Pediatric Urinary Incontinence, Bladder Disorders and Metabolic Stones

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1

Disclosures

- None

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2

Normal Bladder Function

- Store
  - Normal capacity (cc) for age
    - Child < 1 year old: **weight (kg) X 7**
    - Child ≥ 1 year old: **[age (years) + 2] x 30**
  - Sense distension
  - Accommodate increasing volume without change in pressure = remain *relaxed* when not voiding
- Empty
  - Initiate and sustain contraction
  - Empty to completion

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Normal bladder development

- Infants
  - *Multiple small voids with high voiding pressures (hourly)*
  - *Discoordinated voiding is normal (sphincter contraction with detrusor)*
  - Complete emptying
- Toddlers
  - Voiding frequency decreases as capacity increases
  - Voiding becomes more coordinated
  - Toilet training: 2-4 years old
- Teens: 4-6 x daily
  - Basis for CIC Q 4hrs while awake

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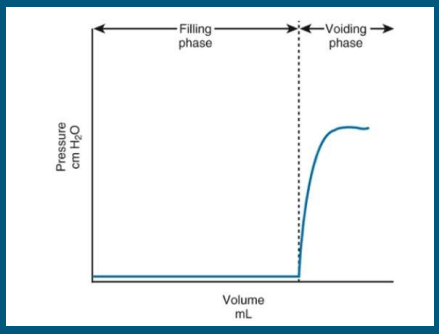
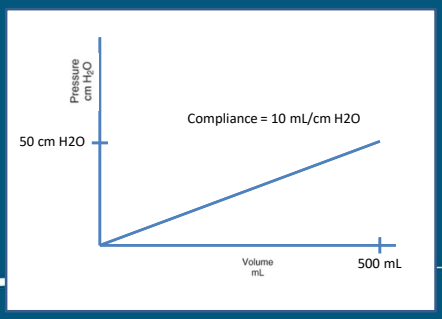
Capacity and compliance

- Estimated capacity
  - < 1 year: weight in Kg x 7 mL
  - > 1 year: (age+2)x30 mL
- Compliance (volume / pressure)
  - *Normal compliance is HIGH*
  - 46-124 mL/cm H2O in Campbell's
  - Estimated bladder size affects normal compliance
    - 10 could be normal for an infant





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Urodynamics in Children

- High Pdet common in < 1 yo
  - As high as 120 cm H_2O in boys and 75 cm H_2O in girls
- Staccato voiding associated with transient DSD is normal
- Remember these UDS tips:
  - Slow fill < 10% predicted capacity/min
  - Fluid should be body temp
  - Catheter size can impact LPP

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Pediatric LUTS conditions

Common

- Voiding dysfunction/bowel bladder dysfunction
- Constipation
- Overactive bladder
- Vaginal voiding
- Voiding postponement
- Underactive bladder
- Nocturnal enuresis

Uncommon, but need to know

- Giggle incontinence
- Ectopic ureter
- Neurogenic bladder
- Non-neurogenic neurogenic bladder
- Posterior urethral valves late presentation
- Pollakiuria, AKA Benign urinary frequency of childhood
- Benign childhood urethrorhaggia
- *Diabetes presentation – don't forget*

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History in clinic

- Daytime incontinence? Nighttime incontinence?
- How frequent? Daily?
- Voiding frequency? *If does not void in AM -> voiding postponement*
- Any urge prior to leak? *Leak soon after voids -> vaginal voiding*
  Continuous leakage -> ectopic ureter
- Bowel habits? *Daily soft BM is goal. Often unknown.*
- Birth history? *DM during pregnancy -> Sacral agenesis*
- Any other developmental concerns? ADHD? Abuse/neglect? Stress?
- Dysfunctional voiding symptom score (DVVS), etc

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Physical exam in clinic

- GU exam
  - Labial adhesions, Skin irritation from incontinence
  - Ectopic ureter – hard to see in clinic
  - *Urine pooling in vagina -> vaginal voiding*
- Spine
  - *Sacral dimples or other abnormal findings -> spinal dysraphism/etc*
- Abdominal
  - Masses, palpable stool burden
- Neurologic
  - Gait, LE strength (walk on toes, walk on heals), perineal sensation/reflexes

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Evaluation in clinic

- Voiding diary; consider UA and uroflow
- *If febrile UTIs or recurrent non-febrile UTIs:*
  - *Consider renal US – assess bladder emptying, upper tract health*
  - *Possible VCUG*
- If continuous incontinence – renal US, MR urogram for ectopic ureter
- If abnormal spine, neuro exam, or refractory case – MRI of spine to evaluate for tethered cord, spinal dysraphism

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Bladder Function Disorders

1. Mono-symptomatic nocturnal enuresis: 80%
2. Daytime urinary incontinence disorders: 20%
   - Prevalence decreases with age:
     - 4-6 year-old children: 10 – 20 %
     - 6-18 year-old children: 4- 5 %
   - Risk factors for urinary incontinence:
     - Female gender
     - History of UTIs
     - Fecal incontinence

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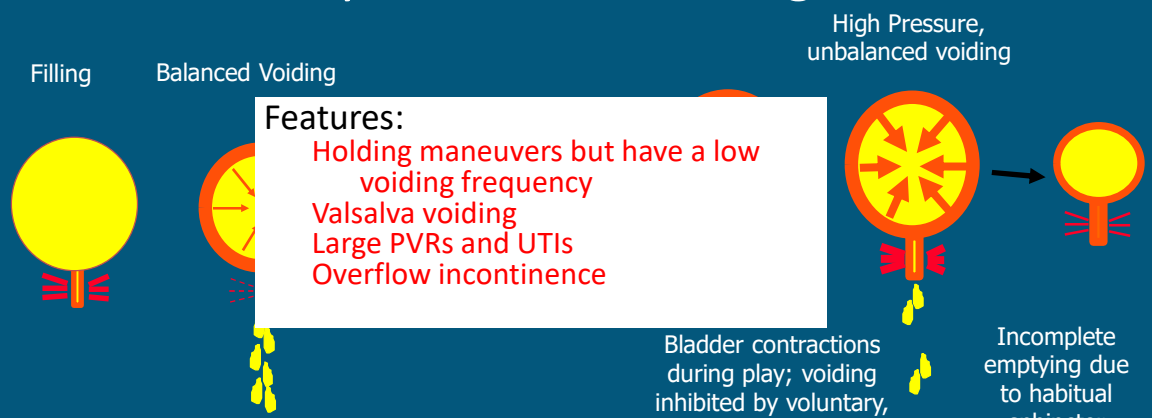
Dysfunctional voiding



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Bowel bladder dysfunction (BBD)

- Frequency, urgency, daytime incontinence, holding behaviors constipation UTIs, pain
- No structural or neurologic cause
- Discoordination of sphincter likely acquired during toilet training
- Associated with VUR and UTIs
  - TREAT BBD PRIOR TO OR ALONG WITH VUR
- 20-40% have neuropsychiatric disorder- ADHD, ASD

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Image from bedwettingandaccidents.com, Steve Hodges, MD

13

Treatment for dysfunctional voiding/BBD

- Urotherapy
  - Inform, instruct, and demystify condition
  - Behavioral modification
    - Q2 hr timed voiding, double voiding, relaxation techniques, good posture, good hygiene, timed voiding watch
  - Treat constipation if present
    - Timed toileting (AM/PM), fiber supplements, miralax, enemas, etc
- Neuropsychiatric condition should be treated (ADHD, etc)

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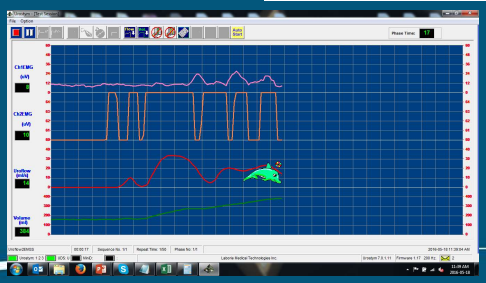
Biofeedback for BBD

- Adhesive pads measure pelvic floor activity
- Computer game controlled by contracting and relaxing pelvic floor activity
- Uroflow/EMG/PVR also used
- Each session 45-60 min
- At least 5 years old, better if over 7



Images from laborie.com

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Overactive bladder

- Urinary urgency, frequency, +/- incontinence, +/- nocturia
- UA – evaluate for infection
- PVR – evaluate for poor emptying
- PE – concern of neurologic cause?
- Treatment
  - *Treat constipation -> common cause of frequency/urgency*
  - Urotherapy
  - Anticholinergics (oxybutynin[1], tolteradine[1]) or mirabegron[2] (liquid form available)

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[1]FDA approved for >5 yrs
[2]Not FDA approved indication, only neurogenic

