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

<u>Selected duplication incidences</u>

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

<u>Duplex system ureterocoele</u>

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 ureterocoele 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
  - Ureterocoele prolapse
  - Ipsilateral lower pole reflux and UTI
- Surgical options (depends on upper pole function)
  - (i) Functioning upper pole
    a. Endoscopic ureterocoele incision
       - Simple, minimally invasive
       - Can induce reflux (do it close to bladder wall)
       - Reasonable medium term results (some view it as temporising measure)

    b. Ureterocoele 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
       - Ureterocoele and distal ureter aspirated and left in situ

  - (ii) Non-functioning upper pole
    a. Upper pole nephrectomy
       - aka 'simplified approach'
       - Upper pole moiety excised
       - Ureterocoele and distal ureter aspirated and left in situ

    b. Upper pole nephrectomy, ureterectomy and ureterocoele 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
   Refluxing ureters should be excised, but non-refluxing ureterocoele can be aspirated and left in situ

<u>Single-system ureterocoele</u>
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

<u>Ectopic ureters</u>
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)
Infrasphincteric as 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] NB. Bilateral infrasphincteric ectopia in 10%. Careful identification prior to planning surgery

Management
- Suprasphincteric

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

<u>Single-system ectopia</u>

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 sphichteric imcompetence – Mx = bilateral re-implant, bladder augmentation +/- bladder closure and mitrofanoff

<u>Ectopic kidney</u>

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

<u>Horseshoe kidney</u>

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)

<u>Crossed renal ectopia</u>

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 calcyces due to abnormal development of papillae. Non-dilated renal pelvis and ureter.