Pelviureteric Junction (PUJ) Obstruction

Definition: a restriction to the flow of urine, from the renal pelvis to the ureter, which, if left uncorrected, will lead to progressive renal deterioration.
Epidemiology: Males > females (5:2 ratio). In unilateral cases, left to right side ratio (5:2). Bilateral PUJ obstruction is present in 10% to 40% of cases. No clear genetic inheritance.
Etiology
Congenital or Acquired Obstruction.
The acquired causes are (Trauma, surgery , renal stone , pelvic tumor or cyst, compression by retroperitoneal tumor)
The most common is the Congenital PUJ Obstruction which may be:
Intrinsic causes defect in the circular smooth muscle of the UPJ or an alteration of the collagen fibers results in an aperistaltic segment of ureter at the PUJ **(** a narrowed segment of the ureter at the UPJ that is probe patent) ,
Extrinsic
An aberrant or accessory, vessel is the most common cause of extrinsic UPJ obstruction
Secondary PUJ Obstruction
PUJ obstruction may also be seen with severe vesicoureteral reflux (VUR); these conditions coexist in 10-- 25% of cases. The ureter elongates and develops a tortuous course.
Presentation
Vary according to the age. Most cases diagnosed prenataly by sonography. Later , the pain and vomiting are the most common symptoms. Haematurea and Abdominal mass may be seen. A few patients may have complications as stone, infection, trauma to enlarged kidney, or rarely hypertension. Many of adult patients may be diagnosed incidently by sonoghraphic examination for other problems .
Diagnosis
Renal ultrasound shows renal pelvis dilatation in the absence of a dilated ureter.
IVU demonstrates delay of excretion of contrast and a dilated pelvicalyceal system.
Abdominal CT to exclude a small, radiolucent stone, urothelial TCC, or retroperitoneal pathology which may be the cause of the obstruction at the PUJ.
MAG3 renography with administration of frusemide to establish a maximum diuresis is the definitive diagnostic test for PUJO. Radioisotope accumulates in the renal pelvis, and following iv frusemide it continues to accumulate (a 'rising' curve). **Retrograde pyelography** to establish the exact site of the obstruction
Magnetic Resonance Imaging for evaluating renal blood flow, anatomy, and urinary excretion
Pressure-Flow Studies (Whitaker studies) differential pressure between kidney and bladder could be an indicative of obstruction to the kidney.

SURGICAL REPAIR

Indicated in (1) symptomatic PUJO (2) decrease renal function les than 45% (3) Complicated PUJO (4) massive hydronephrosis.
Endoscopic treatment of a PUJO is called an endopyelotomy (or pyelolysis). Various techniques have been described, but the essential principle is the same—full-thickness incision through the obstructing proximal ureter, from within the lumen of the ureter down into the peripelvic and periureteral fat, using a sharp knife or HolmiumiYAG laser. The incision is stented for 6 weeks to allow re-epithelialization of the PUJ. Relatively minimally invasive. Generally not used for PUJO >2cm in length.
The incision may be made percutaneously or by a retrograde approach via a rigid or flexible ureteroscope, or by using a specially designed endoplyelotomy balloon (the Acucise technique).

Pyeloplasty Common techniques include (1) dismembered pyeloplasty (also known as the Anderson-Hynes pyeloplasty: the narrowed area of PUJ is excised, the proximal ureter is spatulated and anastomosed to the renal pelvis), (2) flap pyeloplasty (3) Y-V-plasty (Foley).
• Open Pyeloplasty : has success rates of 95%, and may also be used after endopyelotomy failure or as a first line technique.
• Laparoscopic pyeloplasty has the advantage of accelerated patient recovery.
Nephrectomy done if split renal function less than 10% or sever pyonephrosis

Ureteral anomalies

are some of the most significant anomalies in all of pediatric urology because they directly affect overall renal function
A duplex kidney is one that has two separate pelvicalyceal systems. A duplex kidney has an upper pole and a lower pole. The ureters may join at any point. If they join at the level of the ureteropelvic junction, the configuration is termed a ***bifid system***. If the ureters join more distally but are still proximal to the bladder level, is termed ***bifid ureters***.
Double ureters are ureters that drain their respective poles and empty separately into the genitourinary tract. This represents a complete duplication.
Ectopic ureter : a ureter whose orifice terminates even more caudally, such as in the urethra or outside of the urinary tract. Continuous incontinence in a girl with an otherwise normal voiding pattern after toilet training is the classic symptom of an ectopic ureteral orifice. While in boy not cause incontinence but may present with recurrent attacks of orchitis.

Ureteroceles: is a cystic dilatation of the terminal ureter between the superficial and deep layers of the trigon. May be either intravesical or Ectopic. It is due to delayed or incomplete canalization of the uretral bud leading to obstruction and expansion of the uretral bud before absorption in to the urogenital sinus. Females/ male ratio (4:1 ratio). 10% are bilateral. 80% of all ureteroceles arise from the upper poles of duplicated systems. Single-system ureteroceles are sometimes called simple ureteroceles and are usually found in adults. If the ureterocele is large enough, it can obstruct the bladder neck or even the contralateral ureteral orifice and result in hydronephrosis of that collecting system. It is diagnosed by sonography. Excretory urography often demonstrates the characteristic cobra-head (or spring-onion) deformity, an area of increased density similar to the head of a cobra with a halo or less dense shadow around it. Cystography demonstrates the ureterocele in the bladder. It appears as a smooth, broad-based filling defect located near the trigone.
Treatment:
Endoscopic treatment of ureteroceles by incising the ureterocele at the base.
Surgical treatment by excision end reimplantation of the ureter.

