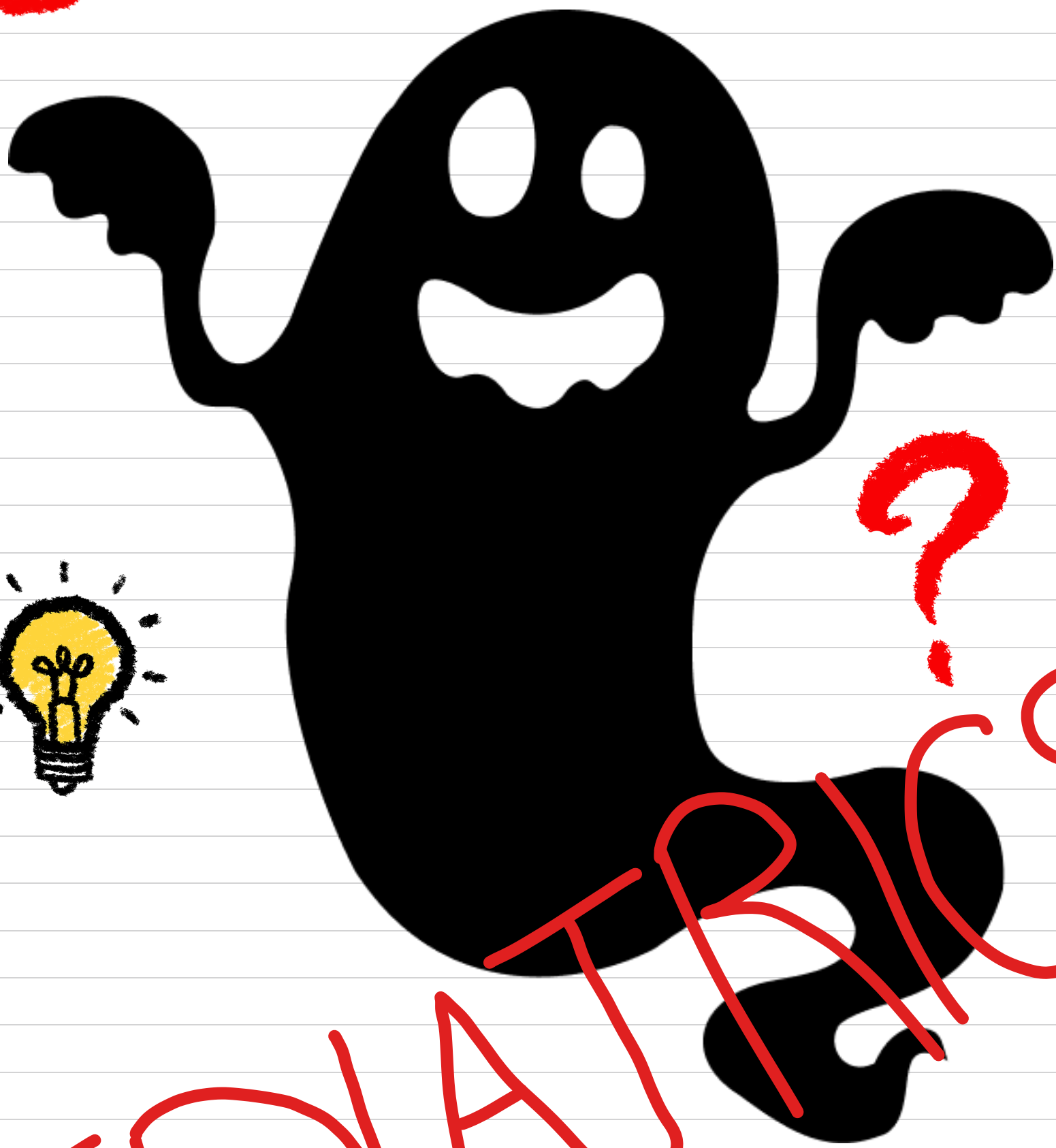
دعواتكم



PEDIATRICS

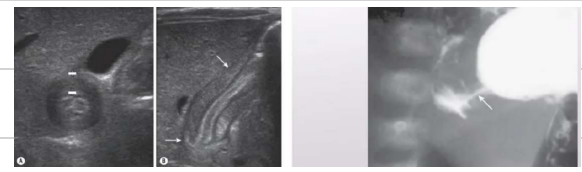
لا تنسونا من دعواتكم !!

