

Disorders of the ureter & ureteropelvic junction

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The ureter is a complex functional conduit carrying urine from the kidneys to the bladder. Any pathologic process interfere with this activity can cause renal abnormalities, the most common sequels being **hydronephrosis & infection.**

disorders of the ureter can be classified as **congenital or acquired.**

Vesicoureteral reflux

Is defined as non physiological back-flow urine from urinary bladder into the ureter or renal pelvis or calyces ,

it is either primary which is due to congenital anomaly of the ureterovesical junction ,or secondary which is caused by anatomical or functional bladder outflow obstruction.

Under normal circumstances, the ureterovesical junction allows urine to enter the bladder but prevents urine from regurgitating into the ureter, particularly at the time of voiding.

In this way the kidneys are protected from **high pressure** in the bladder & from **contamination** by infected vesical urine.

when this physiological valve is incompetent, the chance of development of UTI is significantly increased, & pyelonephritis becomes inevitable.

CAUSES OF REFLUX:

1- PRIMARY

2- SECONDARY

A- ANATOMICAL REFLUX

B- FUNCTIONAL REFLUX

Causes of vesicoureteral reflux.

A -Primary reflux, is a congenital anomaly of the ureterovesical junction in which deficiency of the longitudinal muscle of the intravesical ureter results in inadequate valvular mechanism. the normal ratio of the tunnel length to ureteral diameter is 5:1 in normal children without reflux.

B – secondary reflux which is either due to:

---- **Anatomical**. Like bladder outlet obstruction & its consequent elevated intravesical pressures. The most common anatomical causes are:

(1) posterior urethral valves. Which are associated with reflux in about 50% of affected boys.

(2) ureteric duplication ,

(3) ectopic ureteric orifice and

(4)urethrocele (rarely causes vur)

--- **Functional causes** : include neurogenic bladder & dysfunctional bladder.

Grading of vesicoureteral reflux.

Grade 1 reflux into the non dilated ureter.

Grade 2 reflux into the pelvis & calyces without dilatation.

Grade 3 reflux causes mild dilatation of the ureter renal pelvis & calyces.

Grade 4 reflux causes moderate dilatation of the ureter renal pelvis & calyces.

Grade 5 reflux causes severe dilatation of ureter, pelvis & calyces.

Incidence.

- About 1-2 % of all children have vur.
- In newborns the incidence of reflux is higher in boys than girls but later in life the girls are 4- 6 times more frequent than boys .
- Vesicoureteral reflux occurs in 50% of children with UTI .

Diagnostic work up;

A basic diagnostic study is composed of;

- detailed history ,
- physical examination
- urine analysis & urine culture ,
- renal function test,
- diagnosing imaging by x ray (voiding cystourethrogram),
- us ,and
- radionuclide cystography like dimercaptosuccinic acid(DMSA) study ,
- Cystoscopy.

Clinical findings

Are either

- symptom related to reflux like symptomatic or asymptomatic pyelonephritis
- Symptoms of cystitis
- Symptoms of renal dysfunction .

- symptoms related to underlying diseases like BOO (bladder outlet obstruction) or spinal cord disease
- Most patients with reflux present initially with symptoms of UTI whether lower uti or upper uti
- newborns typically show nonspecific symptoms like failure to thrive & lethargy

By examination

- General look of the child .
- Other systemic causes like spinal cord insult.
- Renal tenderness may be present,
- palpation & percussion of suprapubic area may reveal distended bladder secondary to BOO

.

Laboratory finding.

- Urine exam ;

Pyurea or hematuria.

- The blood urea and serum creatinine may be elevated in the advanced stage of renal damage, but it may be normal even when the degree of reflux & hydronephrosis is marked specially if more severe in one side (pop off phenomenon).

- urine and blood culture specially in septicemic patients.

x-Ray finding.

- **abdominal ultrasound:**

-- **Plain film(KUB)** may reveal evidence vertebral anomalies which points to the neurologic deficit.

-- **Excretory urograms** may be normal, or dilatation of whole or part of ureter or hydronephrosis.

-- **voiding cystourethrography** .

