DUPLEX SYSTEM URETEROCELE

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DUPLEx SYSTEM URETEROCELE

• Definition and Incidence
• Embryology
• Pathology
• Clinical Presentation
• Investigations
• Management
• Long Term Prognosis / Follow-up
Embryology

• Complete duplication
  - Two ureteric buds arise separately from the mesonephric duct
The caudal abnormal ureteric bud (red tip) arising from the mesonephric duct at around 6 weeks (a) subsequently incorporated into the superolateral trigone (b) with a short submucosal tunnel leading to VUR into the lower pole.
The abnormal ureteric bud (blue tip) arising cephalad on the mesonephric duct (a) subsequently draining to an abnormally distal ectopic location (Meyer-Weigart law).
Meyer-Weigart Law

• An accessory ureteric bud that arises more cranially on the mesonephric bud than normal and drains the upper renal pole will enter the urinary tract distally in an ectopic location.

• This may be in the bladder, urethra or urogenital sinus
Opening of the Ectopic UP ureter

• In females
  → suprasphincteric
  → at the level of the striated sphincter
    (usually below bladder neck)
  → infrasphincteric

• In males
  → always suprasphincteric
  → vas, SV or ejaculatory ducts (most common)
Duplex-system Ureteroceles-
Terminology

• Definition → cystic dilatation of the terminal ureter draining the upper pole

• May lie entirely within bladder (intravesical) OR encroach beyond bladder neck (ectopic/extravesical ureterocele)
Incidence of Duplex-system ureteroceles

• 0.02% of the general paediatric population

→ Of these, 80% are females

→ Bilateral = 10%
Duplex-system Ureteroceles

Associated with:

• Coexisting VUR in ipsilateral LP
  → present in 50%
  → moderate severity (Grd I-III reflux)

• Obstruction of ipsilateral LP ureter
  → rare, occurs d/t extrinsic compression by tense ureteroceles or dilated upper pole ureter
  → LP function preserved, not associated with dysplasia

• PUJO of ipsilateral LP
• Dilatation of ipsilateral UP ureter
  → usually (not always) associated with dysplasia of renal UP
  → dilatation may be massive & is d/t primary dysmorphism of ureteric wall

Upper pole dysmorphic and contributes only 1/3\(^{rd}\) of function
• Bladder outflow obstruction
  → caused by ectopic ureteroceles at bladder neck
  → prolapsed ureteroceles in girls

• Contralateral VUR
  → incidental finding, present in 25%
  → rarely exceeds Grade III severity
Embryology and Classification Of Ureterocele

(a) Delay in canalization of upper ureteric bud at time of contact with metanephric blastema results in cystic dilatation and subsequently resulting in (b) formation of duplex ureterocele. (c) Single-system orthotopic ureterocele. (d) Duplex-system ureterocele. (e) Caecoureterocele – Large ureterocele extending distally down from deficient trigone towards perineum in plane between urethra and vagina
Clinical Presentation

• 60% of children identified by antenatal USG

• Clinically present at infancy with UTI

• Rare → urinary retention because of sprolapsed ureteroceles in females.
Investigations – Ultrasound (USG)

• Look for
  → ureterocele—may be compressible with filling of bladder
  → dilatation of ureter(s)
  → kidneys—duplication, hydronephrosis, renal parenchymal thickness of UP
Investigations - MCU

• Essential in pts

• Look for
  – Ipsilateral LP VUR -50%
  – Contralateral VUR -25%
  – Reflux into ureteroceles-10%
  – Prolapse of ureteroceles into urethra in boys
Investigations - DMSA

- Dimercaptosuccinic acid (DMSA) scintigraphy
  → should be done routinely
  → to assess the distribution of f(x) in duplex kidney
Investigations - IVU

• Occasional role-historical
• Detection of cryptic duplication (absent upper calyx; drooping flower sign; scalloped appearance of LP ureter)
Investigations - MRI