الكاتبة : سارة جمال

بالتوفيق جميعا

Miscellaneous topics:

1 HPS:



M:F → 4:1

* presentation: non-bilious, progressive, projectile vomiting of recent feedings
* 2-8 weeks
* well but if late presentation: ① signs of dehydration
  ② visible gastric peristaltic waves
  ③ palpable pylorus "olive sign" 70-90%
* investigation: hypochloremic hypokalemic metabolic alkalosis. // loss of fluid, K^+, HCl then contraction alkalosis, paradoxic aciduria
* diagnosis: US → muscle thickness ≥4mm + pyloric length ≥16mm + upper GI series.
* management: pre op: NPO +/- gastric decomp
  ⓐ: IV fluid resuscitation
  ⓐ: correction of electrolytes

  Surgery: * non emergent
  * laparotomy or Laparoscopic pyloromyotomy

2 Intussusception: * proximal bowel (intussusceptum) into distal (intussuscipiens) that will compress the mesentery resulting in venous obstruction and bowel edema → arterial insufficiency, ischemia and bowel necrosis

m.c.c of small bowel obstruction in this age group (after hernia) * 80% ileocolic

- primary: no leading point, likely due to hypertrophied peyer patches within the bowel wall // 4-9 months // 2/3 boys.
- Secondary:

classic presentation: ⊛ infant or young child with intermittent, cramping abd pain every 15-30 min asso with current jelly stools and palpable mass on physical examination (seen in <25%)

⊛ pain is asso with vomiting (gastric early, bilious later)
⊛ abd destension
⊛ hyperextension and flexion of the knees up.
⊛ lethargy (encephalopathy)
⊛ on exam → signs of dehydration
empty RIF (Dance sign)
abd distension.

Management:
* NGT
* NPO
* IVF
* correct electrolytes
* non op management
* Hydrostatic/pneumatic reduction (fluoroscopy/usguid)

operative management (lap or open)
① non-op reduction → incom
② peritonitis/pneumoperitoneum
③ Lead point (sec intussusception)

Dx: Xray
US: Target or "donut" lesion → Transverse plane + pseudoKidney → Longitudinal plane.

3 Congenital abd wall defects

Ⓐ Gastroschisis : central abd wall defect , 4cm right to umbilicus , no covering membrane , only midgut

DX: US by 20 weeks gestation (prenatal) : bowel loops freely floating in amniotic fluid + defect in abd wall

elevated α FP

↑ acetylcholinesterase

IUGR : intrauterine growth restriction.

* characteristic appearance : Matted intestines.

* Management: ① Resuscitation (NPO / NG / IUF / rectal tube decomp

② bowel should be wrapped in warm saline-soaked gauze and placed in central position on abd wall.

③ Surgery → primary closure or staged closure.

Ⓑ omphalocele:

asso with : ① Trisomies (13, 18, 21, 45x)

② Beckwith-Weideman → omphalocele / hypoglycemia / macroglossia / organomegaly

③ pentalogy of Cantrell → [omphalocele / defective sternum / ventral diaphragmatic hernia, cardiac anomalies / ant pericardial defect]

④ Cardiac anomalies (MC)

⑤ CNS anomalies.

Long term morbidities: GERD / pulmo insuff / reccurent lung infection / asthma / feeding difficulty with failure to thrive.

antenatally: US + ↑ AFP

Deliver: vaginal delivery (except if giant omphalocele + containing liver to avoid [shoulder dystocia + sac rupture + bleeding]

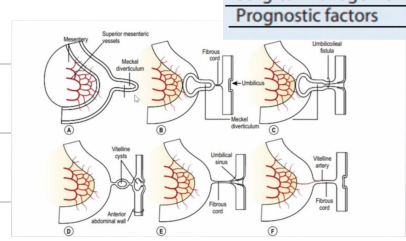
Management: ① Resuscitation ② Sac ③ Surgery

prognostic factors : asso anomalies. / Cx: ① hypothermia bc loss of fluid

② abd compartment or hepatic vein torsion.

Table 48.1 Differentiating Characteristics Between Gastroschisis and Omphalocele

Characteristic	Omphalocele	Gastroschisis
Herniated viscera	Bowel ± liver	Bowel only
Sac	Present	Absent
Associated anomalies	Common (50%)	Uncommon (<10%)
Location of defect	Umbilicus	Right of umbilicus
Mode of delivery	Vaginal/cesarean	Vaginal
Surgical management	Nonurgent	Urgent
Prognostic factors	Associated anomalies	Condition of bowel



4 Meckel diverticulum:

* m.pt are asympt

* M:F → 2:1

* rule of 2's → 2% of popu / 2:1 M:F / discovered by 2 years of age / 2 feet from ileocecal value / 2 cm diameter / 2 inches 5cm / 2 types of heterotopic mucosa (Gastric is the m.c followed by pancreatic).

