Vesicoureteral reflux

(VUR) is an abnormal backward movement of urine from the bladder into ureters or kidneys.
VUR

- May present before birth as prenatal hydronephrosis, an abnormal widening of the ureter or with a urinary tract infection or acute pyelonephritis.
- Newborns may be lethargic with faltering growth,
- Infants and young children typically present with pyrexia, dysuria, frequent urination, malodorous urine and GIT symptoms
- 18% in children
- 85% girls
- 15% - first signs before 6 months of life
- Suspect where a child presents with a urinary tract infection
- When suspected anatomical causes should be excluded first.
- Increases the frequency of UTI's,
- Increases the risk of damage to upper urinary structures.
VUR

1. Anatomically ureters enter the urinary bladder obliquely and run submucosally for some distance.

2. Ureter's muscular attachments, helps secure and support them posteriorly.

1 i 2 together - produce a valvelike effect that occludes the ureteric opening, during storage and voiding of urine.

Failure of this mechanism cause retrograde flow of urine.

Hardikar syndrome can include Vesicoureteral reflux, hydronephrosis, cleft lip and palate, intestinal obstruction
VUR

1. Primary VUR

- Insufficient submucosal length of the ureter relative to its diameter causes inadequacy of the valvular mechanism. (ratio diameter:length 1:5).
- Congenital defect/lack of longitudinal muscle of the intravesical ureter resulting in an ureterovesicular junction (UVJ) anomaly.
- nerve prolapsy – patients with MMC, anorectal disorders

2. Secondary VUR

- The valvular mechanism is intact and healthy to start with but becomes overwhelmed by raised vesicular pressures associated with obstruction on the ureterovesical junction. The obstructions may be:
  - Anatomical: posterior urethral valves; urethral or meatal stenosis. (mainly treated surgically).
  - Functional: Bladder instability, neurogenic bladder and non-neurogenic bladder.
- Urinary tract infections may cause reflux due to the elevated pressures associated with inflammation.
International Classification of Vesicoureteral Reflux

Grade I – reflux into non-dilated ureter

Grade II – reflux into the renal pelvis and calyces without dilatation

Grade III – mild/moderate dilatation of the ureter, renal pelvis and calyces with minimal blunting of the fornices

Grade IV – dilation of the renal pelvis and calyces with moderate ureteral tortuosity

Grade V – gross dilatation of the ureter, pelvis and calyces; ureteral tortuosity; loss of papillary impressions
VUR diagnosis

- Nuclear cystogram (RNC)
  - is preferred for subsequent evaluations
- Fluoroscopic voiding cystourethrogram (VCUG)
  - the method of choice for grading and initial workup, while RNC as there is less exposure to radiation. A VCUG and abdominal ultrasound should be performed in these cases
- x-ray examination during urination
- Ultrasonic cystography
- Abdominal ultrasound.
- Early diagnosis in children with VUR and UTI (urinary tract infection) or associated acute pyelonephritis are more likely to develop permanent renal cortical scarring than those children without VUR.
VUR – diagnosis - whom? When?

- infants diagnosed during pregnancy with urine blockage affecting the kidneys
- children younger than 5 years of age with a UTI
- children with a UTI and fever, called febrile UTI, regardless of age
- males with a UTI who are not sexually active, regardless of age or fever
- children with a family history of VUR, including an affected sibling
- Girls with UTI under 6 month of life.
VUR

- The higher the chance of spontaneous resolution - The younger the age of the patient and the lower the grade at presentation.
- Most of grade I & II cases of VUR will resolve spontaneously.
- Approximately 50% of grade III cases and a lower percentage of higher grades will also resolve spontaneously.
- The goal of treatment is to minimize infections and renal scarring.
- Minimizing infections is primarily done by prophylactic antibiotics in newborns and infants with VUR.
- Habbits – children holding their bladder or constipated have a greater number of infections.
- If progressive renal scarring occurs - surgical interventions.
VUR

- Medical management is recommended in children with grade I-III VUR as most cases will resolve spontaneously, sometimes in patients with Grade IV VUR especially in younger patients or those with unilateral disease.

