Pediatric surgery

➢ CDH

- Defect in the diaphragm
- Male
- Mostly isolated
- Pentalogy of Cantrell: 1. Omphalocele 2. Lower sternal defect 3. Pericardial defect 4. Diaphragmatic hernia 5. Cardiac ectopia
- Types:

1. Bochdalek: posterolateral, most common, usually on the left side
2. Morgagni: anterior, less common, associated with trisomy 21

- Diagnosis:

- Prenatal US: polyhydramnios, intrathoracic fluid-filled bowel loops, an echogenic chest mass, mediastinal shift, intrathoracic stomach/ liver, observed to expected lung head ratio —> severe when <25%, observed to expected total fetal lung volume —> better prognosis if >35%, may have right to left shunt due to pulmonary HTN
- Postnatal CXR showing bowel loops in the chest

- Clinical features: respiratory distress at birth, scaphoid abdomen, decreased breath sounds, displaced cardiac impulse
- Management: immediate respiratory support: intubation, hypercapnic ventilation, NGT
  *if didn't work—> HFOV *if didn't work —> ECMO
  **ECMO contraindications: 1.Severe chromosomal anomalies 2.Intracranial hemorrhage 3.Preterm<34 weeks 4.Birth weight <2kg
  Surgery: FETO (risky) or post delivery when stable
- Most common cause of death is **lung hypoplasia** both lungs are affected (not rare!)

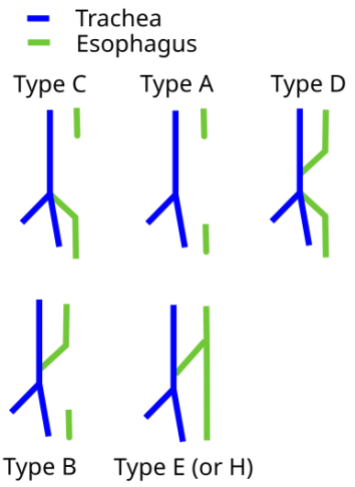
➢ Eventration

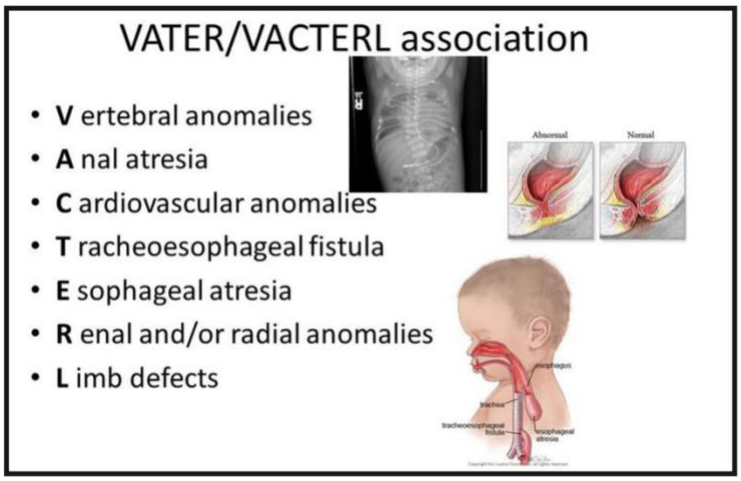
- Abnormal elevation of diaphragm
- Same presentation as CDH
- On CXR: elevated hemidiaphragm
- On US: paradoxical movement
- If severe do plication

➢ TEF

- Incomplete fusion of the lateral tracheoesophageal folds —> defective septum
- Male
- Associations: prolonged use of OCPs, progesterone and estrogen exposure, maternal DM, thalidomide exposure, fetal alcohol syndrome, maternal phenylketonuria
- Types: most common is type C (esophageal atresia with distal fistula)

- Clinical features: **excessive drooling** (most common), chocking, cyanosis, inability to pass NG tube (confirmatiry)