3 m.c presentation in children ① intestinal bleeding
② = obst
③ diverticular infla

- Less common signs include a cystic abdominal mass and a newborn with an umbilical fistula resulting from a patent vitelline duct, In elderly, neoplasia can develop within the Meckel diverticulum. (Carcinoid is the most common tumor)

Management: * stabilize the pt in case of bleeding
* Surgery : open or Laparoscopic diverticulum resection or segmental bowel resection + anastomosis

5 Biliary atresia: sclerosing colangiopathy, m.c.c of end stage liver disease, m.c indication for liver transplantation in children.
* F > M
* isolated in 85%
* it occurs as part of a syndrome → m.c → BASM (splenic malform / malrotation) → #Biliary atresia.

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

Type I: c. bile duct
IIa: c. hepatic duct
II b: both
III: all extrahepatic up to porta hepatis.

presentation: jaundice / pale stool / hepatomegaly / anemia, malnutrition, growth retardation b/c malabsorption of nutrients and fat-soluble vitamins.

Surgery: Roux-EN-Y limb and enterostomy for portoenterostomy (Kassi procedure)
* following successful Kassi op → pigmented stool within 2-3 weeks.

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

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

Inguino-scrotal disease:

1 Inguinal hernia and hydrocele

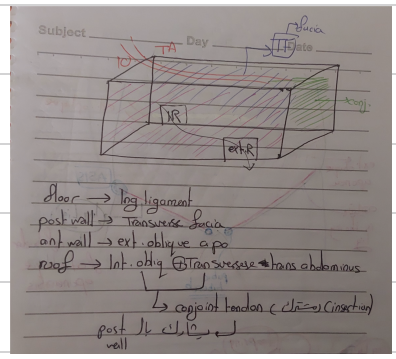
- What is Process vaginalis

→
- In the inguinal canal → gradually obliterates after birth
- In scrotum → forms the tunica vaginalis around the testis

Ⓐ Inguinal hernia : ⟶ pathogenesis: failed obliteration of patent process vaginalis.

*more in premature
* M>F full term but premature M>F
* family history
* Rt>Lt

anatomy



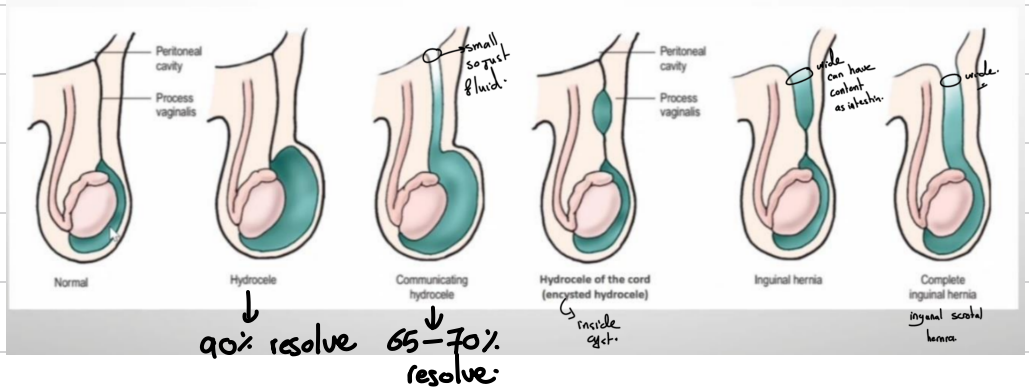
contents of inguinal canal:

♂: ilioinguinal nerve + spermatic cord

♀: : = = + Round ligament

*spermatic cord structures:

① cremasteric muscle (from internal oblique)
② genital branch of genitofemoral n.
③ Testicular artery
④ pampiniform plexus
⑤ Lymphatic channels.
⑥ VAS
⑦ processus vaginalis.



① Hydrocele
② Communicating hydrocele
③ encysted hydrocele (of the cord)
④ Inguinal hernia
⑤ complete Ing hernia (inguinoscrotal)

* sliding hernia : [may contain] → [fallopian tube / ovary / side wall of urinary bladder]
* Amyand's hernia: if @ppendix herniated
* Litter's hernia: meckel diverticulum herniated
* Ritcher hernia : ischemic antimesenteric bowel
* pantaloon hernia: direct + indirect Ing hernia. more in neonates.

most are asymp. ⟶ so can simply be observed for 1-2 years of age
Indications of surgery : pain / fails to resolve / clinical hernia is apparent.

- NO DIFFERENCE IN RECURRENCE (< 0.5%)
- ↓ INCIDENCE OF METACHRONOUS HERNIA
- ↓ OP. TIME FOR LAP. BILATERAL REPAIRS
- ↑ OP. TIME WITH LAP. UNILATERAL REPAIR

Incarcerated hernia ⇒ Try to reduce it
* with sedation (midazolam → apnea) monitor O2
* firm + continous p
* if reduced → admit and repair in 24-48h
* if failed or incomplete reduction or contraindic ↳ EMERGENT surgery

Dont reduce if
① signs of peritonitis ② septic shock

* open v.s. Laparoscopic ppV Ligation
↳ prematurity
↳ younger age
↳ female gender
↳ Lt sided unilateral hernia.