-- **Radionuclide cystography.DMSA**

Cystoscopy. Is very important step in the work up which may show ,Puv ,Ureteral duplication , ureterocele, or ectopic ureteral orifices. These findings imply the possibility of reflux.

Treatment ;

The objective of treatment of vur is to:

- keep normal renal growth
- prevent renal parenchymal damage (reflux nephropathy).
- keep the patient free of infections.

The therapeutic options are:

- conservative medical treatment by antibiotic ,
- interventional therapy by (cystoscopy ,laparoscopy ,and open surgery).

Treatment.

A -Medical treatment.

It has become increasingly apparent that medical treatment will be effective for many children with reflux.

Medical management consists of low-dose prophylactic antibiotic continued until reflux resolves.

In toilet trained children bladder emptying by timed voids, double voiding, help to achieve the goals of medical management.

Urine cultures are obtained every 3 months to evaluate breakthrough infections,

& yearly radiologic studies are also necessary.

B –surgical management.

indications of antireflux surgery include:-

1- breakthrough UTI despite prophylactic antibiotic.

2- noncompliance with medical management.

3- severe reflux grade 4 or 5 ??????.

4- deterioration of renal function .

5- reflux persists to puberty specially in girls.

6- reflux associated with congenital abnormalities such as bladder diverticulum.

Interventional therapy ;

----open surgery ; although different methods had been made but all share the basic principle of lengthening the intramural part of the ureter by submucosal embedding.

----Laparoscopic treatment ; is Not superior than open surgery.

----Endoscopic therapy .

with the availability of the biodegradable substances, endoscopic submucosal injection of this (bulking agents) have become an alternative to long-term antibiotic prophylactic and surgical treatment of vuv.

Note; In boys with low grade vuv only circumcision can resolve the problem

Duplication of the ureter

Complete or incomplete duplication of the ureter is one of the most common congenital malformation of the urinary tract.

it is seen in about 0,9 % of autopsy.

Incomplete (Y) type of duplication is caused by branching of the ureter before it reach the metanephric blastema (kidney in future).

Disorders of the peristalsis may occur near the point of union causing pain .

Complete duplication of ureter in which 2 ureteral buds lead to formation of 2 separate ureters & 2 separate renal pelves.

The ureter draining the upper segment ending in the bladder medial & inferior to the ureter draining the lower renal segment. So the upper segment ureter is longer and usually obstructed and the lower segment ureter is shorter and refluxing due to short intravesical ureter.

Clinical finding.

Patients may be asymptomatic or have recurrent UTI.

In females, the ureter of the upper pole may be ectopic with an opening distal to the external sphincter. Such patients have classical presentation of incontinence with constant dribbling & normal voiding pattern.

In males the opening **always** proximal to the external sphincter so incontinence does not occur.

----- Ultrasound,

----- excretory urography &

----- voiding cystourethrography to asses reflux in the lower pole ureter.

-----Tc 99 dimercaptosuccinic acid DMSA

Treatment

is that of reflux if present ,

If upper pole ureter is ectopic or obstructed the treatment is by surgery like:

----- pyeloureterostomy or

-----Ureteroureterostomy or

-----ureteric reimplantation .

----- Heminephrectomy in cases of non functioning segment.

Ureterocele

cystic dilatation of the intramural part of the ureter, due to incomplete canalization of the ureteric bud . It may be either

-intravesical

- Or ectopic,

Ureterocele 7 times more common in girls than boys &

in 10% is bilateral.

Clinical finding.

Patients commonly present with infection or bladder outlet obstruction or incontinence.

Occasionally ureterocele may prolapsed through the female urethra.

Calculi can develop secondary to urinary stasis.

Ultrasound may be diagnostic,

excretory urography may show cobra head appearance,

Treatment.

Depending on the degree of obstruction & age of presentation and function of the kidney

Vary from transurethral incision of the ureterocele or reimplantation of the ureter or nephrectomy .

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Obstructed megaureter

Obstruction at the ureterovesical junction is 4 times more common in boys than girls. It may be bilateral .

It is clear that in most cases there is no stricture at the ureterovesical junction.

At operation, retrograde catheter or probe can usually be passed through the area of obstruction.