- Diagnosis: antenatal US: polyhydramnios, small/absent stomach bubble, pouch sign, cardiac/renal anomalies
  Postnatal CXR with an NG tube coiled in the esophagus
- Management: not an emergency, supportive care to prevent aspiration (NPO, NG, upright position and on left side, intubation if in distress) until surgical correction (fistula ligation and primary anastomosis) +- bronchoscopy
  Do Echo before surgery – associated cardiac anomalies which may be fatal

➢ Intestinal obstruction

➢ Intestinal atresia

- Part of the intestine is blocked/absent due to failure of canalization
- Male
- 50% associated with trisomy 21, 50% premature, gastroschisis association
- Mostly it's distal to the ampulla (biliary vomit)
- Types:
  1. Type 0: stenosis
  2. Type 1: incomplete obstruction by a membrane or a fenestrated web
  3. Type 2: complete obstruction -> two blinded ends connected by a fibrous cord
  4. Type 3: complete obstruction-> gap between bowel ends there is 3a and **3b (apple peel atresia)**
  5. Type 4: multiple atresias **(string of sausages)**
- Diagnosis: *antenatal US: polyhydramnios, dilated bowel loops, echogenic bowel
  *postnatal: bilious emesis within the first hour of life in an otherwise stable baby + upper abdominal distention + scaphoid abdomen + abd X-RAY: double bubble sign-> duodenal atresia, triple bubble sign-> jejunal atresia
  Contrast enema is done to rule out other differentials

- Management: resuscitation, NPO, NG, Echo, **not urgent**, duodenoduodenostomy (late complication: delayed gastric emptying)

➢ Colonic atresia

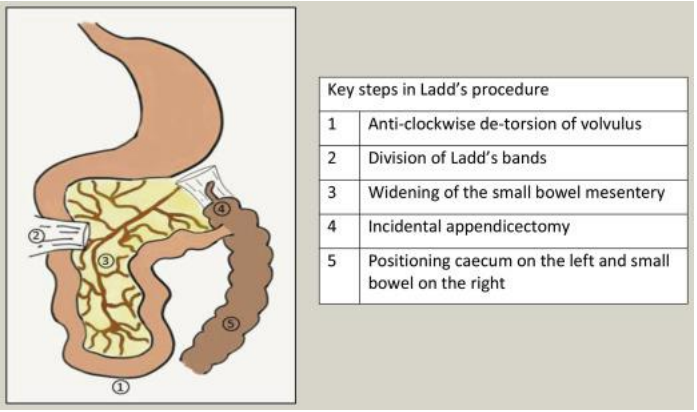
- Less common than intestinal atresia
- Unlikely to be detected on prenatal US
- 3 types: type 1 —> mucosal atresia with intact bowel wall & mesentery (mostly distal to splenic flexure)
  Type 2 —> fibrous cord between the atresic ends
  Type 3 —> most common, the atresic ends are separated by a V-shaped mesenteric gap
- Presentation: abdominal distention, bilious emesis, failure to pass meconium
- Diagnosis: abd X-ray—> dilated bowel loops, ground-glass appearance
  Contrast enema —> small diameter distal colon then abrupt halt
- Management: resection and primary anastomosis and **take a biopsy for HPD**

➢ Malrotation

- Normally, bowel rotates 270 degree counter clock wise around the SMA axis (herniation – rotation – retraction – fixation)
- A spectrum of:

1. Non rotation of midgut: failure of rotation
2. Mixed rotation & volvulus: twisting of bowel causing ischemia
3. Reversed rotation: clockwise rotation leads to abnormal position
4. Internal hernia & midgut volvulus

- Presentation: bilious vomiting in a one month year old otherwise healthy term neonate, can present at any age (infant—adulthood)
- Diagnosis: **upper contrast study** -> **corkscrew** which indicates volvulus, **beak** sign which indicates complete obstruction
  Color Doppler US -> **whirlpool** sign (twisting of mesenteric vessels) normal: SMA on left
- Management: resuscitation, NG, Abx, surgery (Ladd's procedure)
- Cecum will be in the **LUQ**



Key steps in Ladd's procedure	
1	Anti-clockwise de-torsion of volvulus
2	Division of Ladd's bands
3	Widening of the small bowel mesentery
4	Incidental appendicectomy
5	Positioning caecum on the left and small bowel on the right