16

Voiding postponement & underactive bladder

- Holding behaviors and postures
- UTIs, overflow incontinence, sudden urge but infrequent voids
- May lead to overstretching detrusor -> weak contractions -> underactive bladder/AKA “lazy bladder”
- Can have large PVRs, Valsalva voiding
- Treatment
  - *Primary = Urotherapy (timed voiding/double voiding)*, ? Alpha blockers (off label usage), CIC

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Vaginal Voiding

- Classic history
  - overweight prepubertal female, incontinence within 10-15 minutes of voiding
- Exam
  - Urine pooling in vagina, also look for labial adhesions
- Treatment
  - Pull down underwear all the way, sit with legs spread wide, lean forward, dry well

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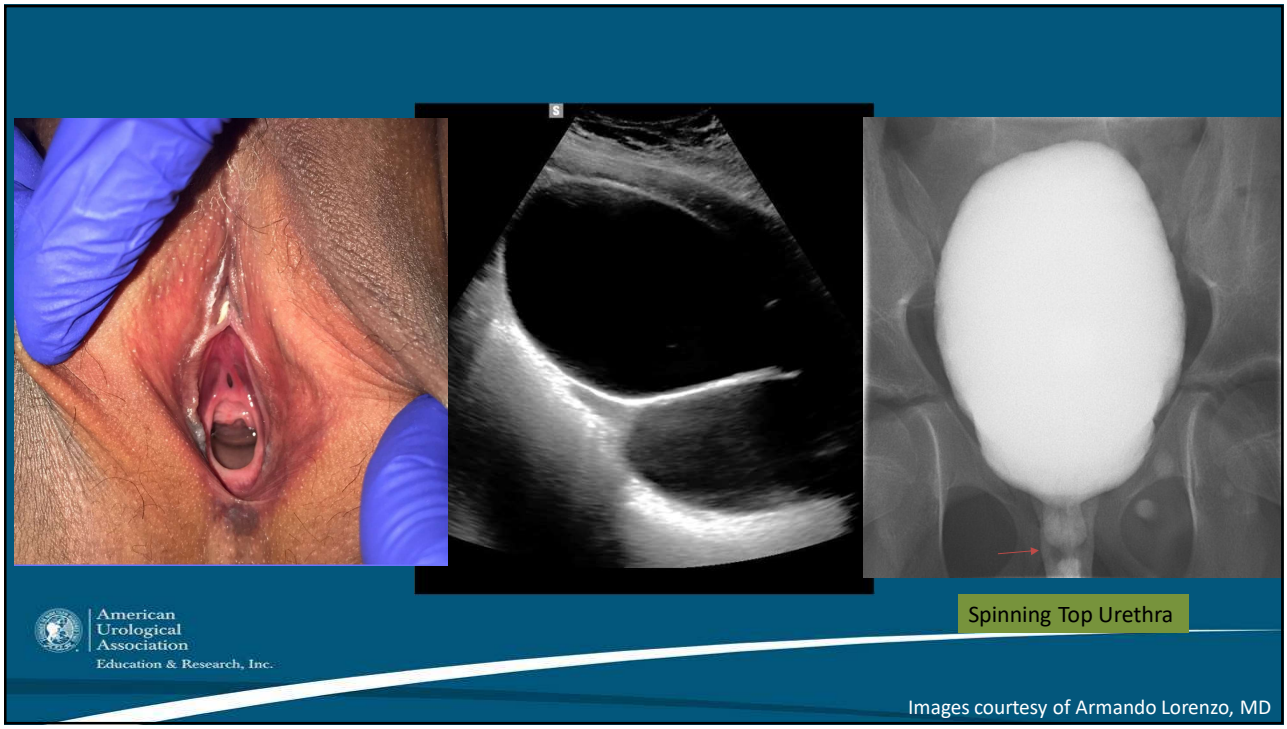
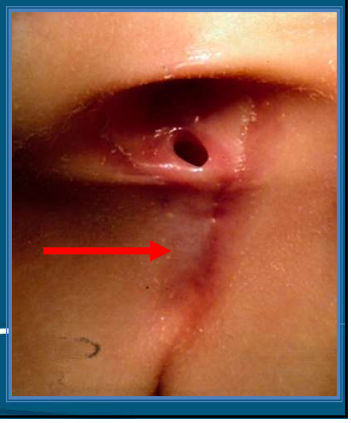


19

Labial adhesions

- If not causing problems (UTIs, vaginal voiding), leave alone
  - *Natural history is resolution over time, even if 95% closed*
  - *Once you start treating, may require re-treatment until puberty*
- Estrogen creams and steroid creams have been used successfully if sxs = pruritis, UTIs, post void dribbling
  - Premarin, betamethasone, estrace
- Lysis of adhesion with local anesthesia or in OR for refractory cases with symptoms



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Nocturnal enuresis

- Primary nocturnal enuresis = present since toilet training
  - Deep sleepers, runs in families, ~15% of 5 year olds
- Natural history is resolution over time
- Secondary nocturnal enuresis = wetting after >6 mo dry
- *Treat daytime incontinence/constipation/LUTS first*
- *If secondary think constipation and sleep apnea*
- *Treatments:*
  - *Observation, limiting fluids in evening, wake up on schedule, bedwetting alarms, DDAVP, anticholinergics, imipramine (2nd line)*

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Giggle incontinence

- Prepubertal females
- Significant incontinence episodes triggered by laughing, not just mild stress incontinence
- Otherwise normal urinary/bowel habits
- Significant adverse effect on life
- Treatment
  - Observation: improves with puberty typically
  - Biofeedback
  - Methylphenidate (ADHD medication, off label use)

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Pollakiuria, AKA benign urinary frequency of childhood

- Classic history
  - Toilet trained male 4-8 years old
  - Rapid onset, VERY frequent voiding, no incontinence
  - Sleeps through the night
  - Can have recent traumatic event
- Evaluation: Clinic evaluation with exam, UA, PVR
- Natural history: Resolution over several months
- *Treatment: reassurance*

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Benign childhood urethrorrhagia

- Classic history
  - Boy at beginning of puberty (8-10), clear urine but few drops of blood at end of void, can have terminal dysuria
- Evaluation: H&P, UA, PVR, possible renal US for reassurance
- Natural history: resolution 6-12 months, +/- association with strictures
- Treatment: Reassurance, hydration, void regularly, treat constipation
  - Possible cystoscopy if persists for many months

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Ectopic ureter

- *Classic history*
  - *Female child who toilet trained and can void but has "constant" wetness*
  - *Males will not have incontinence with ectopic ureter*
    - *Recurrent epididymo-orchitis or pelvic symptoms*
- Evaluation
  - May see ectopic orifice on physical exam – hard in clinic
- Imaging
  - Renal US may miss small atrophic upper pole
  - MR urogram

