, annular pancreas
    - VATER syndrome
    - Skeletal: sacral agenesis, hemi-vertebra, spina bifida, and hip dislocation
    - Cardiovascular: congenital heart defects
    - perineal: hemangioma, lymphangioma, and lipoma.
    - Miscellaneou: Down's syndrome, cleft-lip /palate, meningomyelocele.
- Classification:
    - Males: 50% high type; 50% low type
    - Females: 10% high type; 90% low type
    - To classify an imperforate anus into high or low, look at the pubo-coccygeal line (PC line). If the lesion is high, it lies above this line. It might affect the continence mechanism. However, if the lesion is below this line, it is of the low type. It affects the terminal end of the anal canal and has a good prognosis.
    - The high type correlates to rectal agenesis, while the low type correlates to anal agenesis.
    - There are some intermediate types present
    - The most common type in males is the rectobulbar, while the most common type in females is the rectovestibular type.
- Diagnosis:
    - History and physical examination
        - Pregnancy history: polyhydraminos (more with high type)
        - Perianal inspection
        - Description of the normal anal location, normal hymen, and normal urethra. One of the most commonly found problems is an anteriorly located anal opening.
        - Description of the perineum; look for meconium through the fistula (epithelial pearls) at 24-48 hours.
        - Description of the number of orifices, presence or absence of anal, vaginal or urethral orifices.
        - Meconium or air per urethra or vagina
        - Associated anomalies
    - Radiography:
        - Wagenstein-rice invertogram: used to determine the level of the terminal bowel in related to the anal pit and levator sling. It has been replaced by the prone jackknife position at 24-36 hours of age.

        - The PC line is drawn from the posterior portion of the pubis symphesis to the junction of the sacrum and coccyx. In the neonate, the PC line correlates with the levator sling. Thus, if the terminal bowel is above the PC line, the lesion is high. If the terminal bowel is below the PC line, the lesion is intermediate or low.
        - The invertogram may show air in the bladder in cases of a recto-urinary fistula
        - False information on the invertogram can be due to:
            - Contracted levator
            - Valsalva maneuver
            - Fistula that allows air entry
        - Other radiographic investigations:
            - Injection with a dye or contrast
            - Chest X-ray
            - Spine X-ray
            - Renal ultrasound
            - Vaginoscopy
            - Perineoscopy
            - Cystoscopy
            - Colostogram
- Treatment:
    - Medical: resuscitation of the neonate for fluid and electrolytes like any case of intestinal obstruction.
    - Surgical:
        - Anoplasty: one stage procedure done in the neonatal period. Successful for the low type
        - Colostomy: used for the high, the doubtful, and intermediate types. It is done in the neonatal period followed by a definitive procedure called the pull-through procedure. The pull-through can be performed in many different ways, the most famous of which is called PSARP (posterior sagittal anorectoplasty)
- Results and prognosis:
    - Mortality and morbidity are directed by the associated anomalies rather than the anomaly itself.
    - The functional results of a low lesion repair in both sexes are excellent.
    - In the case of high lesions and complex cases, the prognosis is related to the degree of sacral development, perineal, and pelvic muscle development.

Neck swellings

- Neck lesions and inguinoscrotal disorders represent more than 75% of cases of pediatric surgery
- Neck swellings include:
    - Cervical lymphadenopathy:
        - The most common and most important neck mass in childhood
        - Usually comes due to lymph node enlargement, which can be due to:
            - Infection: enlargement of deep cervical, superficial cervical, submental, or submandibular glands.
                - Infections of the ear, tonsils, pharynx and presence of bad teeth causes enlargement of the draining lymph nodes.
            - Acute submandibular suppurative adenitis:
                - Formation of an abscess on top of a case of simple adenitis. If the treatment of simple adenitis was delayed or the patient was given the wrong choice of antibiotics, an abscess will form. When the abscess forms, it should be treated with incision and draining.
                - Signs of abscess formation include swelling, redness, tenderness, and indurations
            - Hodgkin's lymphoma:
                - Usually occurs during late childhood and adolescence
                - Characterized by non-tender, progressively enlarging, and matted lymph nodes.
        - The cases of lymph node enlargement can be either benign or malignant. Tender lymph nodes are usually infected. Hard lymph nodes imply higher risks of malignancy.
        - Treatment and investigations:
            - Treatment of the underlying cause
            - In adults, you have to investigate lymph node enlargement very carefully because of the higher risk of malignancy.
            - Investigate using trioscopy (bronchoscopy, esophagoscopy, and laryngeoscopy). If these are negative, take a biopsy and search for a tumor anywhere in the body.
            - Solitary, non-tender, mobile, soft, and enlarged lymph nodes in a child (in the absence of an infection) are considered benign ones. The presence of the opposite criteria is suggestive of malignancy and prompts further investigations using a biopsy.
    - Torticollis: tilted neck:
        - Causes:

        - Congenital: due to abnormal fetal position in the uterus. It is a form of trauma that causes fibrosis (inelastic tissue) of part of the neck muscles (sternocleidomastoid). The function of this muscle is the approximation of the mastoid process to the sternum. Fibrosis of the muscles causes its shortening. This might lead to the formation of a prominent mass on top of the muscle
        - Acquired:
            - Trauma that leads to cervical injury, which leads to cervical spasm on one side
            - Fibrosis of the SCM muscle due to any cause
        - Infection: lymphadenitis in that area causes muscle spasm. This muscle spasm is needed to protect the area from pain caused by movement.
    - Presentation:
        - Torticollis occurs at any age; however, it commonly presents in the first few months of life
        - You may see a palpable hard mass that is 1-4 cm in diameter in 1/3 of the patients
        - The baby with torticollis usually sleeps on the same side; this lead to a craniofcial deformity.
    - Treatment:
        - Conservative: in 85% of patients, using physiotherapy for 2-3 months gives excellent results.
        - Surgical: if there is no improvement or if the child develops a craniofacial deformity, surgery is indicated. We cut the short SCM to free it.
- Thyroglossal duct cyst:
    - It is the most common congenital anterior neck mass. It located at the midline.
    - Differential diagnosis:
        - Lymph node enlargement
        - Abnormally located thyroid gland
    - Most of thyroglossal duct cysts present during the first two decades of life.
    - Pathophysiology: the embryological thyroid gland is formed at the level of foramen cecum. Then, it descends anterior or posterior to the hyoid bone until it reaches its final place anterior to the thyroid cartilage. Its tract should be obliterated with time. If this tract remains, a thyroglossal duct cyst forms.
    - Males are affected more than females.
    - Its epithelial lining is cuboidal to columnar or pseudostratified.

        - Diagnosis:
            - Physical examination: as the patient to open his moth. The, protrude the tongue. The cyst will move upward upon tongue protrusion.
            - Ultrasound: appears as a cystic lesion
            - Thyroid scan: controversial. It is used to detect whether this cyst is the only functional thyroid tissue present in the body. If the case is so, some physicians debate that the operation should be withheld. However, the majority of surgeons agree that this tissue is abnormal tissue and should be removed no matter what (higher risk for malignancy)
        - Treatment:
            - Excision along its tract. In many cases, the hyoid bone must be resected (the track passes through it)
            - If you don't remove the hyoid bone, the recurrence rate is 50-60%; however, if it was removed, the recurrence rate drops to less than 10%.
            - Recurrence is due to the fact that some of the tract cells can be found within the hyoid bone. If they were not removed, they can multiply and form a new tract.
            - The thyroglossal duct cyst is removed to prevent infection and abscess formation.
            - The idea treatment regimen is administration of antibiotics followed by incision and drainage, then surgery.
    - Branchial anomalies (cysts, sinuses, or fistulas)
        - Most of them arise from the second branchial cleft
        - Branchial sinuses in the second cleft have a small cutaneous opening between the lower third and mid third of the anterior border of the SCM. Second branchil cysts present usually at the junction between the upper third and middle third of the anterior border of the SCM.
        - A fistula is a tract connecting two epithelial surfaces. In this case, the internal opening is in the tonsillar fossa and the externa opening is on the cutaneous surface. A fistula presents as drained saliva from the baby's neck. It is managed by excision of the whole tract with careful care to the carotid artery and adjacent nerves.
        - First branchial cleft anomalies: their external opening is located at the angle of mandible and by definition they communicate with the external auditory meatus. The preauricular sinus was thought to arise from the first branchial apparatus, but now it is well known that this is completely different. It is caused by abnormal fusion of ear pinna during its formation.

