# Congenital abnormalities of the renal pelvis and ureter

## Duplication of a renal pelvis

- Found in about 4% of patients.
- Usually unilateral and left sided.
- The small upper renal pelvis drains the upper group of calyces
- The larger lower renal pelvis drains the middle and lower groups of calyces.

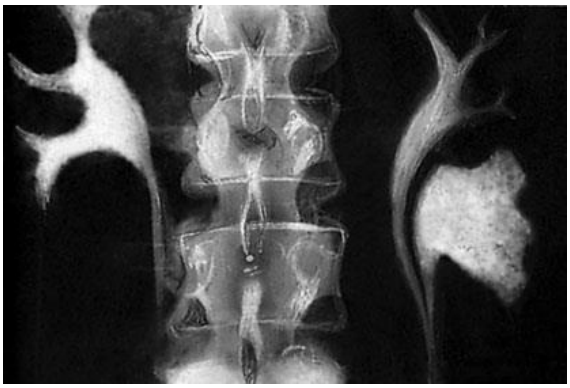
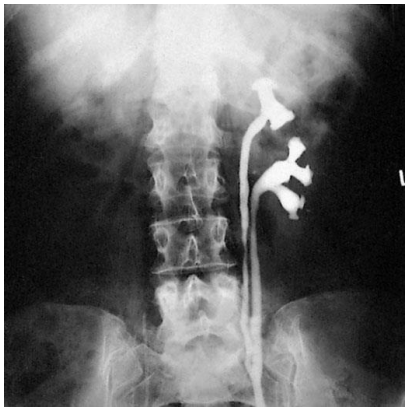
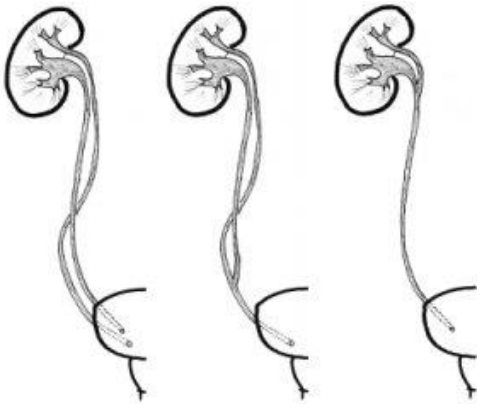


**Figure Urogram showing a left kidney with double pelvis.**

## Duplication of a ureter

- Found in about 3% of urograms.
- The ureters often join in the lower third of their course and have a common ureteric orifice.
- When the ureters open independently into the bladder, the ureter from the upper pelvis opens distal and medial to its fellow.





**Retrograde ureterogram showing a double ureter on the left**

***Clinical features***

- Duplication of the renal pelvis or ureter is often accidentally discovered on renal imaging
- But ***infection***, ***calculus*** formation and ***pelviureteric obstruction*** are more common than in normal kidneys.
- One of the moieties may be **dysplastic** and non-functioning.
- When the two ureters open separately, one may be abnormal in function, position or both.
- In children, this may result in a refluxing lower pole ureter and an upper pole ureter terminating in ***ureterocele***.
- In such cases, the duplex kidney is at risk of infection and/or obstruction.
- An ectopic second ureteric opening is a rarity but it may cause confusing symptoms.
- In the female, an ectopic ureter opens either into the urethra below the sphincter or into the vagina.
- The diagnosis can often be made from the history alone and is confirmed by urography.
- A girl or woman who voids normally but who has dribbled urine for as long as she can remember probably has an ectopic ureteric orifice. The orifice is difficult to see because it is guarded by a valve: it may help to give an intravenous injection of a dye to colour the urine leaking from it.
- In the male patient, the aberrant opening is above the external urethral sphincter so the patient is continent.
- The ectopic ureteric orifice in male may also be at the apex of the trigone, the posterior urethra, in a seminal vesicle or in an ejaculatory duct is likely to be functionally abnormal, and infection is common.

**Notes**

- **Dysplasia** is abnormal development or growth of tissues, organs, or cells.
- **Moiety** means half, part, or portion.