➢ Anorectal malformation

- Common
- Male
- Most common type in males: rectourethral bulbar fistula
- Most common in females: rectovestibular fistula
- Cloaca: one way leads to 3 cavities (digestive, urinary, reproductive)
- Diagnosis: on clinical exam no anus seen
- Management: initially watchful waiting for 24 hrs, NPO, NG, rule out VACTERL (spinal and sacral radiographs, spinal and pelvic US, MRI spine, echo, NG tube, renal US, limbs radiographs)
  *__Double barrel__ stoma if rectum further than 1cm or meconium in urine then do a distal colostogram @ 8 weeks to prepare for PSARP
  *Primary PSARP if rectum within 1cm on cross table lateral film or meconium on perineum
  *If cloaca do renal & pelvic US as a first step
- Complications: if high malformation: fecal & urinary inconvenience
  If low malformation: constipation, recurrent fistula

➢ Hirschsprung disease

- Functional intestinal obstruction
- Mostly there is a transition zone
- Male
- Presentation in neonatal period: abdominal distention, bilious vomiting, feeding intolerance, delayed passage of meconium, Hirschsprung-associated enterocolitis
- Presentation **later** in childhood: severe chronic **constipation**
- Diagnosis: gold standard-> rectal biopsy: absent calretinin +increased acetylcholinesterase activity in affected ganglions, Anorectal manometry: absence of normal rectoanal inhibitory reflux, abd X-ray, contrast enema, **DRE: tight rectum, release of stool upon finger withdrawal**
- Management: rectal washouts if still stable do a colostomy
  Definitive is pull-through procedure

➢ Meconium ileus

- Obstruction usually at iliocecal valve
- Earliest sign of **cystic fibrosis** (family history)
- Diagnosis-> antenatal US: hyper-echoic intraabdominal mass, dilated bowel, no visualization of gallbladder
- Postnatal X-ray shows dilated bowel loops, on exam doughy, narrow anus and rectum

- Management: if simple -> water soluble contrast enema **(gastrografin)**
  If failed conservative -> enterotomy and irrigation (N-acetylcystine) followed by enterotomy closure/enterostomy tube/ileostomy
  If complicated (perforation, volvulus, gangrene) -> resection of ischemic bowel + diverting stoma/primary anastomosis
- Later complications: distal intestinal obstruction syndrome, fibrosing colonopathy

➢ Necrotizing enterocolitis

- Premature neonate
- VLBW <1 kg
- Low birth weight & younger GA are associated with higher mortality
- Presentation: feeding intolerance, abdominal distention, late sign: bleeding per rectum
- Diagnosis: abdominal X-ray -> **pneumatosis intestinalis** (donut like), labs -> high lactate, **metabolic acidosis**, low platelets, hyponatremia
- Management:NPO, NG, if stage 3b -> surgery: laparotomy with resection/ with stoma/ peritoneal drainage for (VLBW)
- 30% mortality
- NEC is the leading cause of pediatric intestinal failure

Table 33.1 Modified Bell Classification for NEC

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

➢ Intussusception

- **Most common cause of SBO** in ages 4-9 months (**children** & **infants** not neonates)
- (**Intussuceptum**) is acquired invagination of the **proximal** bowel into the **distal** bowel(**intussuscipiens**)
- Starts with venous obstruction
- 2 types: primary-> no leading point (hypertrophied Peyer's patches) mostly in distal ileum // secondary-> leading point like Meckel diverticulum
- Presentation: abdominal pain + **current jelly stool** + hyper extension & flexion of knees
- Dance sign: RUQ pass and empty RIF (50%) -> bacteremia & bowel necrosis

- Diagnosis: gold standard-> **US**: transverse plane-> **donut or target** sign, longitudinal plane-> **pseudo kidney**
- Management: *non-operative: **hydrostatic/pneumatic reduction** (if stable not in perforation or hypotension) if unsuccessful or there is a leading point or signs of peritonitis do SURGERY