- Only at infants Grade V VUR may succeed medical management before surgery, if its occur in older patients surgery is the only option.

  1. **Endoscopic injection**

  - involves applying a gel around the ureteral opening to create a valve function and stop urine from flowing back up the ureter. (gel consists of two types of sugar-based molecules called dextranomer and hyaluronic acid. e.x. Deflux and Zuidex).

  2. **Medical Treatment**

    - low dose antibiotic prophylaxis - administered nightly at half the normal therapeutic dose f.ex. Amoxicillin or ampicillin, co-trimoxazole, Nitrofurantoin, Nalidixic acid, Bactrim, Trimethoprim, Cephalosporins
VUR – prevention of UTI

- Urine cultures are performed monthly/weekly to exclude breakthrough infection.
- Repeat radiological investigations
- Good perineal hygiene,
- To treat bladder dysfunction with the administration of anticholinergics
VUR treatment

A surgical approach is necessary:
- breakthrough infection results despite prophylaxis
- non-compliance with the prophylaxis
- if the VUR is severe (Grade IV & V),
- there are pyelonephritic changes
- congenital abnormalities.
- failure of renal growth
- formation of new scars,
- renal deterioration
- VUR in girls approaching puberty.
Current recommendations from the American Urological Association include the following:

- children younger than 1 year of age—continuous antibiotics should be used if a child has a history of febrile UTI or VUR grade III through V that was identified through screening

- children older than 1 year of age with BBD—continuous antibiotics should be used while BBD is being treated

- children older than 1 year of age without BBD—continuous antibiotics can be used at the discretion of the health care provider but is not automatically recommended; however, UTIs should be promptly treated
VUR surgical treatment

1. endoscopic (STING/HIT procedures);
2. laparoscopic;
3. open procedures (Cohen procedure, Leadbetter-Politano procedure).
Hydronephrosis

- obstruction of the free flow of urine from the kidney due to distension and dilation of the renal pelvis and calyces
- Untreated, it leads to progressive atrophy of the kidney
- The most common urinary tract disorder in childhood
- Range boys twice more
- 10% bilateraly
Hydronephrosis Causes

- Structural abnormalities of the junctions between the kidney, ureter, and bladder can occur during fetal development.
- Other structural abnormalities could be caused by injury, surgery, or radiation therapy.
- Abnormally placed vein, artery, or tumor.
- Bilateral compression of the ureters can occur during pregnancy - enlargement of the uterus, changes in hormone levels
- Other sources of obstruction - kidney stones, blood clots, or retroperitoneal fibrosis.
- The obstruction may be either partial or complete
- Also as a result reversing flow of urine from the bladder back into the kidneys - impaction of feces in the colon, neurological dysfunction or other muscular disorders
Hnph Causes

1. Internal
   - Obstruction of ureteropelvic junction
   - Hypoplastic proximal part of ureter
   - 'high' attachment of ureter in renal pelvis

2. External
   - Inappropriate vessels, fibres,
   - Isthmus of a Horseshoe kidney

3. Second
   - VUR
   - Anatomical obstruction of ureterovesical junction
   - Posterior urethral valves, Urethrocoele
Symptoms of hydronephrosis

- depend upon whether the obstruction is acute or chronic, partial or complete, unilateral or bilateral.
- intense pain in the flank area (between the hips and ribs).
- nausea and vomiting may also occur.
- can lead to the development of additional stones,
- Lead to UTI - fever, and blood or pus in the urine
- Blood tests may show impaired kidney function (elevated urea or creatinine) or electrolyte imbalances such as hyponatremia or hyperchloremic metabolic acidosis.
- Urinalysis may indicate an elevated pH due to the secondary destruction of nephrons within the affected kidney.
- Physical examination may detect a palpable abdominal or flank mass caused by the enlarged kidney.
Hydronephrosis

- Prenatal diagnosis is possible—routine screening ultrasounds obtained during pregnancy.
- Imaging studies—an intravenous urogram (IVU), ultrasound, CT or MRI—IVU is useful for assessing the anatomical location of the obstruction.
- Antegrade or retrograde pyelography
- Many stones are not visible on plain X ray examination
- Whittaker's test (or pressure perfusion) test.- the collecting system of the kidney is accessed directly through the skin (percutaneously)
- A renal pelvis greater than 10mm in a neonate is considered abnormal and suggests significant dilation
Hydronephrosis

