**Duplex, ureterocoele and ectopic ureter**

Duplication abnormality 0.8% postmortem

Majority incomplete and asymptomatic

Complete duplications less common (<0.1%) but more often symptomatic

Incomplete duplications bilateral in 40%, complete bilateral in 25%

**Embryologically:**

Arise as a result of second ureteric bud developing adjacent to normal bud. The upper pole ureter is quite close to mesonephric duct and drawn down with duct during fetal growth, whereas the lower pole ureter is under less influence, explaining why the upper pole ureteric opening is always caudal to the opening of lower pole ureter (Meyer-Weigart Law). Degree of dysplasia (usually upper pole) related to degree of ectopia (indicates more extreme penetration of metanephric blastema, with resultant abnormal nephrogenesis)

NB. dysplasia is a histological diagnosis. When referring to x-ray features suggestive of a dysplastic kidney, more correct to use the term hypoplasia

**Classification of associated abnormalities:**

- **Lower pole**
  - VUR
  - Occasionally dysplasia

- **Upper pole**
  - Duplex system ureterocoele*
  - Suprasphincteric ureteric ectopia
    - Boys – vas, seminal vesicle, ejaculatory duct
    - Girls – bladder neck, proximal urethra
  - Infrasphincteric ureteric ectopia
    - Girls only – introitus, distal vagina

**Selected duplication incidences**

- Unilateral incomplete duplex 1:100
- Bilateral incomplete duplex 1:250
- Unilateral complete duplex 1:1000
- Bilateral complete duplex 1:4000
- Unilateral duplex with ectopic ureter 1:10000
- Bilateral duplex with ectopic ureters 1:100000
- Duplex system ureterocoele 1:5000
- Bilateral duplex system ureterocoele 1:50000
- Single system ureterocoele 1:25000
- Bilateral single system ureterocoele 1:250000

**Duplex system ureterocoele**

Incidence 1:5000

80% females

Whites >> blacks

Left > right

10% bilateral
Upper pole moiety affected
May be orthotopic (entirely intravesical), or ectopic (more common; often at bladder neck or rarely level with urethra/introital in girls (caecourethrocoele))
Typically a/w upper pole dysplasia and impaired renal function
Ipsilateral lower pole
   mild VUR in 50% of cases
   occasionally obstruction 2’ to ureterocele itself
   usually preserved renal function
Usually diagnosed prenatally (60%); otherwise UTI, AUR, or rarely prolapse
Readily evident on USS; further investigation includes DMSA to identify dysplasia, and MCUG to identify reflux [reflux involving both kidneys always a/w incomplete duplication]
Management
Complex and depends upon presence of symptoms and associated lower pole and ipsilateral findings
Indications for surgery
   Symptoms
   Preserved ipsilateral upper pole renal function
   Ipsilateral lower pole obstruction
   Bladder outflow obstruction
   Ureterocele prolapse
   Ipsilateral lower pole reflux and UTI
Surgical options (depends on upper pole function)
   (i) Functioning upper pole
      a. Endoscopic ureterocele incision
         Simple, minimally invasive
         Can induce reflux (do it close to bladder wall)
         Reasonable medium term results (some view it as temporising measure)
      b. Ureterocele excision and re-implantation
         For non-dilated units
         Re-implantation of both ureters in Waldeyer’s sheath (typically Cohen cross-bladder technique)
      c. Pyelopyelostomy
         For dilated lower pole moiety
         Ureterocele and distal ureter aspirated and left in situ
   (ii) Non-functioning upper pole
      a. Upper pole nephrectomy
         aka ‘simplified approach’
         Upper pole moiety excised
         Ureterocele and distal ureter aspirated and left in situ
      b. Upper pole nephrectomy, ureterectomy and ureterocele excision
         Gold standard
         Requires 2 incisions
         Risk of damage to bladder neck and vesicovaginal fistula in girls
Reimplantation of ipsilateral lower pole ureter often required

**c. Nephrectomy**
For globally reduced ipsilateral function
Reflexing ureters should be excised, but non-refluxing ureterocoele can be aspirated and left in situ

**Single-system ureterocoele**
Boys > girls
Almost always orthotopic
Prenatal vs. clinical (incidental, UTI, AUR)
Typically renal function is preserved
Management
- Observation
- Surgical intervention for symptoms or obstruction
  - Endoscopic incision vs. ureterocoele excision and re-implantation

**Ectopic ureters**
1:10,000
Usually females
Contralateral duplication (usually incomplete) in 80%
Bilateral infrasphincteric ectopia (leaking girls) in 10% of cases
Typically detected prenatally
Suprasphincteric usually present as UTI (often epididymo-orchitis in males)
Infra- or suprasphincteric in constant dribbling incontinence in girls (or persistent vaginal discharge in those with poorly functioning upper pole – minimal urine, pools in vagina)

**Clinical diagnosis**

- **USS**
  - Dilated distal ureter behind bladder
  - Dysplastic upper pole moiety may be missed (cryptic duplication)

- **DMSA**
  - Documents function

- **MCUG**
  - May identify suprasphincteric reflux

- **IVU**
  - ‘Absent’ upper pole calyx
  - ‘Drooping flower’ deviated lower pole moiety
  - ‘Scalloped’ lower pole ureter (deviated laterally by grossly dilated upper pole ureter)
  - May identify infravesical ectopia

Cystoscopy combined with EUA
- Suprasphincteric ectopia typically identified at bladder neck: often misses infravesical ectopia. Then instil methylene blue into bladder and insert vaginal pads [if blue and wet = bladder; clear and wet = infravesical ectopia]

**Management**
- Suprasphincteric
Non-functioning upper pole
  Heminephrectomy usually suffices
  Ureterectomy for reflux
Functioning upper pole
  Reimplantation
Infrasphincteric
  Non-functioning = heminephrectomy
  Functioning = re-implantation

**Single-system ectopia**
Single vaginal ectopic ureter
  Slightly less common than duplex
  Similar presentation – often referred as unilateral renal agenesis and incontinence
  DMSA or MRI may help locate renal tissue which may itself be ectopic
  Nephrectomy curative
Single proximal urethral ectopic ureter in girls – re-implant vs. nephrectomy
  Very rarely may be bilateral in girls. a/w small bladder, BN and sphincteric incompetence – Mx = bilateral re-implant, bladder augmentation +/- bladder closure and mitrofanoff

**Ectopic kidney**
  1:100 to 1:500
  90% unilateral
  left > right
  60% pelvic
  Often hypoplastic & irregular; associated with other GU abnormalities

**Horseshoe kidney**
  1:400
  95% lower polar fusion; 5% upper pole
  Level
    40% orthotopic
    40% below IMA
    20% pelvis
  Ascend limited by origin of IMA
  20% have associated PUJ abnormalities
  Associated with congenital abnormalities (10% of patients with Turner’s syndrome)

**Crossed renal ectopia**
  Uncommon
  Incidence ranges from 1:1000 to 1:2000 (from Campbell’s)
  Males > females
  Occasionally a/w VACTERL (typically solitary crossed renal ectopia)
  4 types:
    Crossed fused (85%)
    Left-to-right more common than right-to-left
    The superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney
Crossed non-fused (<10%)
Solitary
Bilateral
Clinical sequelae uncommon – occasionally low grade reflux

Megacalycosis
Non-obstructive enlargement of renal calyces due to abnormal development of papillae. Non-dilated renal pelvis and ureter.