MESH is ALMOST NEVER used in CHILDREN
except: reccurent herneas in children with CONNECTIVE TISSUE DISORDER or MUCOPOLYSACCHARIDOSES

Ⓑ Hydrocele : accu of peritoneal fluid in non obliterated process vaginalis.

Surgery : high ligation of process vaginalis + drainge of hydrocele. ⟶ Lord's / Bottle / Jaboulay's procedure

Deep inguinal ring
- abd descent guided by the gubernaculum and anti-mullerian Hormone
- Inguino-scrotal descent along the processus vaginalis driven by testosterone

2 UDT: undescended Testes

* RF: premature or Low brith weight
* usually descend spontaneously by 6-12 months of age
* classification:
  - Non palpable UDT
    - → Testicular agenesis
    - → Intraabdominal UDT
    - → Vanished testis (atrophied due to previous vascular insult as perinatal torsion/trauma/iatrogenic)
    - → small testis / obese child / no experienced examiner.
  - palpable UDT:
    - → inguinal udt
    - → retractile testis (cremastric overactivity)
    - → ascending testis (acquired iatrogenic)
    - → peeping testis
    - → ectopic testis

- ASSOCIATED ANOMALIES:
  - PATENT PROCESSUS VAGINALIS
  - EPIDIDYMAL ABNORMALITIES
  - PRUNE-BELLY SYNDROME
  - GASTROSCHISIS
  - BLADDER EXSTROPHY
  - PRADER–WILLI, KALLMAN, NOONAN SYNDROMES
  - TESTICULAR DYSGENESIS
  - ANDROGEN INSENSITIVITY SYNDROMES

MALIGNANCY RISK
FERTILITY

presentation: emtpy hemiscrotum during neonatal checkup or later visits

- HISTORY IS IMPORTANT (GESTATIONAL AGE, PRESENT AT BIRTH, HISTOY OF TRAUMA/TESTICULAR TORSION , PREVIOUS INGUINAL SURGERY)

on exam ① scrotum → size
- darker skin color
- presence of rugae
- signs of development

② palpate scrotum bilateraly + testis + ing region.

MANAGEMENT

HORMONES (LH-RH AGONIST)
CONTROVERSIAL

SURGERY
WHY WE DO SURGERY ?

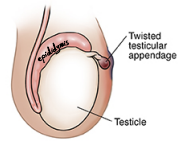
- REDUCES THE RISK OF MALIGNANCY AND INFERTILITY
- REDUCES THE RISK OF TORSION
- EASIER EXAMINATION
- PSYCHOLOGICAL : NORMAL-APPEARING SCROTUM
- ENHANCE ENDOCRINE FUNCTION

3 Acute scrotum:

* = acute scrotal pain
* most are nonurgent
* age → l.m clue

torsion of the appendix testis/epididymis → prepubertal boys

testicular torsion → neonates + adolescents



Torsion of testis
torsion of the appendix testis/epididymis
Trauma/sexual abuse
Tumor
hernia/hydrocele
epididymitis/orchitis
idiopathic scrotal edema (dermatitis)
cellulitis
vasculitis. (Henoch-Schönlein purpura)

Ⓐ **testicular torsion** = twisting of the spermatic cord → compromise vasculature → infarction

* salvage ↓↓ after 6 hours.
* befor 3y + after puberty
* presentation: sudden, severe, unilateral testicular pain/lower thigh/lower abd pain, nausea and vomiting
* if intermittent → incomplete torsion with spontaneous detorsion.
* enlarged testis/retracted up/transverse orientation/ant. located epididymis/severe generalized testicular tenderness/swelling/erythema

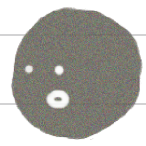
Cremastric reflex is often absent.

2 types:

- Extravaginal: spermatic cord twists proximal to the tunica vaginalis (tunica + testis spin on the vascular pedicle)
- Intravaginal: more in children and adolescents. ♀ spermatic cord twists within the tunica vaginalis "Bell-clapper deformity"

Tx: ⊛ exploration under GA/detorsion/placement in warm saline-gauze/fixation/contralateral fixation.
⊛ if nonviable → remove

Ⓑ **Torsion of testicular appendages** → m.c.c of acute scrotum



* btw 7 and 10
* presentation: sudden onset of pain and nausea, appendage can be palpated [Blue dot sign]
* self limited, NSAIDS, restricted activity, warm compresses.
* physical: induration, swelling, tenderness of hemiscrotum, ⊕ urinanalysis and culture/urethral swab in sexually active suggest dx.
* Tx: Abx + supportive (self limited)
* Viral: Mumps orchitis (rare), Adenovirus, enterovirus, influenza, and parainfluenza virus infections
* scrotal pain + swelling + slow onset + worsening overdays.