• Anatomical and functional information without radiation
• BUT requires GA or sedation
• Justified for complex cases or elusive forms
Management Aims

- ↓ risk of UTI
- Preserve $f(x)$ of LP moiety & contralat. kidney
- Avoid bladder dysfunction
- Prevent incontinence
Management Options

• Influenced by
  - mode of presentation, age
  - presence of any associated or secondary effects of ureterocele on the renal UP, ipsilateral renal LP, bladder and contralateral kidneys

• In a relative proportion of ptts detected on antenatal USG, the ptts are asymptomatic and the tract is normal other than the ureterocele and small, severely dysplastic renal UP → any surgical intervention?? DEBATABLE
Management Options

• Conservative
• Endoscopic
• Surgery
  - Open
  - Laparoscopic
Management-Conservative

• In asx ptts without high grade HUN or VUR
• Absence of bladder dysfunction.

• Prior UTI is not a contraindication
• Need abs prophylaxis
• How long abs?

  Mean 1.5 years (Direnna & Leonard)
  Until resolution of VUR or child is toilet trained (Coplen & Austin)
  Until the age of 5 years if persistent VUR (Shankar et al.)
Management

Endoscopic ureterocele incision (TUI)

- Indication
  - antenatally detected cases where ureterocele is prolapsing or risks obstructing the bladder outlet
  - temporizing method in ptts with gross UP sepsis
- Least invasive
- Risk of UP new reflux 15-25% (to minimize risk → incision close to bladder wall)
- Majority will need some definitive form of surgery though some do respond well with this treatment alone
Endoscopic Incision - Controversies

• When to do? Earlier prevents UTI?

  *Infection rate of 50% in those awaiting surgery (Besson et al.)*
  *
  *No UTI in any pts awaiting surgery (Decter et al.)*
  *
  *Comparable rates of infection in early decompression vs. abs prophylaxis and delayed surgery [9% vs 8%] (Husmann et al.)*

  **Soooo... No Need for Decompression to Prevent UTI!!** (in asx pts without high grade HUN/VUR)

• Conservative
Management – Upper Tract Surgery

Upper pole heminephrectomy

- First choice in pts with severely compromised renal UP function
- Open approach – upper moiety excised with most of the attached ureter (same incision), ureterocele aspirated via stump (left in situ)
- Laparoscopic – advantage → ureter can be completely excised
- Simplified approach – Removal of UP and varying length of ureter (definitive tx in majority); subsequent excision of stump and ureterocele and reimplantation of LP ureter (in 10-20% only)
Management – Upper Tract Surgery

Pyelopyelostomy

• Only appropriate in small number of pts with useful f(x) of UP with dilatation of lower renal pole pelvis / proximal ureter
Problems – Upper Tract Surgery

• Long term f/up → Prevalence of *de novo* ipsilateral LP or contralateral reflux of 40-50% after upper tract surgery

• Damage to LP vessels during heminephrectomy
Controversies – Upper Tract Surgery

• Is surgery needed for non-f(x) / poorly functioning UP moiety if ptt asx?

No cases of HPT & only 2 nephrectomies for sx infections out of 48 patients over 9 yrs after decompression

(Chertin et al.)

Non f(x) upper moieties left in situ seldom cause sx or increase long term morbidity irrespective of chosen treatment approach

(Gran et al., Wang et al.)

• The use of scintigraphy to determine conservative mx?