A preauricular sinus, if followed, will end in the auricle not in the external auditory meatus. Treated by excision.

- 3rd branchial cleft anomalies are like those of the second.

- Cystic hygroma:
  - Rare
  - It is problem with high recurrence rate after treatment
  - It is a multi-lobular cystic structure that results due to abnormal fusion of the lymphatic system with the venous system. It results in dilation of lymphatics at the junction with the venous system.
  - Sometimes, it involves the whole neck and anterior mediastinum until the retro-peritoneum; it can even reach to the scapula. It is rare condition that interferes with respiratory airways.
  - It can be diagnosed in utero, and once delivery occurs, it must be treated in special centers.
  - Clinical diagnosis: usually located in the posterior triangle of the neck. It may present in any lymph rich region like the groins and axilla. It transilluminates like hydroceles.
  - Differential diagnosis:
    - Hemangioma
    - Encephalocele
  - Management:
    - A cystic hygroma that interferes with breathing should be treated at special centers
    - Other cases are treated by:
      - Repeated excision (if it recurs)
      - It was noticed that a cystic hygroma that gets infected can resolve spontaneously. Based on this idea, they extracted a substance from streptococci and injected it into the cyst. The substance works like a sclerosing agent, and it can be used to stimulate fibrosis and obliteration of the cystic hygroma

Gastro-esophageal reflux disease in children

- GER is defined as retrograde passage of gastric content to the esophagus due to incompetence of the lower esophageal sphincter. There is no demonstrable histological or anatomical sphincter mechanism; however, this area exhibits higher pressures than the rest of esophagus and stomach. This acts as a physiological sphincter
- Gastric contents cause damage to the lower esophagus; this damage might lead to many complications:
    - Stretch
    - Bleeding
    - Metaplasia in the epithelium (squamous to columnar). This is called Barrett esophagus, and it is a premalignant condition.
- In adults, GER is always pathological; however, in children, it can be classified into:
    - Functional (physiological): during the first six months of life the sphincter mechanism is not mature, which may lead to passive regurgitation. Symptoms:
        - Asymptomatic vomiting: no bile or blood
        - Effortless regurgitation
        - No predisposing illness (neurological or GI illness)
        - Improves with age without treatment: 47% in the second month of life had GER, but at 6 months this percentage drops to 4%
        - No complications or growth retardation
    - Pathological: usually after the first 4^{th} month of life:
        - Vomiting
        - Recurrent chest infections:
            - Pneumonia
            - Asthma like picture due to reflex spasm (wheezing, stridor, and apnea)
            - Nocturnal coughing
            - These symptoms lead to growth retardation and failure to thrive
        - Predisposing illness:
            - Cerebral palsy
            - Down syndrome
            - Head injury
            - Developmental delay
- Incidence:
    - Physiological: 85% of infants vomit during the first month of life, so we can consider itt as a part of normal physiology
    - Pathological: common in infants with a reported incidence less than 10%