Ⓒ **idiopathic Scrotal edema**

* swelling + erythema
* 5-9 years
* insidious onset and erythema begins in the perineum or inguinal region and spread to hemiscrotum, pruritis.
* Testis is not tender DDx: contact dermatitis/insect bites/minor trauma/cellulitis

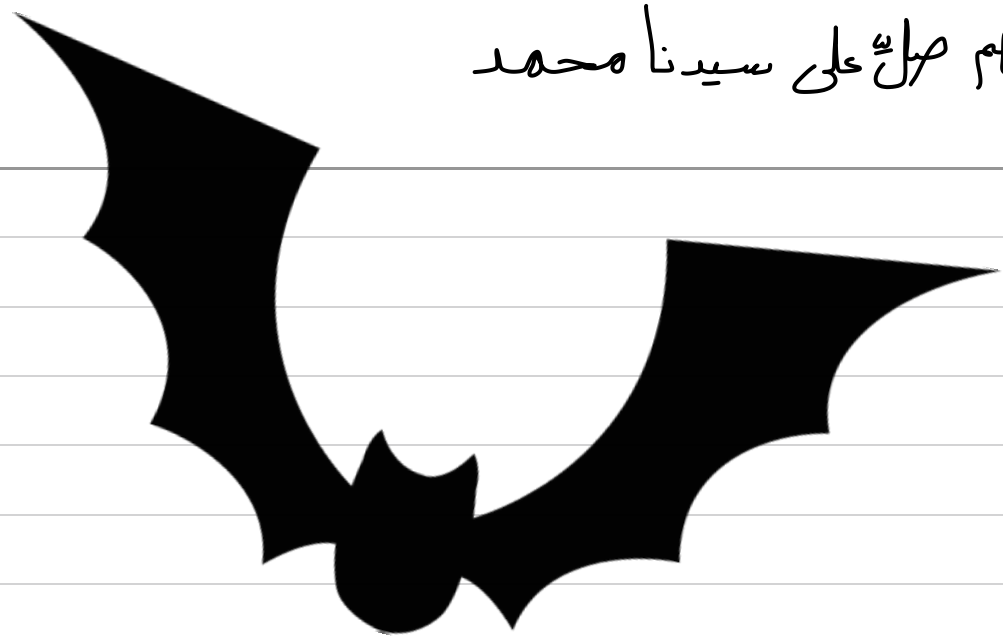
(D) Henoch-schonlein purpura

* vasculitis syndrome ; involve (skin/joints/GI/GU)
* symp: scrotal + spermatic cord pain / erythema / swelling / skin purpura / joint pain / hematuria
* 7y.
* doppler US: normal ↓. flow to testis.
* Management: Conservative.

(E) Testicular trauma:

* injured testis is swollen and tender , swelling and bruising of the scrotum
* evaluate for rupture of tunica albugina
* Tx: exploration ± repair ruptured↑

اللهم صل على سيدنا محمد



Ingestion/Aspiration of FB

→ **esoph. FB**

* more in ≤5y
* Us → coins
  marine areas → fish bone
* eso is the narrowest portion of GI tract.
* 3 areas of narrowing
  1. circopharyngeus sling (70%)
  2. Level of the aortic arch in the mid esoph.
  3. LES (GE junction)
* other areas: 1) underlying eso. patho (strictures (→ caustic ingestion) or esinophilic esophagitis)
  2) prior eso surgery (eso artesia)

* sharp FB may pentrate the mucosa and cause:
  1. mediastinitis
  2. aortoenteric fistula
  3. peritonitis

Hx: → witnessed event or disappear of an object.
Symp can vary:
→ completely asymp
→ drooling
→ neck and throat pain
→ dysphagia
→ emesis
→ wheezing or RS distress
→ abd pain.

PEx:
* majority → normal
* signs of CX:
  1. oropharyngeal abrasions
  2. crepitus
  3. signs of peritonitis.

Management:
1. Neck + chest X-ray
2. ± contrast esophagography
3. ± esophagoscopy

Foley catheter technique
* the balloon filled with contrast
* under fluoroscopy
* care to avoid aspiration
* very cost-efficient.