Close observation by fluoroscope reveals failure of the distal ureter to transmit the normal peristaltic wave, resulting in functional obstruction.

Histologic finding include an excess of circular muscle fibers & collagen in the distal ureter that may account for the problem.

Sonography usually show the pathognomonic configuration of a dilated distal ureter, less dilated proximal ureter, **relatively normal appearing renal pelvis, & calyces.**

Treatment.

A period of observation is nearly always appropriate when the diagnosis is made in an asymptomatic patients,

at least 50% of the cases will have spontaneous resolution.

Surgical management may be necessary in a form of ureteric reimplantation & generally it has excellent prognosis.

Obstruction of ureteropelvic junction

(PUJ obstruction)

PUJ obstruction is probably the most common congenital abnormality of the ureter. It is seen more often in boys than girls (5:2 ratio)

Bilateral obstruction in 10-15% of cases.

The exact cause of PUJ obstruction often is not clear.

(1) Ureteric polyps.

(2) valves have been reported.

(3) True stenosis .

(4) hypoplastic proximal ureter .

(5) High origin of the ureter from the renal pelvis

(6) an abnormal relationship of the proximal ureter to an aberrant renal vessels .

Clinical findings.

Vary depending on the patient's age at diagnosis. Recent improvement in prenatal ultrasonography now allow most cases to be diagnosed in utero.

Later, pain & vomiting are the most common symptoms,

hematuria & UTI also may be seen.

A few patients have complications such as **calculi**,

or rarely hypertension.

The diagnosis is made most often by sonography & EU study which show ballooning of the renal pelvis or non functioning kidney.

CT scan also may be needed.

Treatment.

Symptomatic PUJ obstruction should be treated by:

- open pyeloplasty or
- Laparoscopic pyeloplasty.
- robotic assisted laparoscopic pyeloplasty.
- endopyelotomy.
- Nephrectomy may be indicated in late cases.

Acquired diseases of the ureter

Nearly all acquired diseases of the ureter are obstructive in nature. Their clinical manifestations, effects on the kidney, complications, & treatment are similar to those described previously.

The lesions can be broadly categorized as either intrinsic or extrinsic.

Intrinsic ureteral obstruction.

The most common causes are as follow.

- 1 – ureteral stones.
- 2- urothelial tumour.
- 3- chronic inflammatory changes of the ureteric wall (due to tuberculoses or schistosomiasis) leading to contracture or insufficient peristalsis.

Extrinsic ureteral obstruction

- 1- severe constipation, seen primarily in children.
- 2- secondary obstruction due to kinks or fibrosis around redundant ureters. The primary process is either distal obstruction or massive reflux.
- 3- benign gynecological disorders such as endometriosis.
- 4- local neoplastic infiltration associated with carcinoma of the cervix, bladder, or prostate.
- 5- pelvic lymphadenopathy associated with metastatic tumor.
- 6- iatrogenic ureteral injuries, after extensive pelvic surgery or extensive radiotherapy.
- 7- retroperitoneal fibrosis.

Retroperitoneal fibrosis

Chronic inflammatory process involve the retroperitoneal tissue over the lower lumbar vertebrae may engulf & obstruct the ureter.

Causes:-

Are many,

(1)malignant disease (most commonly Hodgkin disease, carcinoma of the breast, & carcinoma of the colon) should always be suspected & ruled out.

(2)Some medications like methysergide, an ergot derivative used to treat migraine headache.

Or (3) idiopathic.

Symptoms.

Are non specific & include low back pain, malaise, anorexia, weight loss, & in severe cases uremia. Infection is uncommon the diagnosis is usually made by excretory urography. There is medial deviation of the ureter with proximal dilatation.

Ultrasound useful not only to the diagnosis but also for monitoring the response to therapy.

Spontaneous regression has been reported, however, treatment is usually surgical. A course of corticosteroid may start first.

Upper urinary tract dilatation without obstruction:

Sometimes dilatation of the urinary tract is caused by non obstructed causes like:

1– reflux

2– residual dilatation in previously obstructed system.

3– associated bacterial infection the dilatation is caused by direct effect by endotoxin on the ureteric muscles

4– prolong polyurea in diabetes insipitus patient