Complications of Ectopic Ureteroceles & Ureters Seen In Transitional Urology

DA Husmann

Mayo Clinic, Rochester MN
Two Components in the Practice of Transitional Urologic Care

• Provide follow up care for complications that may arise from the treatment of congenital GU anomalies and childhood tumors
  • 97% of practice

• Transition our knowledge to adults to care for a newly diagnosed congenital anomaly
  • 3% of practice
Impact of Technology on the Dx & Rx Ectopic Ureteroceleles and Ureters

• Two Era’s
  • Pre and Post Routine Maternal-Fetal US
    ◦ 1980

• Historically huge impact on
  • Method of Dx
  • Coexisting pathology
  • Operation recommended

Mayo Clinic Patients: Ectopic Ureteroceles and Ureters
Age at Dx: Pre vs Post Maternal-Fetal US

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<tbody>
<tr>
<td>Neonate (&lt;28 days)</td>
<td>13% (14/101)</td>
<td>74% (70/94)</td>
<td>p &lt; 0.001</td>
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<tr>
<td>≥ 28 – 1 year Infant</td>
<td>32% (32/101)</td>
<td>7% (7/94)</td>
<td>p &lt; 0.001</td>
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<tr>
<td>&gt;1 -≤ 14 years (pre pubertal)</td>
<td>40% (40/101)</td>
<td>14% (12/94)</td>
<td>p &lt;0.001</td>
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<tr>
<td>&gt;14 years (post pubertal)</td>
<td>15% (15/101)</td>
<td>5% (5/94)</td>
<td>p = 0.02</td>
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Mayo Clinic Patients: Ectopic Ureteroceles and Ureters
Presentation: Pre vs Post Maternal-Fetal US

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<tr>
<td>Abnormal Maternal-Fetal US</td>
<td>4% (4/101)</td>
<td>71% (67/94)</td>
<td>p &lt;0.001</td>
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<tr>
<td>Febrile UTI</td>
<td>44% (44/101)</td>
<td>7% (7/94)</td>
<td>p &lt;0.001</td>
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<tr>
<td>Urinary Incontinence</td>
<td>19% (19/101)</td>
<td>8% (8/94)</td>
<td>p = 0.037</td>
</tr>
<tr>
<td>Voiding Dysfunction Ureteroceles</td>
<td>10% (10/101)</td>
<td>1% (1/94)</td>
<td>p = 0.007</td>
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Impact of Febrile UTI & Urosepsis
Ectopic Ureteroceles and Ureters

- Pre-US Era
  - ≈ 45% pts Febrile UTI
- Urosepsis
  - 15% -1/3 with urosepsis
    - 10% Mortality
    - 15% Develop CP
    - 25% Require Special Education

In essence = 10% mortality, 40% morbidity

Impact of Febrile UTI on Selection of Surgery

Partial Nx; all children with history of febrile UTI

- Function on DMSA upper pole
  - Poor <15%
    - 35% (30/85 pt) chronic pyelonephritis
  - Good ≥15%
    - 14% (2/14 pt) chronic pyelonephritis

- Significant concern
  - Upper to lower pole anastomosis

Levy et al, J of Urol 158:1997
Hypertension
Multicystic & Segmental Dysplastic Kidney

Incidence of hypertension
- 0.5-0.7%
- DMSA
  - Only reported in functional kidneys
- Nx or partial Nx resolves HPT in 1/3rd
  - Majority of HPT contralateral or lower pole scars

Ectopic Ureterocele
Hypothesis of Intrinsic Deficiency of Bladder Neck

Hypothesis Regarding Long-Term Development of Urinary Incontinence Ectopic Ureterocele

• Congenital malformation of bladder neck
  • Paucity of internal sphincter musculature
• Women > men 4:1
• Age and Parturition status
  o High incidence SUI in adults

Hendren & Mitchell  J of Urol 121: 1979
Three Strikes Its Out
Ectopic Ureterocele - Ureter

• Infection
• Function < 10%
• Bladder neck involvement
Pre-Sono Era

- Surgery of choice – Complete Reconstruction
- Nonfunctional (<10%) upper moiety
  - Partial Nx, ipsilateral lower pole reimplant, excision of ureterocele, BNR +/- contralateral ureteral reimplant
- Function (>10%) upper moiety
  - U-U (upper or lower level), ipsilateral lower pole reimplantation, excision of ureterocele, BNR, +/- contralateral ureteral reimplant

Hendren & Mitchell  J Urol 121: 1979,
Pre vs Post Sonographic Era

• Pre- Sono Era - N=101 pts (1962-1977)
  • Median age - 18 months (range 1 day - 48 yrs)

  • Median age – 3 months (range 1 day - 62 yrs)
  • $p < 0.05$

Significant Concerns Raised

Infant surgery

- Increased technical difficulty
- Risk of renal loss
- Risk of impairment of bladder neck
  - Life long incontinence
  - Scared bladder neck – voiding dysfunction
- Risk of damage to contralateral vas or ejaculatory duct