Coins: * most located in the proximal esophagus.
* majority of proximal will remain entrapped and require retreival → options:
  1. magal forcep
  2. endoscopy (rigid or flexible)
  3. foley balloon extraction with fluoroscopy (80% success)

* if reached Lower esophagus
  ↳ can spontaneously pass into stomach
  ↳ can be observeed
  ↳ can be advanced into the stomach with (NGT in ER)

→ Gastrointestinal FB

* if distal to eso. usually asymptomatic → most pass smoothly through GI tract out through anus
* signs and symp: → ex. in stomach
  - abd pain
  - nausea / vomiting
  - Fever
  - abd distention if intest. obst.
  - peritonitis if perforation

Mx: ① can be managed as an outpatient
② ? prokinetic agents and cathartics
③ if didn't pass → endoscopy (for 4-6 wks)
④ Laparoscopy [from jejenum to terminal ileum → non reachable nither by endoscopy nor colonoscopy]

Special topic Ingestions:

1 Batteries:

* mostly asymp, symp just in <10%
* on radiographs: double contour rim
* problem → contact time btw the battery and eso. bc narrow
* tissue injury → pressure necrosis
  → relase of low-voltage electric current
  → Leakge of alkali solution (liquefaction necrosis)
* mucosal injury may occure in 1 hour of contact time and may continue even after removal.
* Immediate removal
* early and late Cx:
  - esophageal perforation
  - tracheoesophageal fistula
  - stricture and stenosis
  - mortality

* if the battery is confirmed to be distal to the eso AND the pt is asymptomatic → observe

2 MAGNETS

* Sig morbidity if ① multiple magnets
② single magnet + sec metalic FB

if 2 connected magnets swallowed togther → not major problem but if seprated → if 2 diff areas in GIT → pressure necrosis → perfo, fistula, stricture .....

* m.c symp is abd pain
* <40% symp
* plain radiograph (m.commonly used to confirm diag)
* Mx: * close inpatient observation (if 2 magnets or 1 + metallic FB or if in doubt)
  * outpatient observation (if 1 magnet)

* they may attach to each other and lead to: obstruction, volvulus, perforation, fistula.

Sharp foreign bodies
* 15-35% risk of perforation
* Mx: * conservative: smaller objects and straight pins.
  * endoscopic retrieval
  * close inpatient observation

Bezoars: is a tight collection of undigested material
* include
  - Lactobezoars (milk)
  - phytobezoars (plant)
  - trichobezoars (hair)

* presenting symptoms: nausea / vomiting / weight loss / abd distention
* Diagnostic imaging: plain radiographs, upper GI contrast, endoscopy.
* Mx: operation is necessary (phyto + tricho)

- phytobezoar → * vegetable matter
  * usually obst at the ileo-cecal valve level
- Trichobezoar → * hair
  * Rapunzel syndrome (stomach ts. bowel) asso with trichotillomania
  * tipically removed by gastrotomy + Laparatomy + Laparoscopy

→ Airway FB:
* anatomical diff in airway of young children compared with older children
  - ↳ shorter airway / smaller in calibre
  - ↳ anteriorly positioned Larynx (↑ difficulty with oral intubation)
  - ↳ Subglottic region is the narrowest part

Fixed right main stem bronchus:
- Larger in diameter
- airflow is generally greater
- smaller angle of divergence from the trachea

* high index of suspicion is required.

*common presenting symp:

① RS distress

② stridor * inspiratory → Laryngeal FB
* expiratory → tracheal FB

③ wheezing

④ Dysphonia

*many will be asymp

* may completely obstruct the Larynx or trachea producing sudden death.

* Ap + lateral films of the neck and chest (inspiratory and expiratory)

* hyper inflation or "air trapping" ± mediastinal shift

*5%: → normal chest film in 24 of aspiration

* radiopaque FB are easily identified

* radiolucent FB → indirect clues.

* definitive Dx requires Bronchoscopy → flexible → to diagnose
→ rigid → diag + therapeutic

* Fogarty catheter

- Overall **complications** of rigid or flexible bronchoscopy:

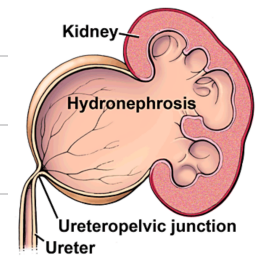
* • Bleeding from local inflammation
* • Laryngospasm
* • Pneumothorax
* • Hypoxia

- Rarely a thoracotomy with bronchotomy or lobectomy is required. if bronchoscopy isn't beneficial

4 topics → ✡✡✡✡

"يارب"

principles of pediatric Urology



✡ UPJO : uretropelvic junction obstruction

* M:F → 2:1 / Lt:Rt → 3:2 / bilateral : 10-40%
* most present as Hydronephrosis detected by antenatal us
* etiology
  - Intrinsic bc → intrinsic narrowing
    - rarely : mucosal valves, polyps, ureteric strictures
  - extrinsic → aberrent renal vessels
    - Kinking as a result of severe vesicoureteral reflux (VUR)