- many cases of prenatal hydronephrosis resolve spontaneously.
- the first few days after birth an imaging study this early may miss some cases of mild hydronephrosis due to the relative oliguria of a newborn
- voiding cystourethrogram (VCUG) – VUR, posterior urethral valves
- Nuclear imaging study such as a MAG-3 scan - If obstruction is suspected-such as a ureteropelvic junction (UPJ) or ureterovesical junction (UVJ)
- RNSC – f.ex. GFR <40% or decrease for 5% in every other examination lead to nephrectomy
Hydronephrosis treatment

- The goal - removal of the obstruction and drainage of the urine that has accumulated behind the obstruction.
- The specific treatment depends upon where the obstruction lies, and whether it is acute or chronic.
- Acute obstruction of the upper urinary tract is usually treated by the insertion of a nephrostomy tube.
- Chronic upper urinary tract obstruction is treated by the insertion of a ureteric stent or a pyeloplasty.
- Lower urinary tract obstruction is usually treated by insertion of a urinary catheter or a suprapubic catheter.
- Surgery is not required in all cases !!!!!
- Plastic of a kidney's pelvic - Hyness -Anderson, Y-V, Culpa, Scardino-Prince'a
Posterior urethral valves - PUV

- an obstructive developmental anomaly in the urethra and genitourinary system of male newborns.

- posterior urethral valve is an obstructing membrane in the posterior male urethra

- It is the most common cause of bladder outlet obstruction in male newborns.

- Varies in degree, with mild cases followed conservatively.

- More severe cases can cause renal and respiratory failure (lung underdevelopment as result of low amniotic fluid volumes)
PUV

- **Type I** - Most common type; due to anterior fusing of the plicae colliculi, mucosal fins extending from the bottom of the verumontanum distally along the prostatic and membranous urethra

- **Type II** - Least common variant; vertical or longitudinal folds between the verumontanum and proximal prostatic urethra and bladder neck

- **Type III** - Less common variant; a disc of tissue distal to verumontanum, also theorized to be a developmental anomaly of congenital urogenital remnants in the bulbar urethra
PUV diagnosis

- Ultrasound - Features suggest are bilateral hydronephrosis, a thickened bladder wall with thickened smooth muscle trabeculations, and bladder diverticula.
- Voiding cystourethrogram (VCUG) is more specific for the diagnosis. Vesicoureteral reflux is also seen in over 50% of cases.
- Cystoscopy- direct visualization
- Cystosonography
PUV treatment

- Fetal surgery is a high risk procedure reserved for cases with severe oligohydramnios, to try to limit the associated lung underdevelopment, or pulmonary hypoplasia (risk of limb entrapment, abdominal injury, and fetal or maternal death)

- *in utero* intervention- infusions of amniotic fluid, serial bladder aspiration, and creating a connection between the amniotic sac and the fetal bladder, or vesicoamniotic shunt.

- Endoscopic valve ablation-
  
  1. *Vesicostomy* followed by valve ablation
  
  2. *Pyelostomy* followed by valve ablation
  
  3. *Primary (transurethral) valve ablation*

- The standard treatment is primary (transurethral) ablation of the valves.
Ureterocele

- Congenital abnormality found in the ureter - the distal ureter balloons at its opening into the bladder, forming a sac-like pouch.
- It is most often associated with a duplicated collection system, where two ureters drain their respective kidney instead of one.
- Simple ureteroceles, a single ureter,
- Mostly female.
- Possibility of diagnosis prenatally (ultrasound)
Ureterocele- classification

- Intravesical
  - Confined within the bladder
- Ectopic
  - Some part extends to the bladder neck or urethra
- Stenotic
  - Intravesical ureterocele with a narrow opening
- Sphincteric
  - Ectopic ureterocele with an orifice distal to the bladder neck
- Sphincterostenotic
  - Orifice is both stenostic and distal to the bladder neck
- Cecoureterocele
  - Ectopic ureterocele that extends into the urethra, but the orifice is in the bladder
Ureterocele symptoms