➢ Hypertrophic pyloric stenosis

- Male
- Younger maternal age is a risk factor
- Presentation: **2-8** weeks full term neonate with non-bilious, progressive projectile vomiting, visible peristalsis, succussion splash, if late will show signs of dehydration
- **Olive sign** (palpable pylorus) in **70-90%**
- Labs: hypochloromic hypokalemic **metabolic alkalosis** (greater loss of Cl-), paradoxical aciduria (in severe cases)
- Diagnosis: US-> muscle thickness >=4 mm & pyloric length >= 16 mm
- Management: supportive NPO, NG, surgery is not emergency (**pylomyometry**) complication of surgery: postoperative emesis

➢ Gastroschisis

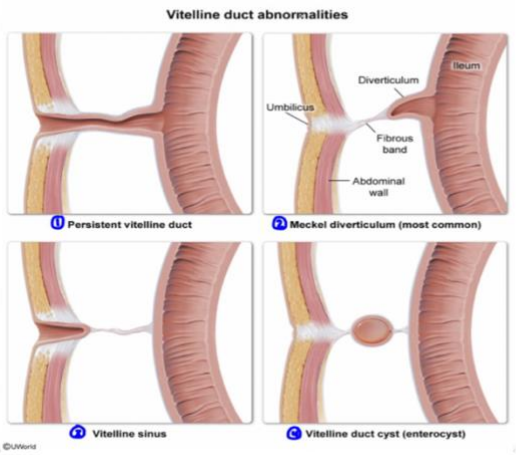
- Mother younger than 21 yo
- Diagnosis: antenatal US: by week 20, freely floating bowel loops in the amniotic fluid and a defect **right** to the umbilical cord + high AFP + IUGR
- Inflammatory thickening of the visceral bowel (matted intestines)
- Associated with intestinal motility disorder, UDT
- Management: wrapping the bowel in warm saline-soaked gauze (silo) then surgery primary closure or staged closure
- Excellent prognosis (depends on bowel condition)

➢ Omphalocele

- Associated with **genetic defects**
- Diagnosis: US at 18 weeks -> well contained sac protruding from umbilicus + high AFP
- Term vaginal delivery except if contains liver (worse prognosis)
- Management: sac should be wrapped in warm saline-soaked gauze
- Surgery options: 1. Primary closure if small defect 2. Staged closure using a mesh or silo 3. Paint and wait (sacrifaction) (honey dressing) if giant or associated with comorbidities

➢ Meckel's diverticulum

- True diverticulum
- **Most common anomaly of GIT**
- Persistence of the vitelline duct
- Rule of 2: 2% of population, 2:1 M:F, discovered by 2 years of age, located 2 feet from iliocecal valve, 2 cm in diameter, contain 2 types of mucosa (**gastric** most common and pancreatic)
- In children presents as: painless intestinal bleeding, intestinal obstruction, diverticular inflammation
- In elderly, neoplasia can develop (carcinoid)
- Diagnosis: technetium-99m pertechnetate (**Meckel scan**)
- Management: stabilize + surgery -> diverticulum resection + anastomosis



Presentation:
① Meconium discharge from umbilicus after birth
② painless hematochezia
③ serous umbilical drainage
④ Asymptomatic

➢ Biliary atresia

- Sclerosing cholangiopathy
- Most common indication for liver transplant
- Female
- Isolated mostly, but can be part of **BASM**: biliary atresia, splenic malformation, and malrotation
- Types: 1) TYPE 1 atresia of CBD 2) Type 2a atresia in common hepatic duct 3) Type 2b (most common) atresia in CBD + common hepatic duct [extrahepatic] 4) Type 3 atresia in extrahepatic and intrahepatic ducts
- Presentation: direct jaundice > 2 weeks + pale stool + cola urine + growth retardation
- Diagnosis: US -> triangular cord sign, MRCP -> contrast will not pass to the small bowel, hepatobiliary scintigraphy -> good liver uptake with no intestinal activity till 24 hr (no excretion)
- Management: surgery **[kassai procedure]**
- Best time to do kassai is before **2 months** of age
- May need liver transplant