- Pathogenesis of GER:
    - Protective mechanism:
        - Sphincter mechanism
        - Intra-abdominal esophagus: the intra-abdominal pressure is higher than the intra-thoracic pressure, so the intraabdominal segment of the esophagus will be exposed to higher pressures, and will be closed. In cases of congenital short esophagus or hiatus hernia, there is no intra-abdominal segment. The incidence of GER is higher in these patients. The length of this segment changes with age, height, and weight. In children, in order for it to effective, it should be 1.5-2 cm long. After surgery, we have to be sure that the intra-abdominal segment is at least 1.5 cm long.
        - Angle of His: it is the angle between the esophagus and the fundus of the stomach. In children, this angle is obtuse, and gastric content can easily regurgitate to the esophagus.
        - High pressure zone: at the lower end of the esophagus, we can see that the pressure is very high (5-10 mmHg). This is higher than the rest of the esophagus
    - Attack mechanism (opening mechanism)
        - When there is an increase in gastric pressure due to gastric contraction or or an increase in the volume of the gastric content with slow emptying, it will lead to regurgitation of gastric contents.
        - Lack of coordination in the contraction of the stomach
        - Transient LES relaxation: normally, after eating or during respiration, the stomach will be filled with. This will lead to LES relation, which lasts for 5-10 seconds. In cases of GER, it will relax for more than 10 seconds
        - Increase in the intra-abdominal pressure: usually, it is protective against reflux; however, it can increase intra-gastric pressure (in cases of cerebral palsy, there will be abdominal contraction, which increases the intra-abdominal pressure). This factor, on its own, is not enough to cause reflux.
        - Esophageal motility
        - Supine position: in this position, the LES allows gastric content to regurgitate easily in to the esophagus. In the prone position, the junction will be higher than the rest of the stomach. This decreases chances of reflux
        - Gastrostomy: leads to a change in the angle of His; the incidence of reflux will increase. Gastrostomy is contraindicated in cases of GER.
        - Drugs: gastrin and dopamine increase the risk of reflux
- Symptoms:
    - GI: heart burn, vomiting, regurgitation, loss of appetite, weight loss
    - Respiratory: apnea, wheezes, stridor, pharyngitis, otitis media

    - Atypical syndrome:
        - Excessive crying
        - Sandifer syndrome: abnormal head posture. Tilting of the head to reduce reflux symptoms. When the reflux is treated, the patient goes back to the normal posture.
- Investigations:
    - Barium swallow and barium meal:
        - Barium goes down through the esophagus. After that, it will appear again due to regurgitation.
        - Barium meal is not diagnostic because GER is episodic, and we may test it when there is no reflux. This means that a negative test doesn't exclude reflux
        - This test is a dynamic test, so we have to take more than one view using X-rays at different times to diagnose the reflux
        - This is used to diagnose a hiatal hernia, as well.
    - pH monitoring:
        - very sensitive and specific for GER. A probe is placed for 24 hours; if there is a reflux it will be detected. This is why it is sensitive.
        - Accuracy: 95-98%
        - Acidity that lasts more than 5 minutes can cause injury to the esophageal wall (esophagitis). However, if the acidity lasts for 4 and a half minutes, there will be no significant damage. Using pH monitoring, we can see for how long acid accumulates in the esophagus.
        - Using this test we can measure the pH during the time of the attack.
    - Manometry: mostly a research tool; not diagnostic
    - Endoscopy: very important, especially in resistant cases. Through endoscopy, we can see if there are any complications such as esophagitis and strictures. Moreover, we can take a biopsy to look for certain organisms
- Management:
    - Conservative:
        - Burp position after feeding
        - Elevation of the head of the bed
        - Prone position: sleeping in this position can cause sudden infant death. This position can decrease reflux; however, we do not adivse using it.
    - Medications:
        - Prokinetic agents: metaclopramide and cesapride. These agents iprove motility of esophagus and increase the tone of the sphincter. Cesapride has been withdrawn from the market because it causes cardiac complications, especially in those with an existing cardiac anomaly.
        - Acid suppression: H_2 blockers and proton pump inhibitors (PPI)

    - Indications for surgery:
        - Resistant cases or the presence of complications
        - Fundoplication is the best surgical treatment for GER> the fundus is sutured partially or completely around the gastro-esophageal junction.
- Prognosis:
    - Most cases are benign.
    - 80% resolve by 18 months of age
    - Surgery is required in a minority of patients