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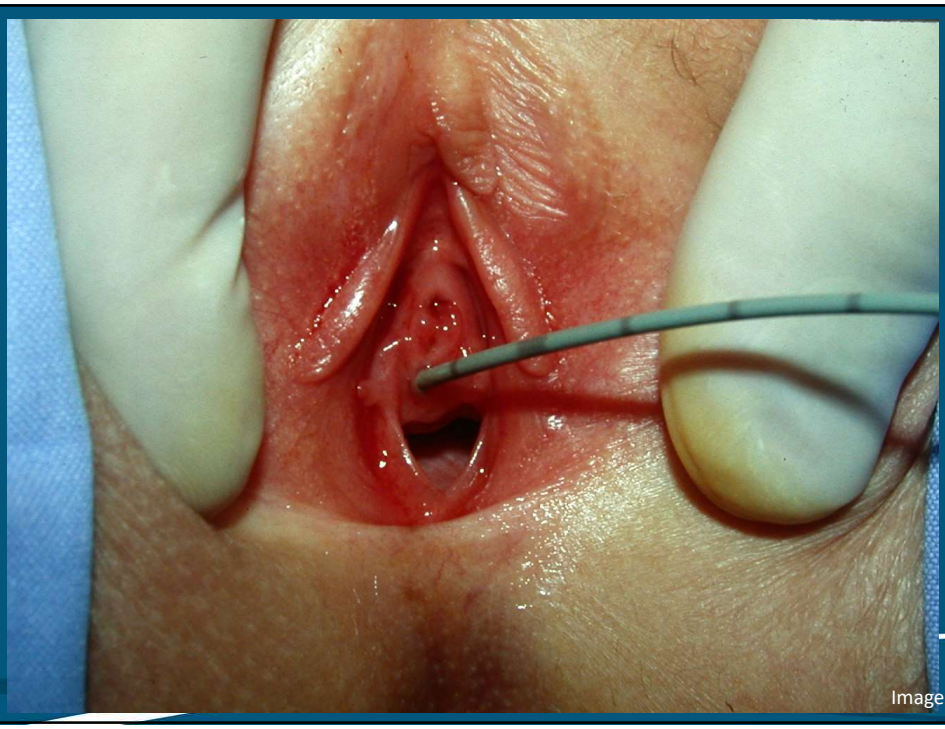
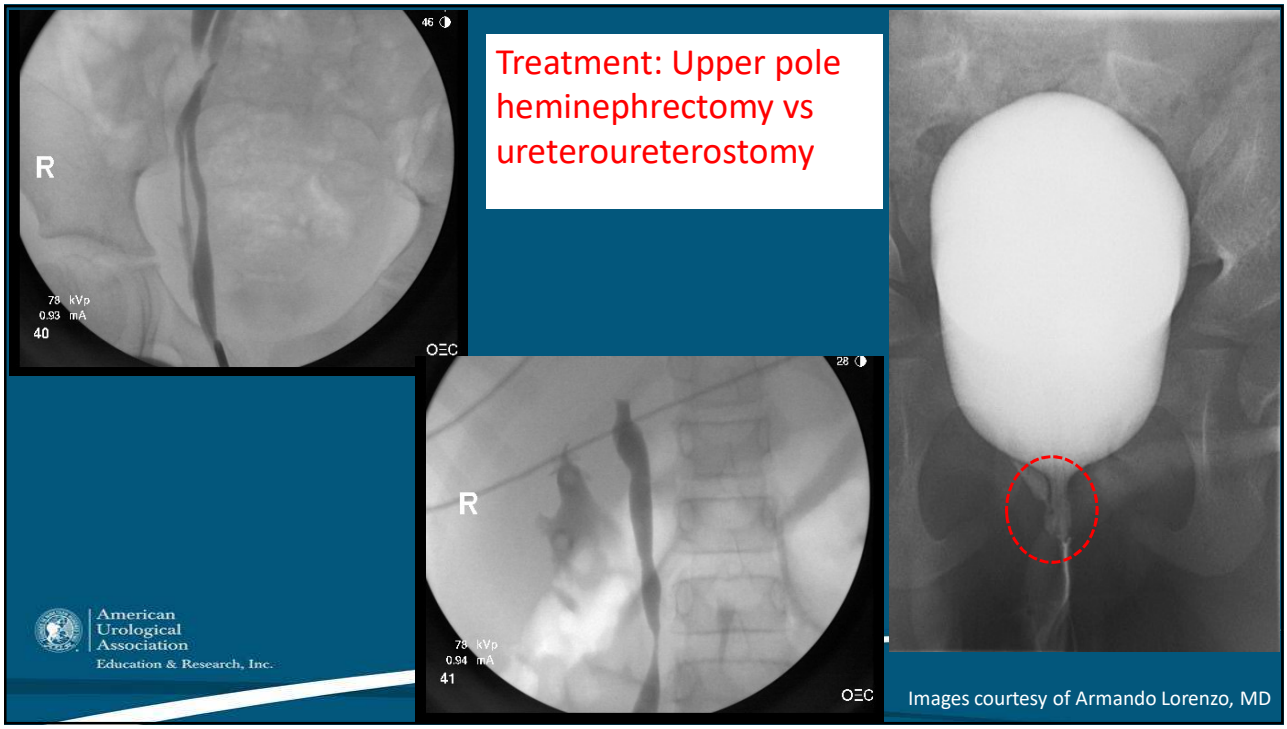


Image courtesy of Armando Lorenzo, MD

26

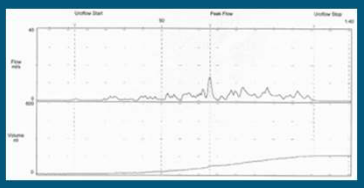


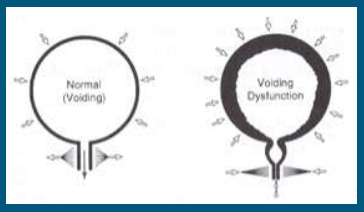
27

(Non-Neurogenic) Dysfunctional Voiding

- Inability to relax sphincter and pelvic floor during voiding in absence of neurological causes
- Staccato pattern & prolonged voiding time w/ active EMG
- Constipation, encopresis & fecal soiling (BBD)
- UTIs, VUR, renal scarring
- Severe form:

Hinman's Syndrome





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Urinary tract infections

- Very common reason for pediatric urology referral
  - 3.5% of children will have UTI every year
- Different etiologies seen at different ages
  - Infants – uncircumcised males, congenital anomalies
  - Toilet training – females with dysfunctional voiding, holding, etc
  - Teenage females – sexual activity
  - *Note: it is always unusual for a circumcised male to get a UTI*

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UTI diagnosis

- Diagnosis requires:
  - *Specimen obtained correctly* – midstream catch, cathed if the child is not toilet trained, or (theoretically) SP aspiration
  - Urinalysis suggests infection (pyuria and/or bacteriuria)
  - Uropathogen on urine culture (>50K CFU)
- Bag specimen in pre-TT children
  - Can rule out a UTI if urinalysis is normal
  - *If urinalysis is abnormal, need to confirm with cath specimen*

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Pediatrics. 2011;128(3):595–610

30

UTI – history/PE

- Symptoms non-specific for infants/young children
  - Fever, nausea, pain, lethargy, irritability
- Older toilet trained children can localize to urinary tract
  - Dysuria, frequency, incontinence, lower abdominal pain
- *Presence of fever important*
- *Prenatal history, family history, history of congenital anomalies*
- *PE similar to incontinence – males note uncircumcised/phimosis*

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AAP UTI Guidelines: Pediatrics. 2011;128(3):595–610

- *2-24 months age range only*
- Proper diagnosis emphasized
- 7-14 day course of antibiotics
- *Renal US indicated for febrile UTI*
- *VCUG not recommended for 1st febrile UTI if renal US normal*
  - Controversial to peds urologists, consider severity of illness
  - Remember the age range of the guidelines
- *VCUG if hydronephrosis, scarring, atypical/complex cases, recurrent febrile*

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Febrile UTI in uncircumcised male infant

- Often admitted to hospital for 2-3 days
- PE normal with expected physiologic phimosis
- Renal US normal
- *Treatment: Consider circumcision or steroid cream to release phimosis. Either can prevent recurrence.*
- VCUG optional if renal US normal

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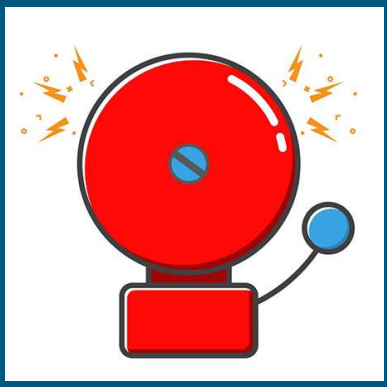
Febrile UTI in circumcised male

- Alarm bells should go off
- *Something is not right*
  - *High grade VUR, posterior urethral valves, nonneurogenic neurogenic bladder, ectopic ureter, etc*
- Not something to ignore



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Febrile UTI in young toilet trained female

- Generally treated as outpatient
- VCUG ~35% with VUR
- *Must treat the bladder/bowel dysfunction*
  - Dysfunctional voiding, Bowel bladder dysfunction
  - Voiding postponement
  - Constipation
- *Subureteric injection of bulking agent less likely to work if BBD present (50% vs 90% per AUA guidelines)*
  - Open surgery works regardless of BBD

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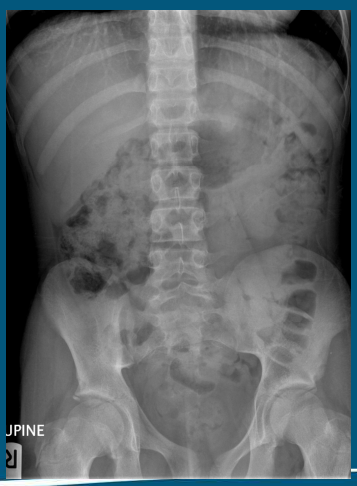
Pediatric Urology pearl

- Everybody is constipated until proven otherwise
- Even when proven otherwise, everybody is constipated
- PEG treats everything ☺



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Courtesy of Armando Lorenzo, MD

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Neurogenic bladder

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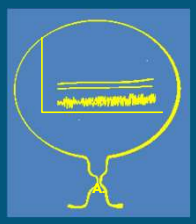
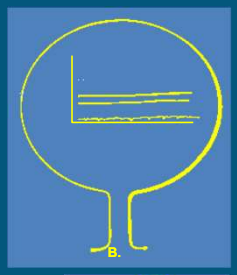
Neuropathic Voiding Dysfunction