VESICOURETERIC REFLUX (VUR)

is the retrograde flow of urine from the bladder into the upper urinary tract with or without dilatation of the ureter, renal pelvis, and calyces. It can cause symptoms and may lead to renal failure (reflux nephropathy).
The overall incidence of reflux is about 10%.Reflux is found in up to 70% of infants who present with UTIs. the vast majority (85%) of reflux occurs in females. The reflux is usually high grade and bilateral in boys, compared with girls. Reflux is the most common inherited anomaly of the genitourinary tract, and siblings of patients with reflux are at much greater risk of having reflux than the normal population.

Pathophysiology

Reflux is normally prevented by low bladder pressures, efficient ureteric peristalsis, and the ability of the vesicoureteric junction (VUJ) to occlude the distal ureter during bladder contraction. This is assisted by the ureters passing obliquely through the bladder wall (the 'intramural' ureter), which is 1-2cm long. Normal intramural ureteric length to ureteric diameter ratio is 5:1. VUR of childhood tends to resolve spontaneously with increasing age because as the bladder grows, the intramural ureter lengthens.

Etiology

***Primary*:** a primary or functional : is a congenital anomaly of the UVJ where in a deficiency of the longitudinal muscle of the intravesical ureter results in an inadequate valvular mechanism and the intramural length of the ureter is too short (ratio <5:1).

Secondary *t*o some other anatomical or functional problem:
• Bladder outlet obstruction (BPH, DSD due to neuropathic disorders, posterior urethral valves, urethral stricture) which leads to elevated bladder pressures.
• Poor bladder compliance or the intermittently elevated pressures of detrusor hyperreflexia (due to neuropathic disorders —e.g. spinal cord injury, spina bifida).
• Iatrogenic reflux following TURP or TURBT (a tumour overlying the ureteric orifice)—this is rare; ureteric meatotomy (incision of the ureteric orifice) for removal of ureteric stones at the VUJ; following incision of a ureterocele; ureteroneocystostomy; post pelvic radiotherapy.
• Inflammatory conditions affecting function of the VUJ: TB, schistosomiasis, UTI.

Associated disorders

VUR is commonly seen in duplex ureters.
The incidence of VUR associated with UPJ obstruction ranges from 10% to 25% .
Cystitis can cause VUR through bladder inflammation, reduced bladder compliance, increased pressures, and distortion of the VUJ. Coexistence of UTI with VUR is a potent cause of pyelonephritis—reflux of infected urine under high pressure causes reflux nephropathy, resulting in renal scarring, hypertension, and renal impairment.

Clinical Presentation

- VUR may be symptomless, being identified during VCUG, IVU, or renal ultrasound (which shows ureteric and renal pelvis dilatation) done for some other cause.
- UTI symptoms.
- Loin pain associated with a full bladder or immediately after micturition.
- Failure to thrive and lethargy are worrisome signs in newborns,

Symptoms of recurrent UTI or of loin pain may have been present for many years before the patient seeks medical advice. A high index of suspicion is required.

Investigations

The definitive test for the diagnosis of VUR is cystography. VUR may be apparent during bladder filling or during voiding (voiding cystourethrography, VCUG —also known as micturating cystourethrography, MCUG).
Urodynamics establishes the presence of voiding dysfunction (e.g. DSD) if this is suspected from the clinical picture.
If there is radiographic evidence of reflux nephropathy check blood pressure, check the urine for proteinuria, measure serum creatinine, and arrange a ^{99m}Tc-DMSA isotope study to assess renal cortical scarring and determine split renal function.

The International Classification System: By VCUG

Grade I Contrast into non-dilated ureter
Grade II Contrast into renal pelvis and calyces; no dilatation
Grade III Mild dilatation of ureter; pelvis, and calyces
Grade IV Dilated and slightly tortuous ureter; moderate dilatation of pelvis and blunting of calyces
Grade V Severe ureteric dilatation and tortuosity; gross dilatation of pelvis and calyces

Management

Medical Management

Medical management consists of continuous low-dosage prophylactic antibiotic therapy until the expected resolution of reflux occurs, Improving toilet hygiene and bladder emptying by means of timed voids, double voiding, proper perineal wiping, and elimination of constipation Bladder dysfunction should also be treated with anticholinergics. Periodic urine cultures are obtained every 3 months to evaluate for breakthrough infections.

Surgical Management

Reimplantation of the ureter with antireflux procedures
Typical indications:

1. Breakthrough UTIs despite prophylactic antibiotics
2. Noncompliance with medical management
3. Severe grades of reflux (grade IV or V), especially with pyelonephritic changes
4. Failure of renal growth, new renal scars, or deterioration of renal function on serial ultrasounds or scans
5. Reflux that persists in girls as full linear growth is approached at puberty
6. Reflux associated with congenital abnormalities at the UVJ (e.g., bladder diverticula)

Secondary reflux: treat the underlying cause