Congenital diaphragmatic hernia

- Congenital diaphragmatic hernia
    - Bochdalek hernia – posterolateral hernia
    - Anterior hernia: Morgagni hernia: retrosternal hernia
- Bochdalek hernia:
    - Incidence: 1/2000-4000 live births. It acounts for 8% of all major congenital anomalies.
    - It constitutes a major surgical emergency in the newborns
    - Symptoms depend on the degree of herniation. Small hernias may initially pass unnoticed; larger ones produce immediate and severe respiratory distress
    - Classification:
        - The 3 basic types of CDH are the posterolateral Bochdalek hernia, the anterior Morgagni hernia, and the less common hiatal hernia
        - The left sided Bochdalek hernia is seen in approximately 90% of cases. Some people say this is due to the presence of the liver. The major problem in a Bochdalek hernia is the posterolateral defect of the diaphragm, which results in either failure of development of pleuro-epithelial folds or improper or absent diaphragmatic musculature migration. Bilateral Bochdalek hernias are rare.
        - The Morgagni hernia is a less common form of CDH. It occurs in 5-10% of the cases of CDH. This hernia occurs at the anterior midline through the sternocostal hiatus of the diaphragm. 90% of the cases occur on the right side. It doesn't interfere with the development of the lungs, and it is often and incidental finding.
        - A congenital hiatus hernia is very rare in neonates. In this form, the stomach herniates through the esophageal hiatus.
    - Diagnosis:
        - In the prenatal period, ultrasonography has high sensitivity. Bowel loops may be seen to undergo peristalsis in the chest
        - In neonatal and infantile periods, one cannot overstate the importance of obtaining a chest radiograph at the first sign of distress in these infants. This image usually permits for an accurate diagnosis. Typically, no bowel gas is evident in the abdomen. Cystic malformation of the lung might look like CDH on an X-ray without contrast. Obtain a contrasted chest X-ray in order to be able to tell the difference between a cystic malformation and bowel hernia.
        - In patients who present in the prenatal period, ultrasonographic features of CDH include: polyhydraminos, absent or intrathoracic stomach bubble, a

mediastinal and cardiac shift away from the side of the herniation, and, rarely, hyrops fetalis.
        - There are some anomalies associated CDH; however, they are usually severe and fatal. Only those patients with an isolated CDH have a chance to live (hidden mortality).
    - Clinical features:
        - Apparent dextrocardia
        - Barrel chest
        - Scaphoid abdomen
        - Respiratory distress or cyanosis on clamping the cord during delivery.
    - Complications of CDH:
        - The main problem lies in the presence of pulmonary hypoplasia, which may be unilateral or bilateral. This hypoplasia my result in persistent fetal circulation
        - Pulmonary hypoplasis is thought to result from the longstanding intrauterine (embryonic) compression of the lungs by the hernia. Mortality in babies with CDH is largely confined to those with bilateral pulmonary hypoplasia, but hypoplasia is always more severe in the lung ipsilateral to the hernia. The pulmonary vasculature is also affected to a greater degree than the bronchial tree
        - Infants with the largest and longest standing hernias have the most hypoplastic lungs and are less likely to survive after birth. If a diaphragmatic hernia develops towards the end of a pregnancy or after birth, pulmonary hypoplasia does not occur
    - Differential diagnosis:
        - Cystic adenomatoid malformation
        - Pneumotocele
        - Mediastinal cyst (bronchiogenic, neuro-enteric, or thymic)
        - Neonatal respiratory distress conditions, RDS, mechonium aspiration, and choanal atresia
    - Treatment:
        - Surgery: it is complicated and the mortality rate is very high. Many approaches have been tries including intrauterine approaches. This was associated with some success; however, it came over the expense of a premature birth.
    - Prognosis
        - Mortality in babies with CDH is related to pulmonary hypoplasia, persistent fetal circulation, and associated anomalies.
        - The reported mortality rate is 25-60%; however, the rate is difficult to determine accurately due to hidden mortality. For this condition, hidden

mortality refers to the death of infants who are severely affected that they die prior to their transfer to surgical unit

- The identification of the stomach within the chest is correlated with a poorer outcome as a result of herniation at an earlier stage of gestation. This results in a greater degree of pulmonary hypoplasia. Poor outcomes have also been associated with pneumothorax (reflects pulmonary hypoplasia) and a right sided defect.
- Favorable features include the presence of an aerated ipsilateral lung and aearation of more than 50% in the contralateral lung.