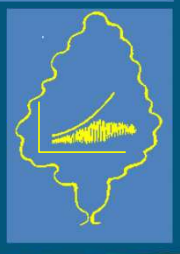
Good bladder
Good sphincter





Good bladder
Bad sphincter

Bad bladder
Bad sphincter



Bad bladder
Good sphincter

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Urodynamics

- Utilize UDS to evaluate bladder capacity, storage pressures, ability to empty
  - If leakage, what kind? (stress, urge, overflow)
  - If voiding, augmented by abdominal straining? Coordinated?
- Determine options for interventions:
- Video/Fluoro additionally assesses:
  - Reflux
  - Bladder outlet – continence options
  - DSD (fluoroscopically)
  - Bladder shape/wall
- Helps with "gestalt" for safety, surgical decisions

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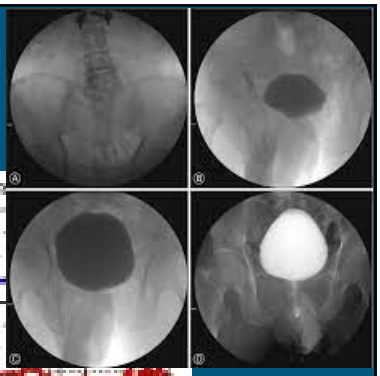
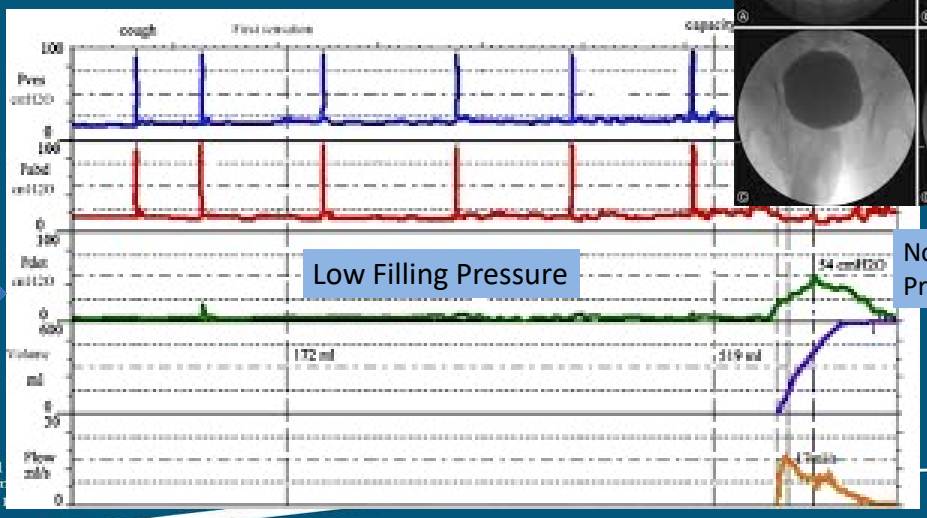
39

What's "Normal"?





Low Filling Pressure

Normal Voiding Pressure Curve

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40

What is Abnormal?

- Dysfunctional voiding
  - Discoordinated activity of BN, EUS, and detrusor
- DSD (neuropathic)
- Bladder pressure of 40cm H_2O is bad (McGuire and Weiss 1981)
  - Impaired ureteral drainage → hydro → decreased GFR
  - Secondary VUR

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Detrusor Sphincter Dyssynergia

- Sphincter either fails to relax or contracts paradoxically with detrusor contraction.
- Cervical or thoracic level spinal cord injury
- Sustained bladder pressure >40cm H2O will inhibit delivery of urine to the bladder and can produce hydronephrosis and renal scarring/functional loss

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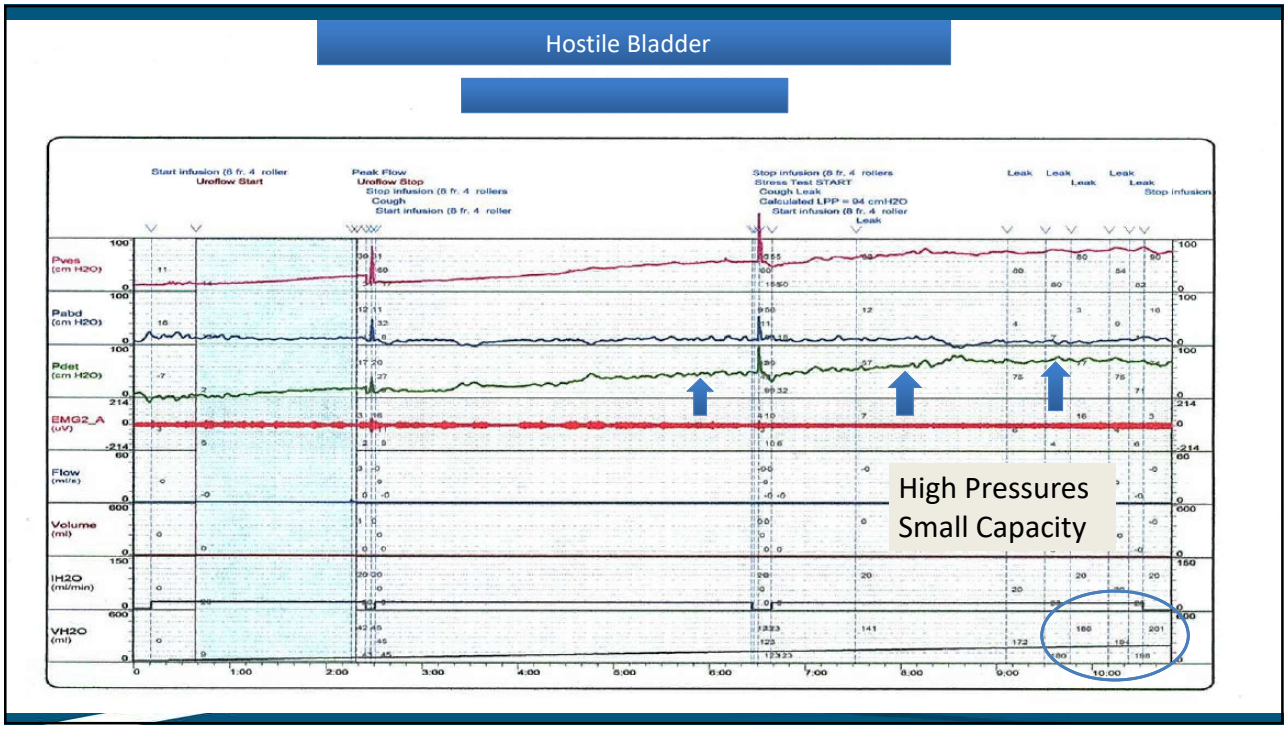

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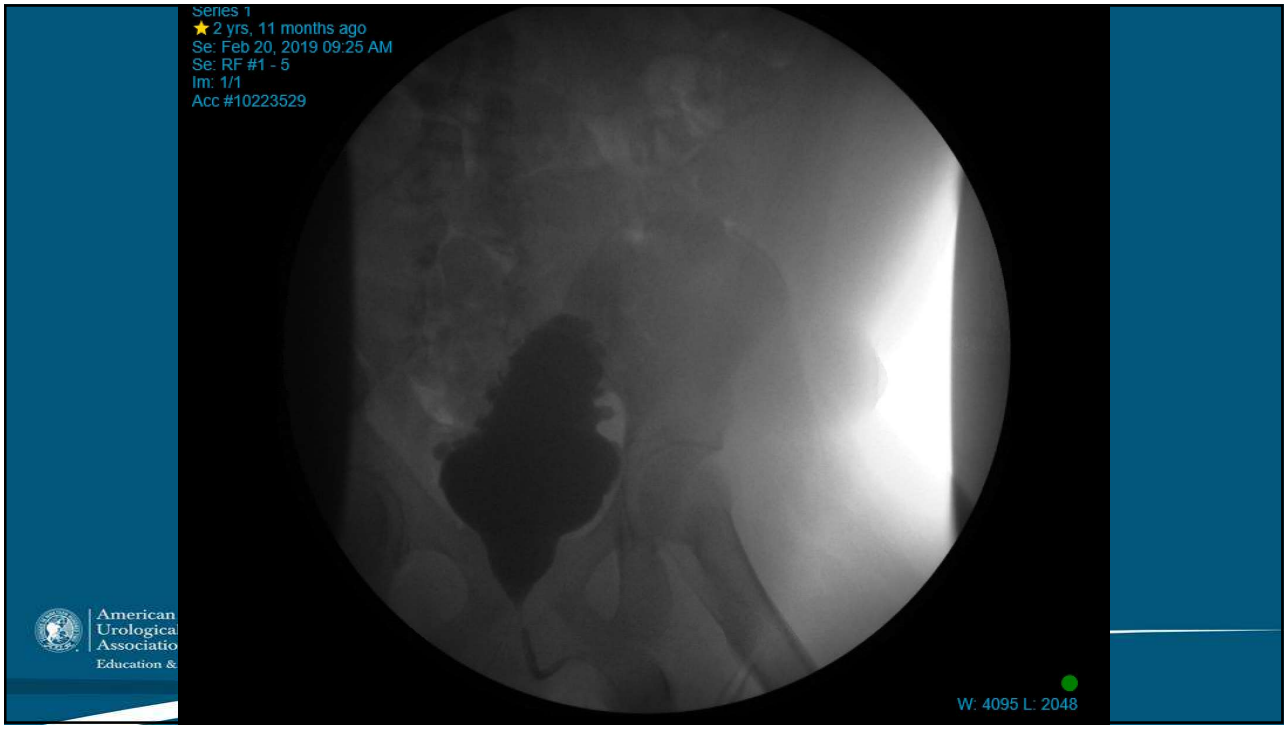