Inguinoscrotal diseases

➢ Anatomy & physiology

Contents of inguinal canal:
Male —> ilioinguinal nerve + spermatic cord
Female —> ilioinguinal nerve + round ligament

Spermatic cord structures:

- Cremasteric muscle
- Testicular artery
- Pampiniform plexus
- Lymphatic channels
- VAS
- Genital branch of genitofemoral nerve
- Processus vaginalis

Walls of inguinal canal:

- Anterior (roof): external oblique aponeurosis
- Inferior wall: inguinal ligament + lacunae ligament + iliopubic tract
- Superior wall: internal oblique muscle + transversus abdominus muscle
- Posterior wall: transversalis fascia + conjoint tendon

Inguinal canal:

- Internal ring: Located in the internal oblique muscle.
- External ring: Found in the external oblique aponeurosis.

➢ Inguinal hernia

- Commonest groin swelling
- More in premature males
- More in **right** side
- Due to failed obliteration of patent process vaginalis
- Sliding hernia: may contain fallopian tube, ovary, bladder
- Amyand's hernia: herniated appendix
- Richter hernia: ischemic antimesenteric bowel border
- Pantaloon hernia: direct and indirect inguinal hernias
- Clinical diagnsos, mostly asymptomatic bulge
- In pediatrics inguinal hernia is commoner
- Management: **PPV ligation surgery**, it is not an emergency but should be done **in first available time** due to risk of incarceration
- Exploration of the free contralateral side if: premature, young, female, left
- If incarcerated: try to reduce with sedation then admit and repair within 24-48 hrs

- If strangulated —> tender, sick looking, skin discoloration, septic shock, signs of peritonitis, or failed reduction: do emergent surgery
- Recurrence rate < 1%
- Mesh is almost never used in children (unless recurrent)

➢ Hydrocele

- Peritoneal fluid
- If asymptotic: observe for 1-2 years of age, usually resolves spontaneously (more if non communicating)
- If didn't resolve or symptomatic or presence of herina then do surgery —> PPV ligation + drainage of hydrocele

➢ Undesended testis

Testicular Descent:

- Testes develop in the abdominal cavity and descend through the inguinal canal into the scrotum.
- Descent occurs in two phases:
    1. Abdominal Phase – Influenced by Insulin-like Hormone 3 (INSL3) from Leydig cells and Anti-Müllerian Hormone (AMH) from Sertoli cells and gubernaculum
    2. Scrotal Phase – Influenced by CGRP and androgen

- More common in premature 40%
- Majority (70-80%) of testes descend within first 6-12 months
- Most common location: superficial inguinal pouch
- Non palpable: agenesis, intra-abdominal, vanished, small, obese child, non-experienced examiner
- Palpable (70%): inguinal, retractile, ascending, peeping, ectopic (not in normal pathway)
- Increased risk of malignancy (if not fixed -> **seminoma**)
- Affect fertility
- High risk of torsion
- PE: empty hemiscrotum
- Managemnt: if unilateral palpable/non palpable -> orchidopexy or orchidectomy
- If bilateral non palpable or ambiguous genitalia -> hormones + karyotyping
- Retractile testes should be observed and examined annually

➢ Testicular torsion

- Neonates and adults
- First **6** hours are important for salvage
- Presentation: sudden sever unilateral testicular pain/lower thigh or lower abdominal pain + enlarged testis + severe tenderness + cremasteric reflex **might** be absent
- Two types: 1. Extravaginal: perinatally 2. Intravaginal: bell-clapper
- Management: exploration under GA, detorsion (medial to lateral), place it in warm saline if it gets better then fix it and do a contralateral fixation. If non viable remove it

➢ Torsion of testicular appendages

- Most common cause of acute scrotum
- **Prepubertal** (hormonal)
- Blue dot sign
- Self limited

➢ Epididymitis / orchitis

- Slow onset
- If bacterial may have positive urinalysis and culture and requires Abx
- If viral, self limited