***Treatment***

- Asymptomatic duplication of the kidney is harmless and does not require treatment.
- If one moiety is severely diseased or atrophic, partial nephrectomy is usually simple and effective.
- A refluxing ureter may need reimplanting.
- An ectopic ureter in the female frequently drains hydronephrotic and chronically infected renal tissue, which is best excised.
- Rarely, the incontinence can be cured and renal function preserved by implanting the ectopic ureter into the bladder or contralateral ureter.

**Duplication of the kidneys and ureters**

■ Is relatively common and often asymptomatic and harmless
■ One or both moieties of the duplex may be dysplastic
■ Abnormalities of the insertion of the ureter into the bladder may cause urinary reflux, incontinence or obstruction

## Congenital mega ureter

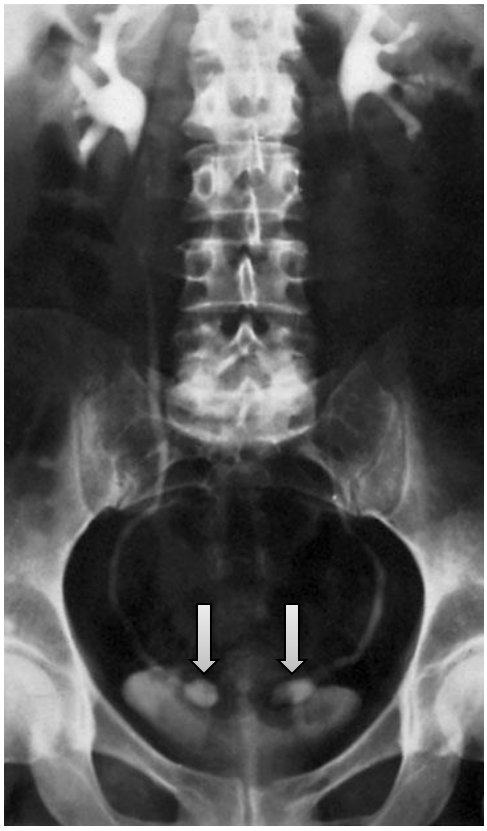
- Congenital megaureter is a rare disease
- May be bilateral and may be associated with other congenital anomalies.
- Functional obstruction at the lower end of the ureter may lead to progressive dilatation and infection.
- The ureteric orifice appears normal and a ureteric catheter passes easily.
- Reflux is not a feature of the untreated condition but is almost inevitable if the ureteric orifice is opened endoscopically.
- Spontaneous improvement can occur but infection or deteriorating function will require refashioning and re-implantation of the affected ureter.

## Post-caval ureter

- The right ureter passes behind the vena cava instead of lying to the right of it.
- If this causes obstructive symptoms, the ureter can be divided and rejoined in front of the cava using a long oblique anastomosis without tension.
- Unusually, the retrocaval portion of the ureter is fibrotic and must be excised.

## Ureterocele

- Ureterocele is a cystic enlargement of the intramural ureter, which is thought to result from ***congenital atresia of the ureteric orifice***.
- Although present from childhood, the condition is often unrecognised until adult life.
- The ‘adder head’ on excretory urography is typical.
- Usually the cyst wall is composed of urothelium only and the diagnosis is confirmed by the cystoscopic appearance of a translucent cyst enlarging and collapsing as urine flows in from above.
- ***Treatment should be avoided*** unless there are symptoms arising from infection and/or stone formation.
- Ureterocele is most common in women; occasionally, the cyst may cause obstruction to the bladder outflow by prolapsing into the internal urethral opening.
- ***Endoscopic diathermy*** incision is usually all that is required for treatment of a symptomatic ureterocele, although a micturating cystogram is advisable to detect postoperative urinary reflux.
- In advanced unilateral cases with hydronephrosis or pyonephrosis, ***nephrectomy*** may be appropriate.



**Adder-head appearance of a bilateral ureterocele**

**بسم الله الرحمن الرحيم**

**رَبِّ اشْرَحْ لِي صَدْرِي (٢٥) وَيَسِّرْ لِي أَمْرِي (٢٦) وَاحْلُلْ عُقْدَةً مِنْ لِسَانِي (٢٧) يَفْقَهُوا قَوْلِي**

**صدق الله العظيم**

**سورة طه**

**د مقداد فؤاد**