- Frequent urinary tract infection
- Urosepsis
- Obstructive voiding symptoms
- Urinary retention
- Failure to thrive
- Hematuria
- Cyclic abdominal pain
- Ureteral calculus
- Cobra head sign is seen in radiography.
- In females: Salpingitis, Hydrosalpinx with sepsis or torsion. T.O. mass.
Ureterocele treatment

- Single-system ureterocele-
  - initial management is usually endoscopic incision of the ureterocele,
  - followed by surgical ureteric re-implantation to preserve renal function and prevent reflux.
- Duplex-system ureterocele
  - endoscopic incision of the corresponding ureteric orifice in case of ureteric meatal stricture;
  - upper pole nephrectomy for a poorly functioning unit with ureterectomy
  - or, where there is useful renal function, ureteropyelostomy can be performed.
Urolithiasis

- = renal calculus = nephrolith = kidney stone
- a calculus formed in the kidneys from minerals in the urine
- At children -50% unknown cause
- 25% anatomical disorders of urinary tract in order to urinary obstruction or a UTI
- 25% metabolic disorders
- Originating anywhere in the urinary system, including the kidneys and bladder
- Most pediatric kidney stones are predominantly composed of calcium oxalate; struvite and calcium phosphate stones are less common
Symptoms

- intermittent pain that radiates from the flank to the groin or to the inner thigh. - **renal colic**, caused by peristaltic contractions of the ureter as it attempts to expel the stone
- urinary urgency
- restlessness,
- hematuria,
- sweating, nausea, and vomiting
- Painful urination.
- Goldflame syndrome +
Ulth risk factors

- Dehydration from low fluid intake
- High dietary intake of animal protein, sodium, refined sugars, fructose and high fructose corn syrup, oxalate, grapefruit juice, and apple juice
- High suplementation of calcium
- Intake fluoridated tap water
- High intake of Vit C
- Underlying metabolic condition: distal renal tubular acidosis, Dent's disease, hyperparathyroidism, primary hyperoxaluria, medullary sponge kidney.
- More common in people with Crohn's disease; associated with hyperoxaluria and malabsorption of magnesium.
- With a 24-hour urine collection. The urine is analyzed for features that promote stone formation.
Urthlth diagnosis

- physical examination,
- Urinalysis (blood, WBC, crystals)
- 24 hour urine collection to measure total daily urinary volume, magnesium, sodium, uric acid, calcium, citrate, oxalate and phosphate;
- radiographic studies
  - Xray (may not detect all types of stones ex. Cystine calculi are radiodense, calcium stones are radiolucent)
  - CT- detect almost all types of stones
  - ultrasound - presence of hydrouphrosis if the stone is blocking the outflow of urine.
- IVP- intravenous pyelogram- (contrast to vein and X ray)
- Chemical analysis of collected stones
Urith treatment

1. Prevention
   - High hydration
   - Elimination diet

2. Pharmacological
   - Urine alkalinization
   - Diuretics - ex. Thiazids
   - Allopurinol - interferes with the production of uric acid in the liver
   - Pain management
Urlth surgery

- **ESWL** - extracorporeal shockwave lithotripsy
  - noninvasive technique, most is carried out when the stone is present near the renal pelvis, upper ureter.
- **PCNL** - percutaneous nephrolithotomy
  - flexible ureteroscopy has been adapted through the percutaneous nephrostomy
  - Dedicated to large or complicated stones.
- **URS or URLS** - Ureterorenoskopy
  - dedicated to medium and lower parts of urinary tract - can directly remove the stone
- **Laser lithotripsy**
- **Ureteroscopic surgery**
  - stent replacement - double j, pigtail,
  - basket extraction,
- **Open surgery**
Urinary Tract disorders

- 10% infants is born with UT disorders
- Most common in all inborn disorders
- 45% of renal failure is connected with UT disorders
- Association: VATER or VACTERL
  - Vertebral
  - Anal
  - Cardio
  - Trachea
  - Esophagus
  - Ren
  - Limb
UT disorders classification