➢ Idiopathic scrotal edema

- Bilateral
- Boys 5-9 years old
- Swelling that begins in perineum or inguinal then spreads to scrotum
- Normal blood flow on US

➢ HSP

- Scrotal pain + skin purpura + joint pain + hematuria
- Normal blood flow on US

➢ Testicular trauma

- Check for sexual abuse
- On US evaluate for rupture tunica albuginea
- Management: exploration and repair

Pediatric solid tumors

❖ Neuroblastoma

- Unpredictable behavior
- Arise from primitive sympathatic ganglion cells
- Can synthesize and secrete **catecholamines**
- Mostly 1-4 years of age
- **Mostly** originates from adrenal **medulla**
- **Least** common site: anterior mediastinum (worst prognosis)
- Presentation: abdominal mass with sick appearance, secretory diarrhea, hepatomegaly
- (Raccoon eyes + bone marrow findings + neurologic symptoms → think neuroblastoma)
- Elevated **VMA** & **HVA** in urine
- Diagnosis: abdominal X-ray-> **tumor calcification**
- **Dark blue round cells**, rosette formation
- For staging **CT**
- Dumb-bell tumor when there is intraspinal extension
- Should do tissue biopsy + **bone marrow biopsy (most common site of mets)**
- Management: chemotherapy with or without surgery depending on stage
- Stage MS: 30%, blueberry muffin spots, mets to skin, liver, bone, 80% spontaneous regression
- No screening recommended

❖ Nephroblastoma (Wilm's tumor)

- Highly malignant but mostly **favorable**
- Most common pediatric renal tumor
- Neuroblastoma is the most common intraabdominal malignancy followed by Nephroblastoma
- Mostly solitary & sporadic
- Arises from **fetal undifferentiated metanrphric blatesma**
- Presentation: **asymptomatic abdominal mass**
- Rare feature: varicocele on the right side!!
- CT/MRI for diagnosis
- Echo may show right atrial involvement
- Management: surgery (nephrectomy) + chemotherapy
- When to do partial nephrectomy? If bilateral WT, contralateral pre-existing abnormality of kidney, WT in single kidney, WT in nephroblastomatosis
- Prognosis is worse as the patient is younger in age (in contrast to Neuroblastoma)

Foreign body aspiration

❖ Esophageal FB

- <= 5 years of age
- 70% at **cricopharyngeus** sling (upper esophageal sphincter)
- Normal physical exam
- Diagnosis: Neck & chest X-ray + contrast esophagography + eshophagoscopy
- How to extract a coin? 1. Endoscopy (nonemergent) 2. Foley ballon extraction with fluoroscopy

❖ Gastrointestinal FB

- Mostly asymptomatic
- **Outpatient** management
- If didn't pass —> endoscopy
- If in ileum —> may need laparoscopy

❖ Batteries

- Double contour rim on xray
- Damage to esophagus may continue after removal
- If passed esophagus and asymptomatic —> can be observed at home then follow up

❖ Magnets

- Most common symptom is abdominal pain
- If 1 magnet —> outpatient observation
- If 2 magnets or 1+metallic FB —> close inpatient observation + urgent surgery
- +- endoscopy or laparoscopy

❖ Sharp FB

- Risk of perforation 15-35%

❖ Bezoars

- Tight collection of undigested material
- **Radiolucent**
- <u>Operation is necessary</u>
- Phytobezoars: causes obstruction at ileo-cecal valve
- Trichobezoars: associated with trichotillomania, rapunzel syndrome when involves stomach + small bowel, removed by gastrotomy or laparotomy

❖ Airway foreign body

- Subglottic region is the narrowest part **(fatal)**
- If stuck on vocal cords —> death
- Usually in the right bronchus: larger in diameter, greater airflow, smaller angle of divergence from trachea
- More in boys
- Inspiratory stridor —> laryngeal FB
- Expiratory stridor —> tracheal FB
- Diagnosis on chest X-ray: hyperlucency due to **hyperinflation** (air trapping), hyperextended lung, flattening of diaphragm, widening of intercostal spaces, straightening of ribs, mediastinal shift
- 56% normal chest film within 24 hrs of aspiration
- **Definitive diagnosis: bronchoscopy**
- Flexible —> to diagnose, rigid —> diagnostic & therapeutic
- FB distal to the main bronchus —> Fogarty catheter
- Complications of bronchoscopy: bleeding, laryngospasm, pneumothorax, hypoxia