Vesicoureteral reflux

- Retrograde flow of urine from the bladder into the ureter.
- Classification:
    - Primary reflux: VUR in an otherwise normally functioning lower urinary tract. Treatment is easier and the outcome is better
    - Secondary: associated with or caused by an obstructed or poorly functioning lower urinary tract (posterior urethral valve or a neurogenic bladder)
- Pathophysiology:
    - The valve mechanism depends on the length of the submucosal tunnel of the distal ureter or its muscular backing
    - The ratio of tunnel length to ureteral diameter must be at least 5:1 to prevent reflux
    - This fundamental observation is the basis of almost all surgical procedures to correct this disorder
- Frequency (incidence)
    - 1-2% of general population
    - 15-70% among children with a UTI (depending on age). The risk increases in younger age groups
    - One third of cases that are identified antenatally tend to be hydronephrosis on ultrasound.
    - Siblings of children with VUR have a 25-33% risk of having a VUR
    - Offspring of parents with a reflux have a 66% incidence. Higher in female offspring.
- Sex predilection:
    - Among all children with UTI, males are more likely to have a VUR (74% of males vs. 29% of males). However, in general, it is more common in females because UTIs are more common in females.
- Age:
    - VUR is more common among infants and resolves progressively with age (increase in tunnel length with age):
        - 70% < 1 year
        - 25% at 4 years
        - 15% at 12 years
        - 5.2% in adults patients presenting with a UTI
- Clinical presentation/history:
    - Most children with VUR present in one two distinct groups:
        - Antenatal hydronephrosis
        - Clinical UTI
- Physical signs:

    - Fever
    - Flank or abdominal tenderness
    - Palpable kidney in cases of hydronephrosis
- Predisposing factors:
    - The case of the defect in primary reflux is unknown
    - Genetic component
    - Congenital abnormality of urinary bladder
- Work up:
    - Diagnosis of UTI is dependent on obtaining an accurate urine culture:
        - Suprapubic aspiration: any growth in such a sample is significant
        - Midstream voided specimen: growth of >100,000 colony forming units is a significant finding
        - Urethral catheterization provides a substantially better specificity: >1000 colony forming units is significant
        - The least reliable method is bag collection (should not be used)
    - Imaging studies:
        - The standard imaging tests are the ultrasound and the voiding cystourethrogram (VCUG)
        - Imaging is indicated after the first UTI in all children
        - Children with antenal hydronephrosis should evaluated postnatally
        - The standard in diagnosis of VUR is VCUG. This test provides precise anatomic findings and allows for grading of the reflux
- The international grading system for VUR:
    - Grade I: reflux into a non-dilated ureter
    - Grade II: reflux into renal pelvis and calyces without dilation
    - Grade III: reflux into a mildly to moderately dilated ureter and pelvis
    - Grade IV: dilated tortuous ureter, dilated calyces and persistent papillary projections
    - Grade V: severe dilation and tortuousity, blunting of the fornices, intrarenal reflux (the reflux extends to the cortex, so you can see the cortex and collecting duct on the VCUG).
- Detailed discussion of investigations:
    - VCUG:
        - Should be taken after the child has fully recovered from the UTI (2-3 weeks after recovery)
        - Performance of the test during an episode of acute cystitis can result in:
            - Overestimation of the grade due to peristalsis and laxity of the ureteral musculature
            - False results: due to edema around the ureter which does not show the reflux well