Post Sono Era

- No longer dealing with Dx following febrile UTI
  - Risk of chronic pyelonephritis - less
- Risk of Dysplasia-Hypertension
  - Low
- Does anyone really know the risk for SUI?
- Need to change to less risky operation

Pathologic Key to Bladder Neck Dysfunction (Outlet) Causing Obstruction

Pathologic Alterations

• Over distension bladder wall = hypoxia
  • Apoptosis – neural & smooth muscle cells

• Reperfusion injury = Toxic oxygen radicals
  • Endothelial leak of fluid into interstitium
  • Damage DNA
  • Alters bladder collagen
  • Apoptosis - smooth muscle & interstitial cells

• What does this mean to the urologist?

Bladder Outlet Obstruction
What does the Urologist Need to Know?

• Hypoxia
  • Kills sensory nerves first
  • Lumbar-sacral ganglia second (contraction)

• Collagen alterations
  • Impairs smooth muscle syncytium (contraction)

• UDS alterations in order
  • Impaired sensation, overactive, overactive poorly contractile, underactive

Abrahamsson, et al, J of Urol 160:1998,
In patients with an ectopic ureterocele:

Is bladder neck dysfunction & primary bladder dysfunction a consequence of surgery or due to the underlying congenital anomaly?
Ureterocele Puncture, Partial Nx Alone, Complete Reconstruction

- Ureterocele Puncture  N= 41 pts
  - Median surgery age - 5 days (1-2 weeks)
  - Median follow up 10 yrs (3-34 yrs)

- Partial Nx  N=87 pts
  - Median surgery age - 3 months (1 month - 16 yrs)
  - Median follow up 10 yrs (3-28 yrs)

- Complete- Total Reconstruction N=60 pts
  - Median surgery age - 4.5 yrs (3 months – 16 yrs)
  - Median follow up 14 yrs (3-28 yrs)
## Ureterocele Puncture, Partial Nx Alone, Complete Reconstruction

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<thead>
<tr>
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<th>Ureterocele Puncture/incision</th>
<th>Upper pole Nx</th>
<th>Total Reconstruction</th>
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<tbody>
<tr>
<td><strong>Delayed Void</strong> ≤ 3 times per day</td>
<td>7% (3/41)</td>
<td>11% (10/87)</td>
<td>12% (7/60)</td>
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<tr>
<td><strong>Urgency Incontinence</strong></td>
<td>20% (8/41)</td>
<td>10% (9/87)</td>
<td>8% (5/60)</td>
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<tr>
<td><strong>Need for CIC</strong></td>
<td>2.5% (1/41)</td>
<td>2% (2/87)</td>
<td>3% (2/60)</td>
</tr>
<tr>
<td><strong>Stress or total Incontinence</strong></td>
<td>2.5%(1/41)</td>
<td>6% (5/87)</td>
<td>3% (2/60)</td>
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<td>2 pts bilateral ureteroceles</td>
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Is there a voiding abnormality present based on treatment used? 
Median F/U 10 – 14 yrs

• Urinary incontinence
  • 25-30% of pts irrespective of Rx
• Timed voiding +/- anticholinergics
  • 75-85% resolution
• CIC required
  • 7-12%
• Could we have prevented by better F/U?
Persistent Stress or Total Incontinence
2-3% of Ectopic Ureterocele Patients
(Median F/U 15 yrs, Range 8-28)

8 pt - BNR with sling

- 4 pt - success
  - 2 pt - AUS
  - Failed 15, 22 yrs
  - 2 pt - BNL & Stoma
  - 2 pt - BNL & Stoma
Ureterocele; What to do with persistent VUR? Females Only

• 29% (54/188 pts) No Reflux

• 49% (93/188 pts) high grade 4-5 reflux
  • We repaired -163 ureters
  • 75% success (122/163 ureters)
    o 41 ureters treated with Deflux
      – 63% (26/41) success
    o 15 ureters
      o 5% (8/188) with U-U
      o 4% (7/188) persistent VUR
Persistent Low Grade VUR (2-3/5)

- 22% (41/188) pts
  - Median F/U 13 yrs (2-28 yrs)
- 76% (31/41 pts) asymptomatic
- 10 pts (18 ureters) recurrent > 2 UTI – catheterized
  - 80% (8/10 pts) successfully Rx with Deflux
  - 20% (2/10 pt) Failed Deflux UNO with success

- Persistent observation - low grade VUR reasonable
Ureteral Stump Syndrome
Ectopic Ureters

• All ectopic ureters into the urethra managed by open partial nephrectomy

• Repeat VCUG performed 3 months post op

• Refluxing stumps ligated distal to iliac vessels
  • 20 pts
    • 50% (10/20) with resolution of reflux

• Non-refluxing stumps left to drain
  • 21 pts
    • 29% (6/21) new onset of VUR

