Ureteroceles and Duplex System

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• Definition
  – Ureteroceles: cystic dilatation of the terminal ureter within the bladder, urethra or both

• Demographics
  – 1 in 5000 children
  – Female : Male (4 : 1)
  – Very common in whites
  – Slight left-sided preponderance
  – 10% bilateral
  – 80% ectopic, from upper pole moiety in a duplex system

• Types:
  – **Single system ureteroceles**
    • Associated with a single kidney, a single collecting system, and a solitary ureter.

  – **Duplex system ureteroceles**
    • Associated with kidneys that have a completely duplicated collecting system and 2 ureters.

  – **Orthotopic ureterocele**
    • Orifice is located in a normal anatomic (orthotopic) position within the bladder
    • Usually arises from a single renal unit with one collecting system and is more common in adults

  – **Ectopic ureterocele**
    • Orifices are located in an ectopic position, such as the bladder neck or urethra.
    • Typically arises from the *upper pole moiety* of a duplicated collecting system
    • More common in the pediatric population
• **Aetiology**
  – Incomplete dissolution of the Chwalla membrane during fusion of the ureteric bud to the urogenital sinus.
  – Weigert-Meyer Law
    • Lower pole orifice migrates **cephalad & laterally** -> **shorter** detrusor tunnel -> **refluxing**
    • Upper pole orifice migrates **caudally & medially** -> **longer** detrusor tunnel
      – If Chwalla membrane complete dissolves -> **ectopic ureter**
      – If Chwalla membrane **incompletely** dissolves -> **ectopic ureterocoele**
  • **Lower pole** -> **Refluxing**
  • **Upper pole** -> **Ectopic & Ureterocoele**

• **Classification (American Academy of Paediatrics):**
  – **Simple**: Intravesical with single ureter
  – **Intravesical**: Entire ureterocoele, incl. stenotic orifice contained within the bladder; duplicated system
  – **Ectopic**: Part of ureterocoele extends into urethra

• [Stephen Classification – no therapeutic relevance]
• **Presentation:**
  – Antenatal ultrasound
  – Urosepsis / UTI
  – Incontinence
  – Voiding dysfunction
  – Hydronephrosis
  – Haematuria
  – Urinary retention
  – Prolapsing mass (female)

• **Examination:**
  – Abdominal pain or mass
  – Flank tendernessness
  – Prolapsing mass
## Imaging studies

<table>
<thead>
<tr>
<th>Ultrasound</th>
<th>VCUG</th>
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<tr>
<td>• Fluid-filled cystic intravesical mass</td>
<td>• Round filling defect along the base of the bladder = ureterocoele</td>
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<tr>
<td>• Hydroureteronephrosis</td>
<td>• VUR (Ipsilateral 50%, contralateral 25%)</td>
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<tr>
<td>• Duplex system</td>
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<tr>
<td>• Cortical thickness / dysplastic changes</td>
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<tr>
<td>• IVU</td>
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<tr>
<td>• Hydronephrosis</td>
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<tr>
<td>• Poorly functioning and hydronephrotic upper pole displacing lower pole - “Drooping lily sign”</td>
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<tr>
<td>• Lower ureteric displacement</td>
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<tr>
<td>• Cobra-head extension of distal ureter</td>
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<tr>
<td>• Stone</td>
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| Renograms (DTPA/DMSA/MAG3)                                                | CT scan                                                              |
| • Differential function                                                   | • If IVU or U/S equivocal                                           |
| • Obstruction                                                             |                                                                      |

| Other investigations:                                                     |                                                                      |
| • Urine C/S, renal function, blood culture                               |                                                                      |
| • Whitaker test                                                           |                                                                      |
| • If other tests to determine presence of obstruction equivocal           |                                                                      |
Treatment

- Medical - Initiate prophylactic antibiotics
- Surgical
  - Treatment goals:
    - Control of UTI
    - Preservation of renal function
    - Alleviate obstruction
    - Maintenance of urinary continence
  - Individualize surgical approach based on:
    - Age of the patient
    - Size and location of ureterocele
    - Degree of renal function
    - Presence and degree of vesicoureteral reflux
    - Comorbid conditions (anaesthetic risk)
Surgical Options

• **Endoscopic:**
  1) **Endoscopic incision**
     - Least invasive
     - Small puncture made at base
     - Indications:
       - Neonates
       - Sepsis
       - Obstructive uropathy
       - Indeterminate function
  2) **Transurethral unroofing**
     - Transverse smiley incision at base to minimize reflux risk
     - Indications:
       - Infection
       - Stone

• **Upper pole heminephrectomy, partial ureterectomy and ureterocoele decompression**
  - Indications:
    - Obstructed ectopic ureterocoele and a dysplastic upper pole, but without associated vesicoureteral reflux
    - Failed endoscopic Rx
    - If assoc. VUR -> add distal ureterocoele ligation
  - May need further surgical Rx if:
    - Severe VUR (Grade III, IV, V)
    - Residual stump Cx – UTI/Stone
    - Poor detrusor backing
  - Outcome: 60% spont. resolution of ipsilat. Grade I and II VUR
• **Ureteropyelostomy**
  - Joins upper pole ureter to lower pole pelvis
  - **Indication:**
    - if affected upper pole show significant function

• **Excision of the ureterocele and ureteral reimplantation**
  - **Indications:**
    - Significant lower moiety reflux
    - Well functioning upper pole
    - ± Significant contralateral reflux
    - Usually older child (>2 yr) but before toilet trained
    - Common sheath reimplantation or ureteroureterostomy
    - May need to taper dilated ureter

• **Nephroureterectomy**
  - **Indication:**
    - Single system ureterocele and non-functioning kidney
Churchill functional classification of ectopic ureteroceles

- Non-functioning upper pole
  - One renal unit in jeopardy (grade I) - Only the upper pole drained by the ureterocoele is affected (other renal units normal, may have grade I-II vesicoureteral reflux): Perform upper pole heminephrectomy.
  - Entire ipsilateral renal unit (grade II) or all renal units (grade III) in jeopardy - Ipsilateral and/or contralateral renal units affected by hydronephrosis or high-grade vesicoureteral reflux: Perform upper pole nephroureterectomy, ureterocoele excision with ureteral reimplantation.

- Indeterminate function: Perform endoscopic incision and reassessment of function.

- Functioning upper pole
  - One renal unit in jeopardy (grade I): Perform ureteropyelostomy and ureterocoele drainage.
  - Entire ipsilateral renal unit (grade II) or all renal units (grade III) in jeopardy: Perform ureteropyelostomy, ureterocoele excision, and ureteral reimplantation.
  - Note: the endoscopic incision is also considered first in infants who are medically unstable because of sepsis or coexistent medical conditions.
Complications

- **Endoscopic incision of ureterocele**
  - Iatrogenic VUR occurs in 40-50% of pediatric patients.
  - This is infrequently definitive therapy, except in cases of a single system intravesical ureterocele.

- **Upper pole heminephrectomy**
  - Highest risk of intraoperative blood loss
  - Vascular compromise of lower pole with potential loss of renal function
  - Need for further lower tract reconstruction due to persistent reflux, infection, or failure to decompress the ureterocele

- **Excision of ureterocele and ureteral reimplantation**
  - Problematic hematuria and/or bladder spasms
  - Damage to bladder neck or continence mechanism
  - Injury to the contralateral ureteral orifice
  - Compromise of blood supply to the lower pole ureter