        - This test can detect a neurogenic bladder and a posterourethral valve.
    - Nuclear cystogram:
        - Done by administration of Technicium 99 into the bladder
        - Less accurate than VCUG
        - Radiation is lower than that used inVCUG (less invasive)
        - Can be used for screening
    - Ultrasound cystogram:
        - The incorporation of doppler technology into ultasound units is used as a radiation free screening tool for VUR
        - These studies look reversal flow in the ureter during filling of the bladder
    - DMSA scan: to detect if there is kidney damage
    - Urodynamic study: reveals functional abnormalities of the lower urinary tract (neurogenic bladder, for example)
- Treatment:
    - Prevent complications such as kidney infection or damage
    - Two methods (surgical and medical):
- General management principles:
    - Spontaneous resolution of VUR is common in young children; however, it decreases as puberty approaches.
    - Severe reflux is unlikely to resolve on its own
    - Sterile reflux does not result in reflux nephropathy
    - Long term antibiotic prophylaxis in children is safe
    - Surgery to correct this condition is highly successful.
- Medical treatment:
    - Supportive care and prompt administration of the proper antibiotic
    - Antibiotic prophylaxis: those dose is 1/9 of the therapeutic dose
    - Antibiotics are continued as long as there is a reflux
- Indications for surgical treatment:
    - Breakthrough febrile UTI despite adequate antibiotic prophylaxis
    - Severe reflux (grade IV and V) with renal scarring
    - Mild or moderate reflux in females the persists as the patients approaches pubery despite several years of observation.
    - Poor renal growth or appearance of new scars
    - Poor compliance to medications
- How is the procedure done?
    - Mobilization of the ureter
    - Elongation of the intramural segment
    - Implantation of the ureter in a site different from its primary one.

Neuroblastoma

- Site of origin:
    - Adrenals (abdomen): 50%
    - Paraspinal (abdomen): 24%
    - Mediastinum: 20%
    - Cervical: 45
    - Pelvic: 2%
- The most common tumor in children 1 year of age
- The third most common childhood tumor after leukemia and brain tumors
- The most common spontaneously regressing or maturing tumor
- It is an embryonal tumor
- Associated with other neurocrestopathies:
    - Beckwithh-Wiedmann syndrome
    - Hirschsprung's disease
    - Fetal alcohol syndrome
    - Van Waardenburg syndrome
- Clinical features:
    - The most common presentation is either antental ultrasound finding or an asymptomatic abdominal mass that is fixed, painful, and crosses the midline.
    - Has a unique immunobiology
    - Most children present with advanced disease
    - Hepatomegaly, subcutaneous nodules, orbital swelling, paraplegia, anemia, and thrombocytopenia
    - Paraneoplastic syndromes, VIP, WADHA, opsocolnus myoclonus of cerebellar ataxia (dancing eye syndrome) due to cerebellar involvement with Antigen-antibody complex rather than direct cerebellar involvement.
- Unusual clinical features:
    - Dumbbell tumor: extension of the tumor through the neuroforamina
    - VIPoma like syndrome
    - Horner's syndrome
- Diagnosis:
    - Plain X-ray: microcalcifications
    - Abdominal CT
    - MRI: intra-spinal extension and osseous involvement
    - Bone survey and bone isotope scan
    - Serum ferritin, NSE, and LDH
    - Urinary VMA, HVA, and metanephrine
    - 90% of tumors are biologically active secreting CVA and HVA
    - NSE (neuron specific enolase is elevated in all patients with metastases

    - Genetic abnormalities are present in 80% of the patients:
        - Deletion of the short arm of chromosome 1
        - Amplification of the n-myc oncogene
    - Triad of myoclonus opsoclonus syndrome, cerebella ataxia, and opsoconus
    - 50% of these patients have the tumor in the chest area
    - MIBG (meta-iodo-benzylguanidine) scan is the best best diagnostic and localizing test
- Stagins:
    - Stage 1: localized to the site of origin: treated by complete excision with or without residual disease and negative lymph nodes
    - Stage 2A: incomplete excision and negative lymph nodes
    - Stage 2B: complete excision of the tumor and positive lymph nodes, but negative contralateral lymph nodes.
    - Stage 3: tumor crossing the midline with or without lymph node involvement. OR a unilateral tumor with contralateral node involvement. OR midline tumor with bilateral lymph node involvement
    - Stage 4: tumor dissemination to distant nodes, bone, bone marrow, liver or other organs.
    - Stage 4S: localized primary tumor as defined for stage 1, 2A, and 2B with metastasis limited to the liver, bone marrow, or skin. Most newborn infants and 30% of those under year of age have stage 4S
- Treatment:
    - Surgery: complete excision (when possible), lymph node sampling and liver biopsy. If the tumor is unresectable, chemotherapy and radiation then surgery (second-look surgery)
    - Chemotherapy when resection is incomplete. Multiple agents are better than a single agent. Ifosfamide, carboplatin, iproplatin. Older patients have a limited response to chemotherapy.
    - Radiation: the tumor is radiosenstive; however, when the tumor is localized, radiation offers no benefit over surgery and chemotherapy. Radiation is used for stage 3 disease and older patients.
    - Bone marrrow transplant after melphalan administration and total body radiation.
- Prognosis:
    - Local tissue extension
    - Liver metastasis
    - Feasibility of excision
    - Tumor tissue DNA flow cytometry (aneuploidy, diploidy)
    - Number of copies of the amplified N-myc oncogene, trk-proto-oncogene
    - Response to monoclonal antibodies
    - Differentiation