Ureteral Stump Syndrome
41 pts - Median F/U 15 yrs (3-28 yrs)

Non-refluxing stumps 21pts
- Never Refluxed – 15 pts
  - 13% (2/15) Recurrent UTI
  - 87% (13/15) Asymptomatic
- Secondary Reflux – 6 pts
  - 50% (3/6) Recurrent UTI
  - 50% (3/6) Asymptomatic

Refluxing 20 pts
- Always Refluxed – 10 pts
  - 30% (3/10) Recurrent UTI
  - 70% (7/10) Asymptomatic
- Initially Refluxed – 10 pts
  - 30% (3/10) Recurrent UTI
  - 70% (7/10) Asymptomatic

Never Refluxed 13% (2/15) Recurrent UTI
Refluxed 35% (9/26) Recurrent UTI
Ectopic Ureters Presenting As Adults (N=14 women)

- Presented with classic voiding history of voiding normally yet wet all of the time since childhood (28%; 4/14)

- Presented with recurrent UTI’s, incontinent only during pregnancy (28%; 4/14)
  - Ectopic just distal to BN
  - Gravid uterus extra pressure
  - Laxity of pelvic floor induced by hormones

Dysplastic Kidney with Ectopic Ureter DX as a Vaginal Cyst
Ectopic Ureters Presenting As Adults (N=14 women)

- Incontinent following excision of “vaginal cyst” by GYN 28% 4/14 pts
  - Terminal segment of ectopic ureter occluded forming vaginal cyst

Eubanks & Gonzalez, Ob & GYN 127:2016
21 y/o woman with severe dysmenorrhea
Vaginal Duplication and Ectopic Ureters
Obstructed Hemi-vagina and Unilateral Renal Agenesis

• Presentation classically with severe dysmenorrhea secondary to obstructed hemivagina

• MRI/CT with vaginal duplication and obstruction with uterine didelphys – agenesis/dysplastic kidney

• GYN – Resected vertical vaginal septum

• Onset of classic hx urinary incontinence yet voiding normally - post resection (16%; 2/14)

50 y/o man with intermittent left flank pain, CT stone protocol
28 y/old male with pain with ejaculation, cystic right pelvic mass on rectal US
Zinner’s Syndrome

- Ectopic Ureters in men 55% empty into prostatic urethra with paucity of symptoms

- Zinner’s Syndrome
  - 45% - empty into seminal vesicle, ejaculatory duct or proximal vas
  - Unilateral renal agenesis on Fetal US
  - Unilateral laterally displaced mesonephric duct cyst develops post puberty
    - Secretions from seminal vesical ducts

Allaparthi & Blute Cn J of Urol 17: 2010
Zinner’s Syndrome

- At puberty or in adulthood - Recurrent prostatitis, epididymitis, pelvic pain, infertility, palpable prostate anomaly
  - 2 pts > 60 yrs of age - underwent transrectal prostate biopsy “abnormal prostate on DRE” pelvic abscess requiring PCN drainage and delayed surgery
Zinner’s Syndrome  
(Beware of Voiding Dysfunction)

• 5 pts referred with obstructive voiding Sx  
  • First pt - excised mesonephric cyst  
  • Continued voiding dysfunction  
  • UDS- underactive bladder on CIC

• The remaining - 4 pts UDS first  
  • All with UDS with underactive bladder  
  • 2 excised on CIC  
  • 2 excised voiding with Valsalva with high PVR observing  

• Should we have operated?
Chronic Pyelonephritis following U-U

- 3 pts with history of recurrent febrile UTI (2-4)
  - Ureteral ectopy into urethra – no reflux
  - Upper to lower UU, transection of stump
- Multiple recurrent UTI in adolescence & adults
  - No voiding dysfunction, refluxing stump, VUR
  - Bacterial Isolation studies - ipsilateral bacteria
  - DMSA scan 20-25% function as adult
- Nephrectomy - chronic pyelonephritis both segments
Points to Remember

• Voiding dysfunction post Ureterocele Rx appears to be primary in origin not surgically induced
  • Timed Voiding – Key

• Persistent incontinence with ureterocele primary defect, rare but does exist
  • ≠ AUS

• Reimplantation high grade VUR post ureterocele
  • 75% success
  • Salvage 50% failures with Deflux
Points to Remember

• Grade 1-3 VUR - post ureterocele Rx
  • Successfully managed observation in 75%

• Ureteral stump syndrome
  • Refluxing stump 1/3\textsuperscript{rd} require surgery
  • Non-refluxing stump 15%

• Ectopic Ureters in adult women
  • Classic voiding history may be delayed
  • Beware of incontinence - I&D Vaginal cyst
Points to Remember

• Zinners Syndrome
  • Voiding dysfunction maybe due to underactive bladder – not cyst

• Beware of risk of chronic pyelonephritis with UU in patients with history of recurrent UTI
Thank- You