Challenges in Stone Management of “Complex” Patients

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Financial and Other Disclosures

- Off-label use of drugs, devices, or other agents: None or FILL IN HERE; including your local regulatory agency, such as FDA, EMA, etc.
- Data from IRB-approved human research is presented [or state: “is not”]

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<th>I have the following financial interests or relationships to disclose:</th>
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Why do we call them complex?

- Complex GU anatomy
  - Congenital/Reconstructed
- Complex anatomy (pelvic, GI, GYN)
- Body habitus
- Comorbidities
- Unusual presentation/asymptomatic
- Compliance
- Patient/Family/Other provider’s priorities
- Logistics/Adults/Out-of-towners

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Risk Factors for Stones in Patients with Neurogenic Bladder

- Urinary stasis and treatment of stasis
- Immobility
- Infection
- Diet
- Medications

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Urinary Stasis

- Accumulation of debris
- Bladder drainage
  - Patients with indwelling catheter 9x more likely to form stones than those who void
  - Patients on CIC 4x more likely to form stones

Urol Clin N Am, 2010

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Immobility

- Immobility releases calcium from the bone
  - Sudden as in spinal cord injury or spinal fusion
  - When young men with Duchenne’s stop walking and start using wheelchair
  - Stone risk does not suddenly start in children who have never walked as in non-ambulatory children with spina bifida

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Infection

- Bacterial infection that hydrolyzes urea to ammonium
  - leads to higher urine pH and stones composed of **struvite** (magnesium ammonium phosphate) or **dahllite** (carbonate apatite)

- Urease producing organisms (Not E coli)
  - Proteus, Klebsiella
  - Others – Morganella, Providentia, Serratia, Staphylococcus, some Pseudomonas

- Acetohydroxamic acid inhibits urease
Infection and Stones

125 patients (PCNL, Mayo Clinic)

<table>
<thead>
<tr>
<th></th>
<th>19 Struvite + stone culture</th>
<th>24 Non-struvite + stone culture</th>
<th>82 Non-struvite - stone culture</th>
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<tbody>
<tr>
<td>NGB</td>
<td>26%</td>
<td>8%</td>
<td>0%</td>
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<tr>
<td>Hypoicitr</td>
<td>47%</td>
<td>32%</td>
<td>26%</td>
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Cogain et al, Urology, 2014  
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Diet

- Poor fluid intake
- Ketogenic diet for seizures
- High protein diet
- Enteric formulas
- High salt intake
Medications

- **Migraine/Seizures treatment**
  - migraine/seizures treatment (Topiramate, Zonisamide, acetazolamide)
  - Carbonic anhydrase inhibitors induce a **mild distal renal tubular acidosis** with a relatively elevated urine pH
  - acidosis, hypokalaemia, hyperuricaemia and hypocitraturia (Dell’Orto VG, Br J Clin Pharmacol, 2014)

- **Steroids**
  - Long-term corticosteroid use can increase enteric absorption of calcium, leading to hypercalciuria and an increased risk for calcium-containing stones

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Children with Spinal Abnormalities have an increased health burden from upper tract urolithiasis

- Ramachandra et al, Urology, 2014
  - data extracted from the Pediatric Health Information Systems database over an 8-year period - 11987 pts with stones
  - prevalence of stones in patients with normal spines was 0.24% compared with 1.40% and 4.03% among children with spinal curvature and spinal dysraphism, respectively (P<.001)
  - children with spinal curvature and spinal dysraphism were more likely to have multiple procedures for stones than those without spinal abnormalities (25% vs 25.7% vs 13.1%, P<.001)

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Risk Factors for Upper Tract Stones in Children with NGB


- Augmented bladders
- History of bladder stones
- Male
- Non-ambulatory
- Thoracic level dysraphism
Case # 1 – “Simple” complex patient

- LY DOB, 07/29/1997
  - MM, NGB
  - 2003 - Continent urinary reconstruction: YDL bladder neck reconstruction, bladder neck fascial sling, Monti Mitrofanoff, appendiceal MACE, bilateral ureteral reimplantation (Cohen)
  - 2004 - Revision of YDL bladder neck reconstruction, take-down of Mitrofanoff, Monti Mitrofanoff, Colocystoplasty
  - Stable upper urinary tract, continent of urine and stool
Case #1

- 2013 - small non-obstructing R renal stone (for several years) by RUS. No pelvocalyectasis or ureteral dilatation
  - KUB – small calcification in R lower pelvis (fecalith, flebolith, ureteral stone?)
Case #1

- CT scan

- A 6.4 mm calculus is seen in the right lower pelvis, which is favored to be vascular in nature rather than associated with the genitourinary system.

1. Further imaging/Conservative treatment
2. Endoscopic approach via Mitrofanoff (stenting/lithotripsy)
3. Percutaneous approach (drainage/stenting/lithotripsy)
4. Open ureterolithotomy
Case #1

- 2 months later - admission for R flank pain and sepsis
  - RUS - “new” R distal ureteral stone with moderate R hydroureteronephrosis

1. Further imaging/Conservative treatment
2. Endoscopic approach via Mitrofanoff (stenting/lithotripsy)
3. Percutaneous approach (drainage/stenting/lithotripsy)
4. Open ureterolithotomy
Case #1

- Percutaneous nephrostomy tube placement by Interventional Radiology (IR) with immediate improvement in symptoms
- Nephrostomy tract dilatation (by IR), percutaneous antegrade flexible ureteroscopy with basket extraction of ureteral stone
- Last follow up (2015) with RUS – “stable R renal stone without pelvocalycectasis, no ureterectasis or ureteral calculi seen”
Case #1

- **Challenges**
  - Confusing initial presentation
    - Asymptomatic patient with no evidence of obstruction and equivocal radiological evaluation – did it change ultimate approach?
  - Complex anatomy - limited retrograde access to ureter: Cohen ureteral reimplantation + augmentation + BN reconstruction
    - Any “tricks” how to approach ureteral stones in this situation?
Case #2

- **CF, DOB 1993**
  - Cloacal exstrophy, XY karyotype, female gender assignment (outside institution)
    - 2 initial attempts of bladder closure
  - Long-term management by CCHMC colorectal team
    - 1995: colon pull-through, vaginoplasty with rectum, vesicostomy, genitoplasty
    - 1996: R ureteral reimplantation, TUU (L to R), L ureteral Mitrofanoff, ileal neobladder
    - History of 2 episodes of urinary obstruction resulting in ARF with creatinine to 3.3
    - History of bladder stones
Case #2

- 2011: Presented to GU team for management of urolithiasis - delightful young lady, successful college student (in Atlanta)

1. Observation
2. Endoscopic lithotripsy via Mitrofanoff
3. Percutaneous Cystolithotomy
4. Open Cystolithotomy
Case #2

1. Observation
2. Endoscopic lithotripsy via Mitrofanoff
3. Percutaneous Cystolithotomy
4. Open Cystolithotomy
Case #2

- SP tube placement by IR: is that necessary?
- Percutaneous cystolithotomy (EHL, CyberWand) with extraction of multiple fragments
- 2 weeks later - second look Cysto, extraction of small fragment

Case #2 FOIU, 2018
Lesson learned - how could I prevent missed stone at first procedure
- Careful bladder examination
  - Small skin incision
  - Ileal neobladder
- Count stones (how?)
- KUB on the table
- All of the above!?
Random urine sampling in children with low muscle mass. Use calcium/osmolality ratio or 24 hour urine.

Persistent hypercalciuria – check bone density

Metabolic stone profile may be helpful even if you have a reason for the stone – like infection