    - Maturation
    - MKI (mitosis karryohexis index)
    - Different stage and maturation in the same patient
- Survival:
    - Stage 1: 100%
    - Stage 2A and 2B: 75%
    - Stage 3: 45%
    - Stage 4: 15%
    - Stage 4S: 70%

Wilm's tumor:

- The most common childhood renal malignancy
- Embryonal tumor (nephroblastoma)
- Nephroblatomatosis: persistent metanephric tissue beyond 36 weeks of gestation present in 25-40% of nephroblastoma patients
- Incidence:
    - 500 cases each year: 1:15,000 live births in the USA
    - No racial, environmental, or ethnic predilection
    - Blacks and hispanics are more frequently affected by the sarcomatous tumor type
    - Equal sex ratio
    - Mean age: 3.5 years
    - Syndromatic variety (10-15%) is associated with
        - WT-1 (11P13), 11P15, WT-2, 16q, 1p
        - WAGR syndrome
        - Aniridia
        - Hemihypertrophy
        - Neurofibromatosis
        - Beckwith-Widmann syndrome
            - GU anomalies, UTI, hypospadias, aniridia
- Wilm's syndromes:
    - Drash syndrome: Wilm's, pseudohermaphroditism and degenrative renal disease
    - Kippel Trenaunnay syndrome
    - Perlman syndrome
    - WAGR syndrome
    - Familial Wilm's tumor caused by deletion of chromosome 11p
- clinical manifestations:
    - asymptomatic abdominal mass
    - abdominal distention
    - microscopic hematuria (25%)
    - abdominal pain

    - polycythemia
    - hypertension that may present with heart failure
    - acute abdomen, shock (rupture)
- diagnosis:
    - history and physical examination
    - clinical features
    - abdominal ultrasound, CT, abdomen and chest MRI scan, bone survey
    - tumor staging, venal caval involvement, and contralateral kidney involvement
    - biopsy, FNA
    - linear calcifications (fine stippled calcifications in neuroblastoma)
    - propensity for venous extension
- pathology:
    - epithelial pathology: well differentiated (favorable) 85-90%
    - sarcomatus type: undifferentiated (unfavorable); 10-15%
    - clear cell: Rhabdoid anaplastic
- staging:
    - stage 1: encapsulated unilateral tumor without capsular or nodal involvement. Completely resected without tumor spill
    - stage II: unilateral tumor with involvement of the renal capsule or hilar fat, adherent to the surrounding structures including the renal vein, without lymph node involvement. Resected without tumor spill
    - stage III: unilateral tumor with regional lymph node involvement. Preoperative tumor rupture with significant intraoperative tumor spill
    - stage IV: hematogenous metastasis to lungs, bone, brain, and distant lymph nodes.
    - Stage V: bilateral tumor, synchronous or not
- Treatment:
    - Surgery: the main form of treatment. Unilateral nephroureterectomy and lymph node dissection
    - Chemotherapy: according to the stage. Most patients receive chemotherapy with different duration and number of drugs. Drugs include: Actinomycin D, Vincristin, Doxrubicin, Eroposide, and cyclophosphamide
    - Radiation: some stage II, all stage III and IV, not with stage I
    - Bilateral: 5% ; treatment is individualized
- Prognosis:
    - Prognosis depends on stage and histopathological type:
        - 90% survival with favorable histology; 54% with unfavorable histology
        - Stage I: 96%
        - Stage II: 92%
        - Stage III: 78%
        - Stage IV + unfavorable histology: 58%