Pediatric urology

❖ Uretropelvic junction obstruction

- Male, left side palpable mass
- **Antenatally asymptomatic** but on **US -> hydronephrosis** (most cases are identified here)
- Etiology: * intrinsic narrowing: functional obstruction, loss of ganglion cells, ureteric stricture * extrinsic: aberrant renal vessel, kinking (due to severe VUR)
- May cause flank pain, flank mass, hematuria, recurrent UTIs
- Diagnosis: **postnatal US**, renal radioisotope scan (**MAG3**), **MCUG** (to rule out VUR)
- MAG3 scan findings: * renal function (when <40% needs intervention) * pelvic drainage curve (after furosemide) if t1/2 > 20 min -> needs intervention
- Mx: observation or pyeloplasty if: function <40% or t1/2 > 20 minutes or symptomatic

❖ Vesico-uretric reflux

- Female, 3 years
- Tunnel length:ureteral diameter should be 5:1 to prevent VUR
- Primary VUR (most common): due to short ureteral submucosal tunnel
- Secondary VUR: posterior urethral valve, neurogenic bladder, anterior urethral valves, uretroceles, bladder diverticula, ectopic ureters
- Clinical features: recurrent UTIs, renal scarring, renal dysfunction, hypertension, reduced somatic growth
- Diagnosis: * urine analysis —> to rule out infection, * KFT —> normal, * US —> hydro-uretero-nephrosis, * DMSA —> renal scars, *MCUG [confirmatory imaging] —> for degree of VUR, * direct isotope cystography (DIC) —> for follow-up

- Treatment of lower grade reflux (1,2,3): antibiotic prophylaxis, resolves spontaneously +- subureteral submucosal injection of **bulking agent**
- Surgery: reimplantation of ureters
- Indications for surgery: 1. Failure of submucosal injection 2. Deterioration of renal function 3. New scars 4. Secondary VUR 5. Grades (4 & 5) 6. Hypertension 7. Single kidney with higher grade VUR 8. Decrease in renal growth
- After surgery: Abx for 3 months
- Surgery prevents further damage but does not reverse scarring nor parenchymal damage

❖ Phimosis

- Foreskin is unable to be retracted to expose the glans
- Physiological phimosis is normal in first years of life
- Pathological phimosis: *primary: true congenital phimosis with pin-hole meatus *secondary: to **bacterial** infection—> balanitis, posthitis or due to balanitis xerotica obliterans
- **Conservative** management with topical steroids and self retraction or do circumcision
- **Paraphimosis**: foreskin is able to be retracted but become **stuck** —> distal congestion and edema of glans.. Surgical emergency: in ER compress with ice or multiple needle punctures if failed —> send to OR to do dorsal slit of the tight band + circumcision
- Circumcision contraindications: bleeding disorder, **hypospadias** (relative)
- Circumcision complications: bleeding, inadequate/excessive skin removal, infection, meatal stenosis in neonates

❖ Hypospadias

- **Complex** of: abnormal ventral urethral meatus, dorsal hooded foreskin, glans defect, underdeveloped corpus spongiosum, phallic torsion, chordee (phalic ventral curvature)
- Associated with: high parity, high maternal age, LBW, +ve family history, inguinal hernia, hydrocele, undescended testes, increased maternal progesterone exposure (IVF)
- Mostly isolated
- 50% distal, 30% middle, **20% proximal (worst)**
- Best time for surgery is before 18 months
- **Tube urethroplasty** + corpus spongioplasty + straightening phalloplasty + glansplasty + circumcision + phalloplasty
- Late complications: meatal stenosis, urethrocutaneous fistula, persistent chordee, urethral stricture, urethral diverticulum

Done by Ruaa Hdeib