- **Number** - agenesis uni – or bilateral, excess kidney
- **Localization** - Pelvis, Thorax, intraperitoneal,
- Collapsed kidney – different vascularization!!!!
- **Shape** - Horseshoe kidney - increasing risk of nephroblastoma!!, S or L shaped, croosed kidneys with or without fusion of ureters
- **Structure** - aplasia, hipoplasia, polycystic, sponge kidney, simple cystis (percutaneus injection or partial excision),
- **Roatation** - ventricular or dorsal

All those increase risk of UTI, urolithiasis or hydronephrosis!!!
Policystic kidney diseases

- the presence of multiple cysts ("polycystic") typically in both kidneys;
- The cysts are numerous and are fluid-filled, resulting in massive enlargement of the kidneys.
- The disease can also damage the liver, pancreas and, in some rare cases, the heart and brain.
- The two major forms of polycystic kidney disease are distinguished by their patterns of inheritance.
- Polycystic kidney disease is one of the most common life-threatening genetic diseases, affecting an estimated 12.5 million people worldwid
Autosomal dominant polycystic kidney disease (ADPKD)

- Autosomal dominant
- Adult type
- is characterized by progressive cyst development and bilaterally enlarged kidneys with multiple cysts.
- There are three genetic mutations in the PKD-1, PKD-2, and PKD3 gene with similar phenotypical presentations. (chromosome 16 and 4)
- Cyst formation begins in utero from any point along the nephron. As the cysts accumulate fluid, they enlarge, separate entirely from the nephron, compress the neighboring renal parenchyma, and progressively compromise renal function.
- Provides to nephrectomy
**Autosomal recessive polycystic kidney disease (ARPKD)**

- Autosomal recessive
- Infatile type
- is the lesser common of the two types of PKD,
- is typically identified in the first few weeks after birth.
- Unfortunately, resulting hypoplasia results in a 30% death rate in neonates
- kidneys retain their shape but are larger than the normal anatomical range with dilated collecting ducts from the medulla to the cortex.
- Non-cured outcome – leads to transplantation of kidney or liver or both
Medullary sponge kidney

- is a congenital disorder
- characterized by cystic dilatation of the collecting tubules in one or both kidneys.
- increased risk for kidney stones and urinary tract infection.
- Wide symptomes spectrum
  - increased chronic kidney pain, renal colic (flank and back pain), UTIs and complications, nephrolithiasis,
  - most cases are asymptomatic
  - hematuria
- Detected by renal ultrasonography or abdominal x-ray
- Leads to renal failure and nephrectomy
Ureter's disorders

1. Number – agenesis, hipoplasia, dupplex, fissus (Y)
2. Structure- giant, vanished, diverticuli,
3. Inappropriate uretrovesical juncion ora ureteropelvic junction
   - leads to VUR, nephrolithiasis, hydronephrosis
4. Inappropriate localization
   - behind v.c.i. Or v. iliaca intera
5. Ureterocele
Duplicated ureter
Ureter's duplication

- It is the most common renal abnormality
- Ureteral duplication is either:
  1. **Partial** - the two ureters drain into the bladder via a single common ureter. Rarely clinically significant.
  2. **Complete** - the two ureters drain separately.
     - one ureter opening normally into the bladder
     - the other being ectopic, ending in the vagina, the urethra or the vulval vestibule
- Cause UTI, hydronephrosis, VUR,
- Urinary incontinence in females occurs in cases of ectopic ureter entering the vagina, urethra or vestibule.
Dx ureter's classification

1. **Duplex kidney**: two separate pelvicalcyecal systems draining a single renal parenchyma

2. **Duplex collecting system**: a duplex kidney draining into:
   - single ureter: i.e. duplex kidney's duplication pelvicalyceal systems uniting at the pelviureteric junction (PUJ)
   - bifid ureter (ureter fissus): two ureters that unite before emptying into the bladder
   - double ureter (complete duplication)

3. **Bifid collecting system**: refers to a duplex kidney with the two separate pelvicalcyecal collecting systems uniting at the PUJ or as bifid ureters

4. **Double/duplicated ureters** (or collecting system): two ureters that drain separately into the bladder or genital tract