Scintigraphy evidence of drainage (half-time < 30 min) suggest demonstrable moiety function

(Han et al.)
Management

**Ureteroureterostomy ± Reimplantation**

- Anastomosis between UP and LP ureters
- If only obstruction of UP found in DSU
- Approach → inguinal or upper
  → large ureterostomy on LP ureter for end-to-side anastomosis (better stent it at start of op)
- If reimplant → just the LP ureter
  → Persistent VUR may be seen (to tx endo or leave it alone)
Management

Ureterocele Excision and Reimplantation (Lower Tract Approach)

• Used when there is useful UP f(x) and NO dilatation of LP
• Transvesical approach
• Ureterocele excised up to level of LP ureter
• UP & LP ureters dissescted as a unit, UP ureter tapered and submucosal reimplantation
• Distal ureterocele excised up to BN, detrusor plicated, may need bladder mucosal flap
• Alternative to excision → fulguration or marsupialization
• Preferable to delay till after 1 yr of age d/t risk of interfering with bladder f(x)
Problems – Lower Tract Approach

• Caution
  -> do not separate the 2 ureters during intravesical dissection (dmg common bld. supply)
  -> care during surgery near the BN (dmg sphincter)

• Reflux (maybe present in 5-10%)
  -> some say no need for surgery
Management

Upper Pole Heminephrectomy, Ureterectomy and Ureterocele Excision + Reimplantation (Total Recon)

- Most extensive and lengthy, technically challenging, requiring 2 incisions
- Care during excision of caecoureterocele (sphincter damage, bladder neck injury, VVF)
- Ureterocele excised from above, repairing detrusor and mucosal defect
- Remaining distal lip resected endoscopically
- Reimplantation of LP ureter often required (careful to not injure blood supply during dissection of dilated UP ureter)
- Another option → lap heminephroureterectomy + open bladder procedure
# Options for Ureterocele Management

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Age</th>
<th>Appearance</th>
<th>Management</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ureteroureterostomy or ureteropyelostomy</td>
<td>Older patient</td>
<td>Functioning upper pole, No VUR</td>
<td>Drains obstructed segment with little risk of obstruction or UTI</td>
<td>May still require lower surgery in bladder, Leaves ureterocele in bladder, May develop reflux</td>
</tr>
<tr>
<td>Ureterocele excision and common sheath reimplantation</td>
<td>Reflux</td>
<td>Functioning upper pole without significant dilation</td>
<td>Eliminates obstruction and reflux, removes ureterocele, No renal risk</td>
<td>Complex surgery, Risk to vagina and bladder neck, May require ureteral tapering</td>
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UTI, urinary tract infection; VUR, vesicoureteral junction.
Combination Treatment

- Endoscopic ureteroceles puncture with subsequent endoscopic deflux injection for tx of reflux (70% 1 inj and 20% 2 inj)

Majority of VUR (asso. with ureteroceles) managed conservatively and 48% resolve or downgraded, 13% new VUR after TUI (5 of this 13% resolved spontaneously) – Chertin et al (J Urol 2007)
Asymptomatic neonate with DSU

Severe HUN in ureterocele or associated moieties +/- BOO

ED + prophylaxis

No severe HUN, no BOO

Follow-up full evaluation (3–6 months):
US, MCUG, renal scintigraphy (alternatively UroMRI)

Symptomatic or persistent severe dilatation

Upper or lower tract approach

Asymptomatic

Observation + prophylaxis and full evaluation if symptoms or progressive dilatation arise
DSU

Asymptomatic
No severe HUN or obstruction

No VUR

Low grade
Observation

High grade or multiple
Bladder surgery or endoscopic management

Symptomatic or severe HUN or obstruction

VUR

VUR

VUR

No VUR

Good function
Ectopic: upper to lower tract anastomosis
Intravesical ED

No/ poor function
Ectopic UPPN
Conclusion

• Tx should be generally individualized based on pre-op evaluation and clinical status up to 6 mths of age
• Asx pts with no BOO, massive HUN or no VUR > Grd III → conservative mx
• Sx pts or those with > Grd III VUR or massive HUN → surgery
• In ptts with massive HUN but no VUR, see if ureterocele intra or extravesical (determine moiety f(x))
• DSUs with high grade or multiple VUR (commonest) can be mx endoscopically (50% success) or open
• Endoscopic mx ± conservative mx of VUR or endo mx of VUR reduces need for open surgery
Thank You