* clinical features:
* most are asymptomatic (detected via prenatal screening us)
* when symptomatic
  - flank or abd pain
  - palpable flank mass
  - Hematuria
  - reccurent UTIs

Investigations:

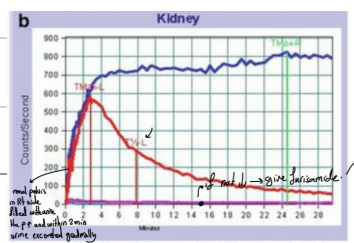
① postnatal us: * the primary investigation tool for HN
* assess kidney anatomy + Ap diameter of renal pelvis

② Renal radioisotope scan (function)

MAG3 is the scan of choice (percentage of perfusion to kidney)

- Differential renal function
  - normally 50% , 50%
  - acceptable down to 40%
  - needs intervention when <40%
- pelvic drainge curve
  - shows pelvic emptying t1/2 after administration of furosemide
  - normally t1/2 < 20 min
  - t1/2 >20 = sig obst. → needs intervention.

③ MCUG → to rule out whether HN is due to VUR (vesicourethral reflux)



Obstructed kidney

Normal kidney

Tx:

*antenatally detected HN

- Conservative management
- surgery → any or all → functional deterioration <40%
  → T1/2 >20 min
  → symp

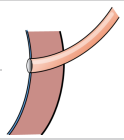
  └ open or lap pyeloplasty 1) excision of the narrowed segment
  2) anastomosing ureter to the most dependent portion of renal pelvis
  3) excision of redundant renal pelvis.

* Endourological pyeloplasty → use of balloon dilations, percutaneous antegrade endopyelotomy, retrograde uretroscpic endopylotomy

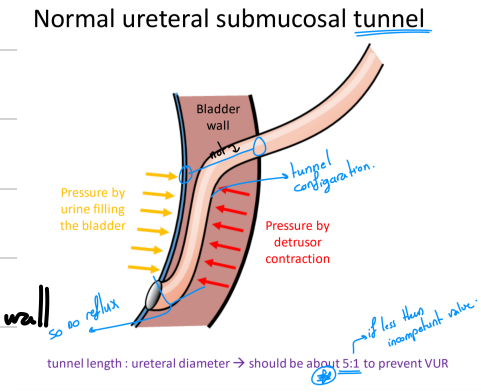
Vesico-Ureteric reflux:

*F predominant

* peak → 3yrs
* familial incidence 2-4%



pathology: M.C.C primary VUR due to short ureteral submucosal tunnel in bladder wall



└ Secondary VUR: due to either → post urethral valve (PUV)
→ neurogenic bladder (NB)

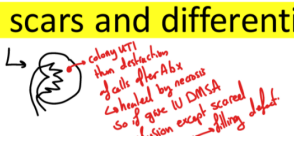
clinical features:

* symp of UTI | recurrent UTIs
* Renal scarring (due to previous pyelonephritis - upper UTI)
* renal dysfunction
* HTN
* reduced somatic growth.

Investigations:

① Urine analysis → r/o infection

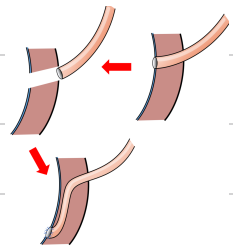
- US → HUN (hydro-uretero-nephrosis)
- DMSA nuclear scan → for renal scars and differential renal function
  ↳ attached to Technetium as MAG3.
- MCUG → for degree of VUR
  Micturating Cystogram



Tx: * Low-grade reflux (I, II, III):
- most likely → resolve spontaneously with age
- antibiotic prophylaxis
- ± subureteral submucosal injection of bulking agent
  - Results better for lower grades of reflux (>80% success)
  - Less successful in children with neurogenic bladder (NB)

Surgical Tx indications

- **Indications:**
  - Failure of chemoprophylaxis and/or submucosal injection therapy
  - Deterioration of renal function &/or appearance of new scars
  - Secondary VUR (due to ureterocele, duplex ureter, PUV, or neurogenic bladder)
  - Higher grades VUR (IV, V)
  - Hypertension
  - Single kidney with higher grade of VUR
  - Decrease in renal growth or somatic growth



= Reimplantation of Ureters

However → doesn't reverse scarring nor parenchymal damage.

circumcision:

* at birth: foreskin is adherant to the glans (non-retractable) (physiological phimosis)

* At 2-4 years: dissolution of adhesion → foreskin can retract

* at 5 years → most boys have normal foreskin retraction # if not → pathological phymosis.