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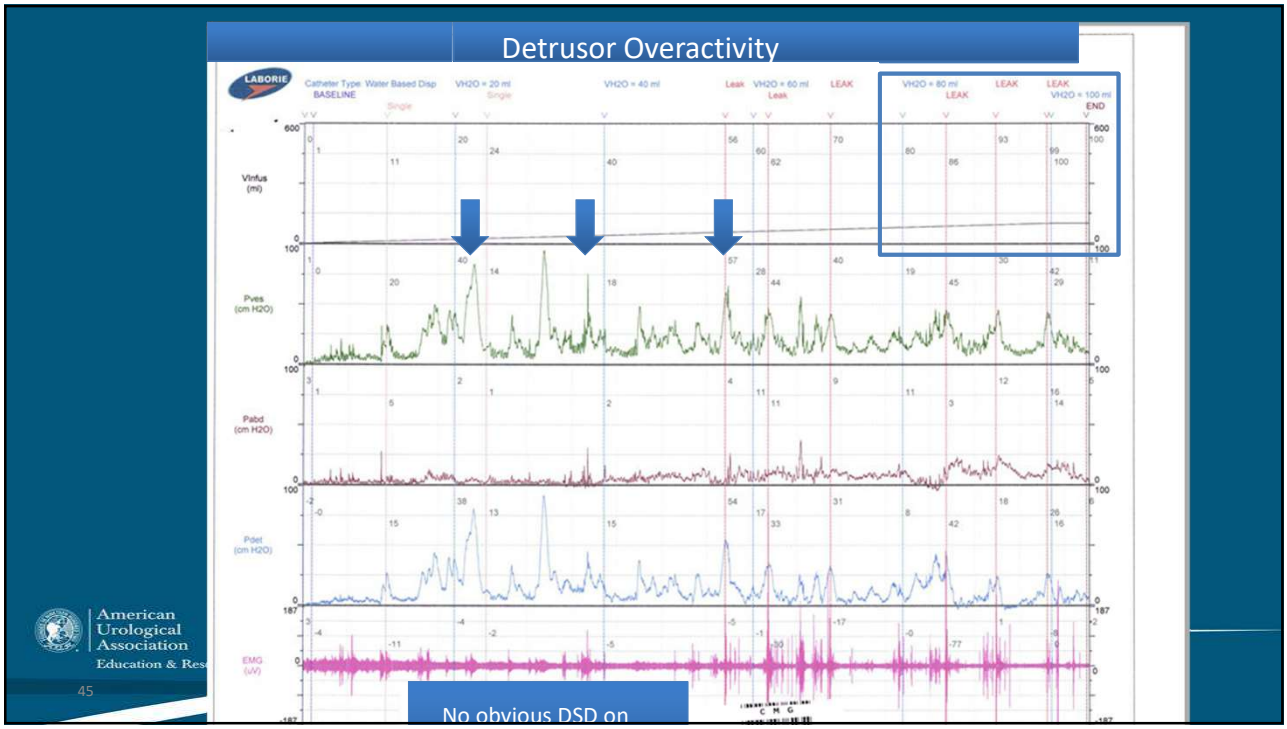

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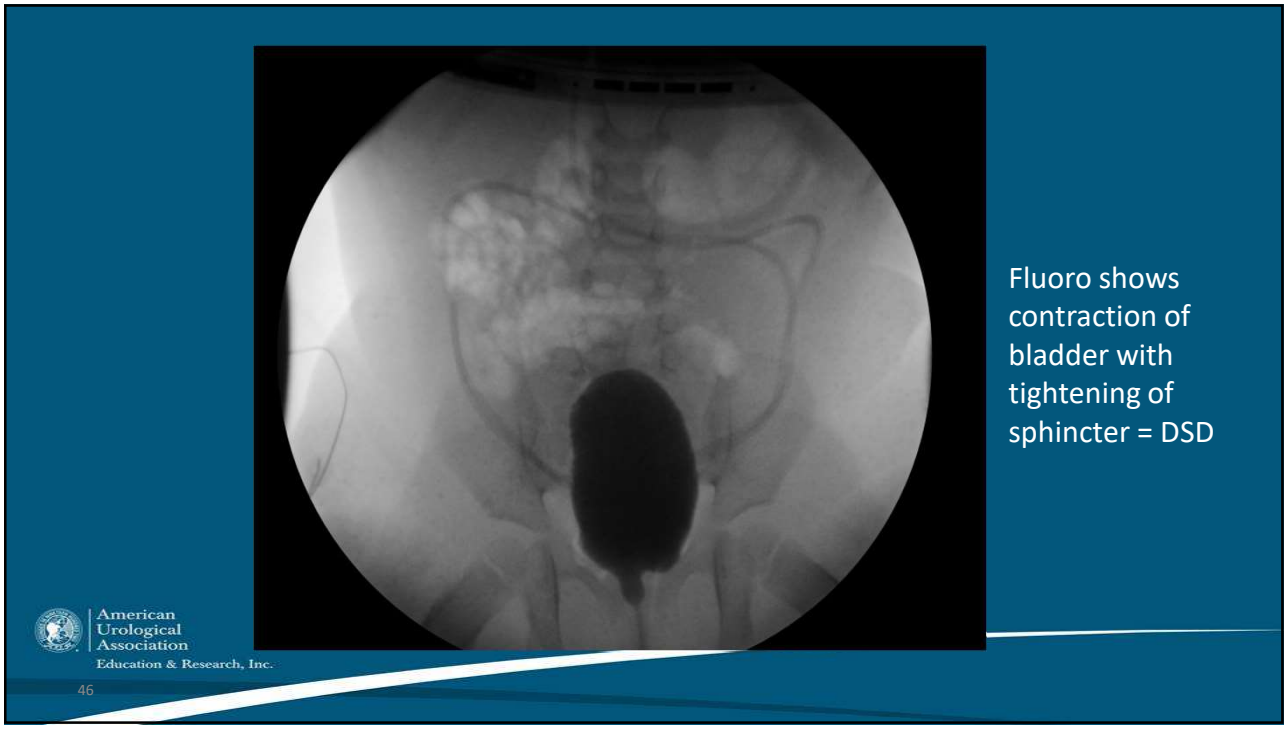
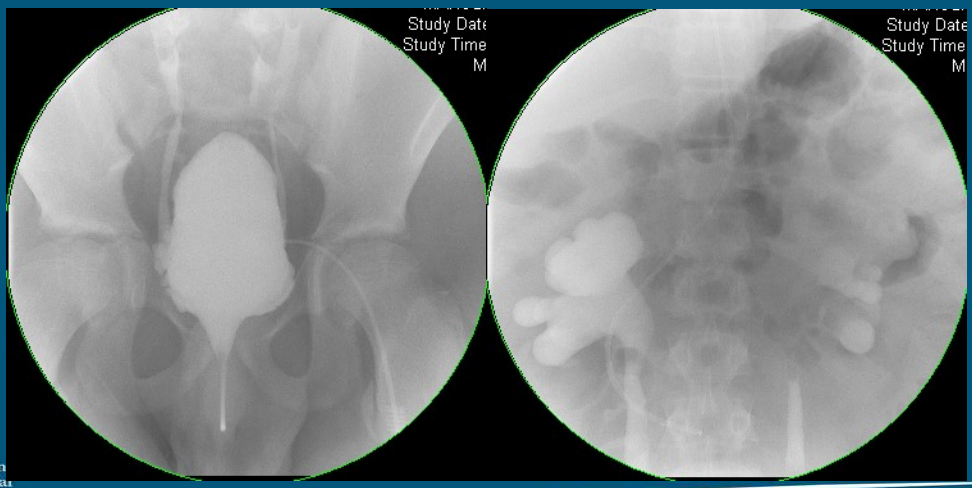


46

In neuropathic bladder, if you have low detrusor pressures during filling on Urodynamics, but early VUR especially if bilaterally, you cannot trust those pressures and should consider the bladder to have abnormal compliance



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Neurogenic bladder - epidemiology

- Causes of neurogenic bladder in pediatric population
  - *Spina bifida, myelodysplasia = most common*
  - Spinal cord injury
  - Non-neurogenic neurogenic bladder
  - Sacral agenesis, VACTERL
  - Other, rare
- Spina bifida is most common: Incidence is decreasing; adults > children now
  - folic acid supplementation decreases risk of spina bifida by 50-70%
  - A mother with a previous child with spina bifida has increased risk of having another

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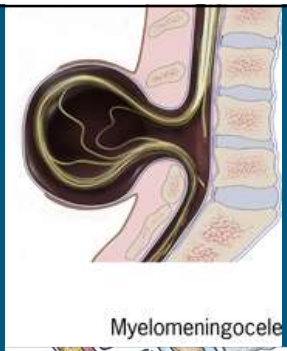
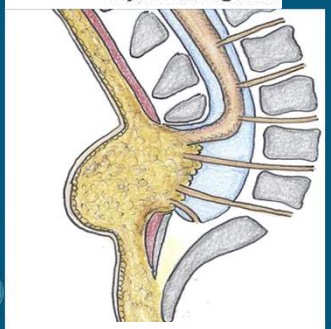
Spina Bifida

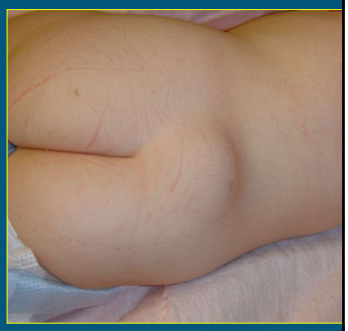


Myelomeningocele



Lipomyelomeningocele

- *Myelomeningocele = meninges and spinal cord protruding*
  - *Major disability, typical spina bifida*
- Lipomyelomeningocele = tethered spinal cord enters an extraspinal meningeal sac and terminates in the lipomatous tissue
  - Range of disability from minor to major
  - Covered by skin

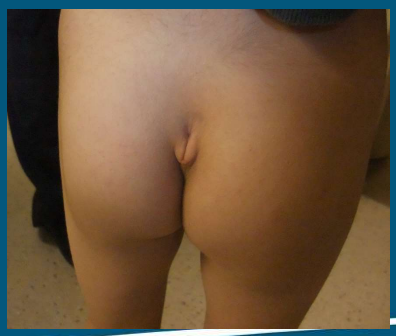


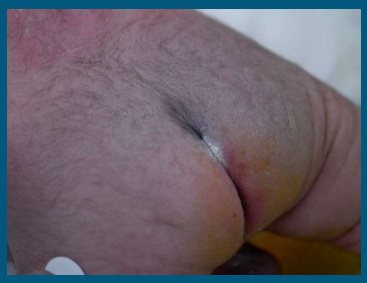
www.cdc.gov/ncbddd/spinabifida/facts.html
Neuropathology. 2017 Oct;37(5):385-392.

49

Spina bifida

- "Closed" spina bifida often diagnosed by sacral dimple or cutaneous abnormality (spinal cord and skin = ectoderm)





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Prenatal closure of spina bifida

- MOMS trial – randomized to prenatal vs postnatal closure
- Clear neurosurgical benefits:
  - *VP shunt 68% vs 98%*
  - *Ambulation 53% vs 23%*
- Unclear if long term urologic benefits
  - MOMS1 at 30 months: equivalent CIC and UDS findings; nonspecific bladder diffs
  - MOMS2 in school age: Decr CIC in prenatal group but criteria subjective
  - Majority will still need CIC with some getting vesicostomy/augment
- Downside: prematurity, respiratory distress, perinatal deaths, risk to mom

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J Urol. 2019 Oct;202(4):812-818.
N Engl J Med. 2011 Mar 17;364(11):993-1004.

51

Spina bifida – infant management

- Spina bifida closure (if not done prenatally), +/- VP shunt
- Assess upper tracts with renal US (most have NORMAL kidneys at birth)
- Assess bladder emptying
  - Q4-6 hr CIC vs bladder scanning
- Use of CIC variable across centers
  - CDC protocol*: continue CIC if >30 mL residuals, DLPP > 40 cm H2O, detrusor overactivity with detrusor sphincter dyssynergia

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*J Urol. 2016 Dec;196(6):1728-1734.

52

Spina bifida – initial imaging findings

- Baseline renal US
  - 56% no hydronephrosis, 40% SFU grade 1-2, 4% SFU grade 3-4
- Baseline cystogram (usually around 3 months)
  - 85% no VUR, 15% VUR
- Baseline DMSA
  - 92% normal DMSA
- *Majority will have normal upper tracts at birth*

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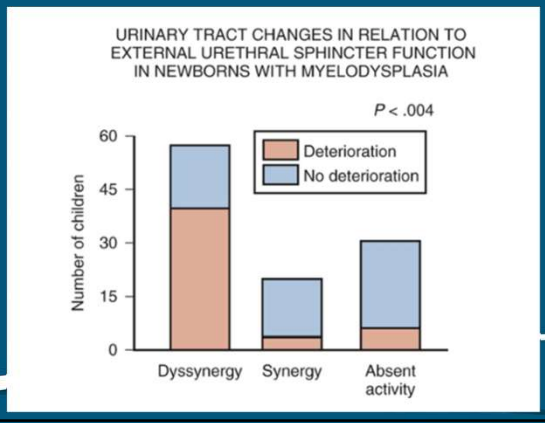
J Urol. 2019 Jun;201(6):1193-1198.

53

Spina bifida – urodynamic findings

- *Presence of detrusor sphincter dyssynergia (DSD) associated with poor outcomes = upper tract deterioration*
  - ~1/3 dyssynergy, ~1/3 synergistic, ~1/3 denervation



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J Urol. 1986 Jan;135(1):90-3.

54

Spina bifida – urodynamic findings

- *A high detrusor leak point pressure (DLPP) is associated with poor outcomes*
  - 40 cm H2O DLPP often cited (based on ~40 patients)

PROGNOSTIC VALUE OF URODYNAMIC TESTING IN MYELODYSPLASTIC PATIENTS

EDWARD J. McGUIRE,* JEFFREY R. WOODSIDE, THOMAS A. BORDEN AND ROBERT M. WEISS

TABLE 3. *Relationship of urethral opening pressure to urethral complications*

	Urethral Opening Pressure	
	<40 cm. Water No. (%)	>40 cm. Water No. (%)
Vesicoureteral reflux	0	15 (68)
Ureteral dilatation	2 (10)	18 (81)

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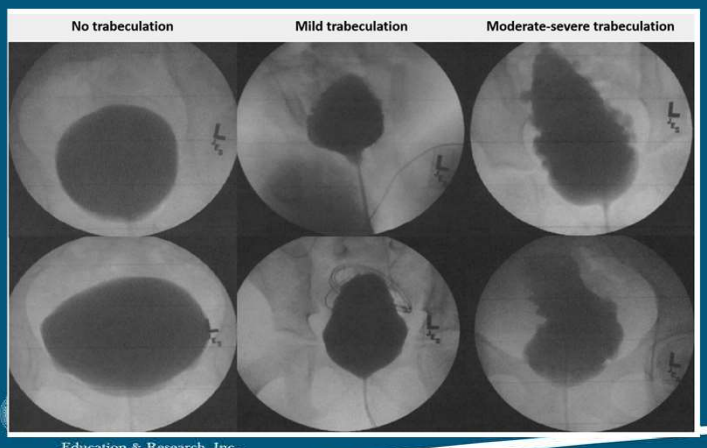
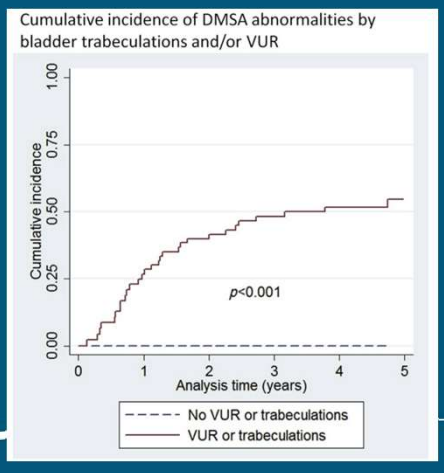
J Urol. 1981 Aug;126(2):205-9.