→ indications: ① religious
② pathological phimosis
③ paraphimosis
④ reccurent UTI with no known cause.

phimosis: foreskin is unable to be retracted to expose the glans.

- physiological (in 1st y of life)
- pathological
  - primary: congenital with pin hole meatus
  - scondary to:
    - Bacterial infection:
      - **balanitis** (inflammation of the glans)
      - **posthitis** (inflammation of the foreskin)
    - Balanitis xerotica obliterans (BXO)

Mx: conservative → reassure, gentle-self retraction.
→ Topical steroids (eg. betamethasone)
Surgical → Circumcision
→ preputial stretch or plasty

paraphimosis: foreskin is able to be retracted but becomes stuck in that position resulting in distal congestion and edema of glans.

Mx: Surgical emergency

- **Manoeuvres can be done in ER:**
  - Compresses with ice or sugar (to reduce the swelling and allow protraction)
  - Multiple needle punctures (to allow fluid to be squeezed out) ↓ edema
- if failed → send to OR
→ dorsal slit of the tight band +/- circumcision (under GA) (→ general anesthesia)

circumcision → freehand
→ surgical clamps

Contraindications:

→ Absolute: * family history of bleeding disorders
* newborn with bleeding tendency /disorder/pathological jaundice.

→ Relative: Hypospadias

Circumcision | Complications

- **Bleeding**
- Infection
- Meatal stenosis
- Insufficient/excessive foreskin removed
- Adhesions, skin bridges, or inclusion cysts
- Entrapped penis or secondary phimosis
- Urethral injury (iatrogenic hypospadias)
- Necrosis of the penis (injudicious use of electrocautery to control bleeding)
- Amputation of the glans (partial or complete)
- **Death** (mainly due to unnoticed bleeding)

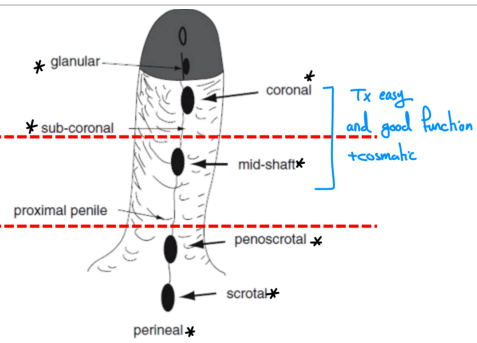
Hypospadias:

- Definition → * abn. ventral urethral meatus
* dorsal hooded foreskin
* glans defect
* underdeveloped corpus spongiosum
± phalic torsion
± phallic ventral curvature (chordee)

* etiolog

Genetic Factors	• Exact mode of inheritance is unknown • Monozygotic twins (x8) • +ve family history (8% fathers | 14% brothers)
Endocrine Factors	• Deficient androgenic stimulation (production, conversion, or sensitivity) • Increased maternal progesterone exposure (x5)
Environmental Factors	• Maternal exposure to estrogenic substances (in pesticides, milk, plastic linings of metal cans, and pharmaceuticals)

classification → all need surgery



Distal (50%)

Middle (30%)

Proximal (20%)

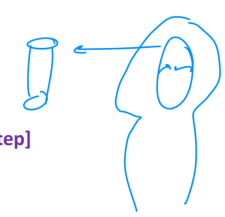
Tx easy and good function + cosmatic

- **Best timing recommendation:**
  - before 18 months of age (minimizes psychological impact of genital surgery)

- **Hormone manipulation preoperative:**
  - penile size can be increased by..
    - weekly IM testosterone or hCG
    - or topical testosterone or DHT

Surgery

Tube urethroplasty
urethral plate is tubularized to neourethra [main step]



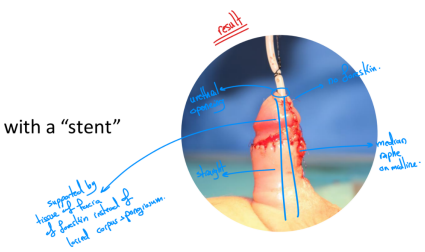
+

- **Corpus spongioplasty:** deficient corpus spongiosum is compensated by fascia (preputial or dartos fascia) to support the neourethra
- **Straightening phalloplasty:** chordee is released
- **Glansplasty:** glans defect is corrected
- **Circumcision:** dorsal foreskin is removed
- **Phalloplasty:** phallic torsion is corrected

Postoperative care

- Neourethra is protected for 1 week with a "stent"
- Simple analgesics
- Oral antibiotic

- **Early complications:**
  - Bleeding
  - Hematoma
  - Infection
  - Breakdown of repair



- **Late complications:**
  - Meatal stenosis
  - Urethrocutaneous fistula (UCF)
  - Persistent chordee
  - Urethral stricture
  - Urethral diverticulum