55

Spina bifida – urodynamics findings

- *Trabeculated bladder and VUR associated with poor outcomes*





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J Pediatr Urol. 2018 Aug;14(4):319.e1-319.e7.

56

Spina bifida – infant management

- Renal US every 3-4 months
- Urodynamics at 3 months and 1 year
- Possible baseline DMSA or if UTIs
- CIC/Anticholinergics:
  - *High risk features on urodynamics*
  - *Febrile UTI*
  - Progressive hydronephrosis
  - "Proactive", "early intervention"

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Spina bifida – high risk bladder

- *Non surgical is first line*
  - CIC, anticholinergics
- Can escalate
  - Overnight catheter drainage
  - Q2 hour CIC
- Intradetrusor botulinum toxin*

Vesicostomy

- Surgical option, can be a relief to families

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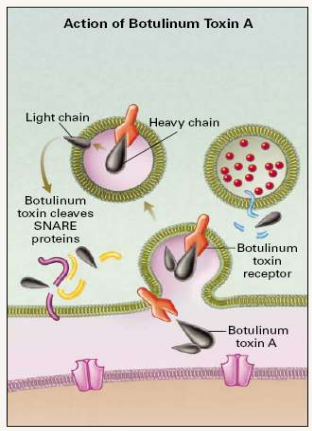
*FDA approved for > 5 years of age

58

Botulinum toxin injection

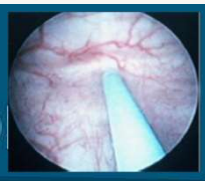


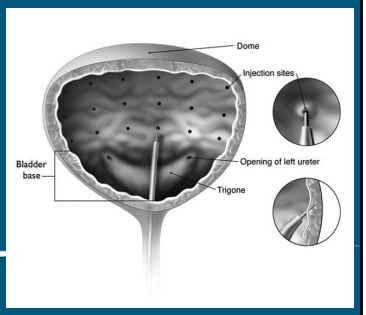
MOA: inhibits presynaptic release of acetylcholine from nerve terminals, reducing stimulation of the muscarinic receptors

For children: 10 international units /kg up to 200 iu

Typically in OR, some specialty clinics done awake

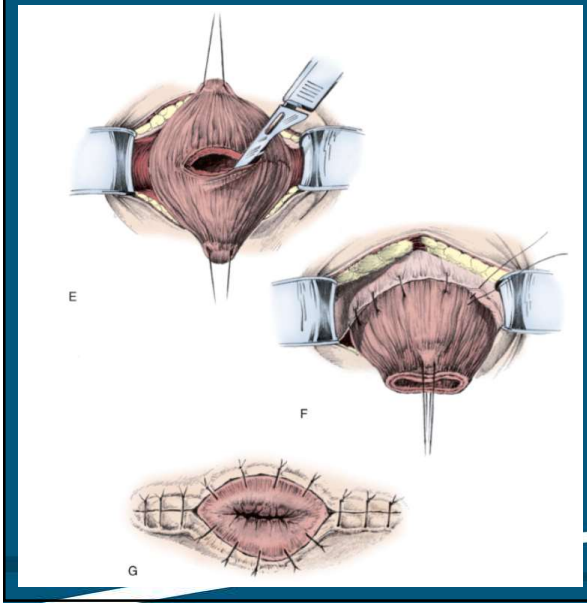
If for high risk, repeat UDS at 2-3 months
Repeat treatment on average every 6 mos





59

Vesicostomy



- Protects kidneys
- Stops need for CIC
- Risks:
  - Prolapse: Reduce manually; revision to secure posterior wall/dome to abdominal wall
  - Stomal stenosis: Dilate; revision
- Will likely need bladder augmentation when taken down
- Can opt to keep forever

60

CIC for continence

- Age 3-5 typical
- *Most will need anticholinergic*
  - Oxybutynin 0.2 mg/kg BID-QID most common (FDA approved > 5 yrs)
  - If not tolerated -> intravesical Ditropan option and botulinum toxin
- Mirabegron suspension available (FDA approved > 2 yrs)
- CIC Q3-4 hours typical
- *A significant minority will not be continent despite Q2 hr CIC, maximum anticholinergic*
  - No good medication for sphincteric deficiency
- Goal is not complete dryness for all families

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Bowel continence

- Bowel continence is also goal and constipation can affect bladder capacity, uninhibited contractions, and ability to empty bladder fully
- Bowel program usually starts around same time as bladder program if not earlier
  - Something to help soften stool
    - Fiber, lactulose, senna, polyethylene glycol
  - Something to help empty
    - Timed toileting, suppository, enema, cone enema, Peristeen© system
    - Antegrade enemas (Malone antegrade continence enemaAKA MACE)
    - Colostomy
- A lot of trial and error
- *Small amount of bowel incontinence affects QOL more than small urinary incontinence**

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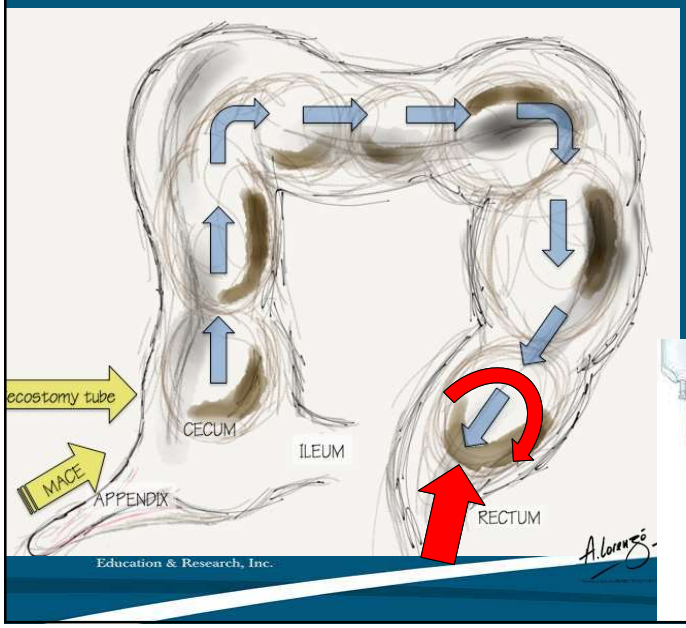
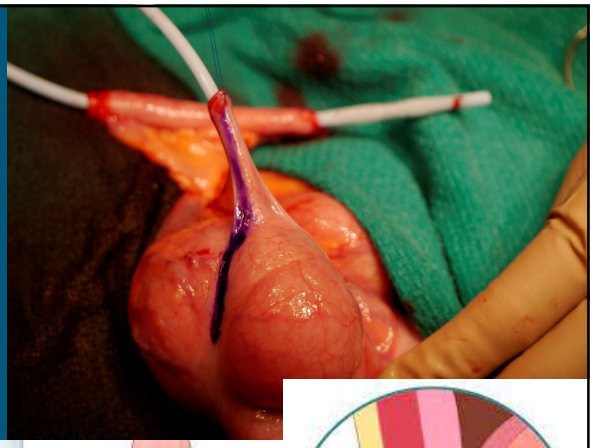
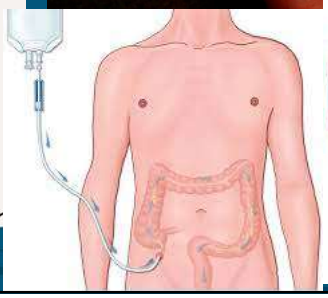
*J Urol. 2017 Mar;197(3 Pt 2):885-891.

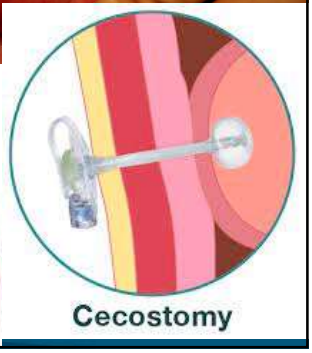
62

MACE or Cecostomy Tube









63

MACE complications

- Stomal stenosis / stenosis at the fascial level
  - Betamethasone, dilation, open revisions rare
  - ACE Stopper
- False Passage
  - Leave a catheter in for a week
- Problems with cleanouts
  - Typically use 500-1000c of water or saline
  - Additives: glycerin, castile soap, miralax, dulcolax

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Reconstructive surgery indications

1. Protect kidneys from high risk bladder

2. Achieve continence

3. Achieve independence

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Reconstructive surgery options

- 3 options for bladder reconstruction
  - Bladder augmentation = make bladder bigger with bowel
  - Catheterizable channel = appendicovesicostomy or Monti channel
  - Bladder neck reconstruction = make less likely to leak from urethra
- *If indication to protect kidney from high risk bladder*
  - *Generally do not need bladder neck reconstruction*
- *If indications are to achieve continence or independence*
  - *Surgery is individualized and should be patient/family driven*

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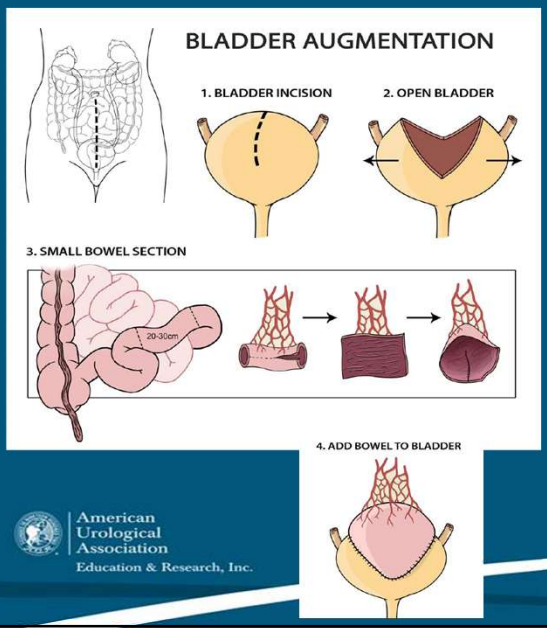
Bladder augmentation



- Ileum most commonly used
  - Less contractions than colon
  - Less mucus than colon
- Detubularize bowel
- Bivalve bladder aggressively (prevent hour glass deformity)
- *Ileum: Hyperchloremic, hypokalemic metabolic acidosis*
- *Colon: Hyperchloremic, hypokalemic, metabolic acidosis*
- *Stomach: Hyponatremic, hypochloremic metabolic alkalosis*

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Augment with Sigmoid

- PROS:
  - Easily mobilized
  - Excellent backing for implants
  - Often redundant
  - Good option for adult re-do cases
- CONS
  - More mucus & unit contractions Higher risk of perforation?
  - Hyperchloremic, hypokalemic metabolic acidosis

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Complications of Augmentation

General Major Abdominal

- Wound infection
- Catheter malfunction
- Bowel obstruction/leak
- Shunt problem

Augmentation-specific

- Mucus
- Inadequate dynamics
- Metabolic acidosis
- B12 deficiency
- Stones
- Perforation
- Tumor

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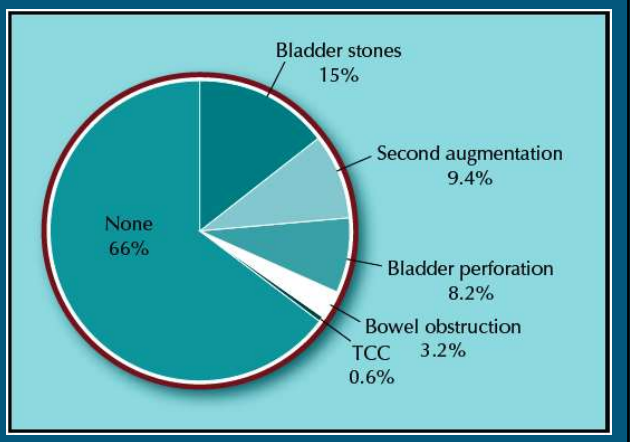
What are the long term risks?

- The Indiana 500+ series
- **34%** surgical complication rate
  - 4 deaths:
    - 3 from **malignancy**
    - 1 from **perforation**
- Concern about bladder cancer
  - Likely not increased over baseline risk
- ~10% re-augmentation



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SHUNT INFECTION AND MALFUNCTION AFTER AUGMENTATION CYSTOPLASTY

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- Very low risk (<2%) with care
- Isolate shunt
- Vigilance postoperatively
  - Headaches
  - Confusion
  - Nausea, vomiting, pain

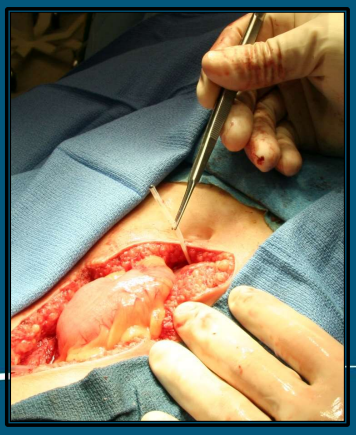

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Table 2. Recommendations for routine surveillance of patients who undergo asymptomatic augmentation cystoplasty.

Annual history and physical examination (to screen for gross hematuria/symptoms)	Recommended
Annual renal/bladder ultrasound	Recommended
Annual electrolyte panel, vitamin B_{12} level	Recommended
Annual screening cystoscopy	Not recommended
Annual screening cytology	Not recommended

Table 3. Recommendations for work-up of patients who undergo symptomatic augmentation cystoplasty.

Cystoscopy/cytology/ultrasound for gross hematuria	Recommended
Renal/bladder ultrasound for suprapubic pain/frequency	Recommended
Electrolyte panel, vitamin B_{12} level for fatigue, anorexia, neurological sequelae	Recommended
Cystoscopy/cytology for recurrent urinary tract infections	Not recommended
Cystoscopy/cytology for microhematuria	Not recommended

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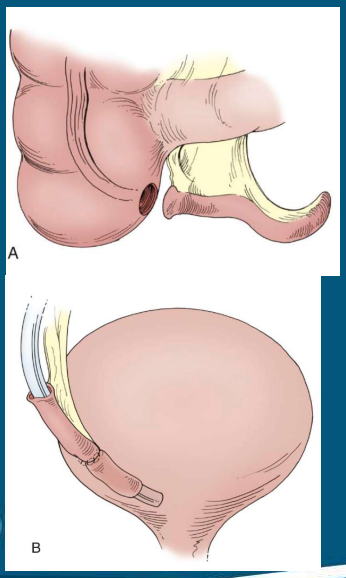
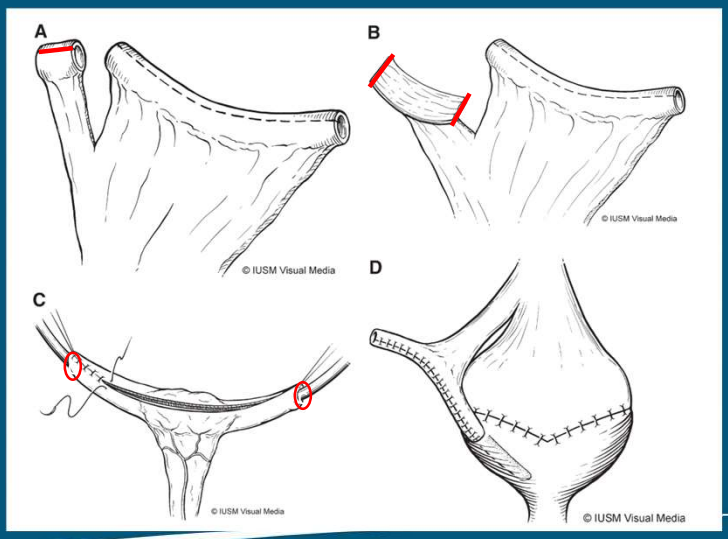
Catheterizable channels





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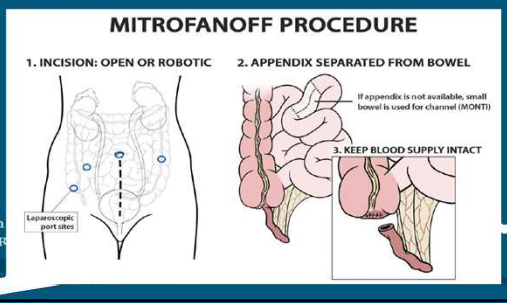
Catheterizable channels

Appendicovesicostomy

- Less long term need for revision
- Sometimes appendix not suitable
  - Short; can't get a 12 Fr at least thru it
- Can split appendix for MACE and APV if long enough



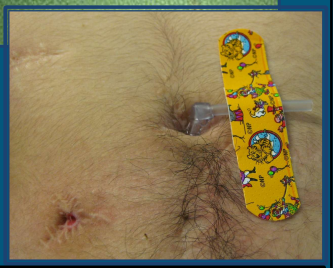
Monti channel

- *Higher risk of revision*
- *Bowel anastomosis needed*
- Can have progressive dilation of channel
- Spiral type or Double Monti can create longer channel
  - Higher complications

74

Stomal Complications

- Stenosis → Steroid ointment, L-stent or MACE stopper (silicone to remodel); revision (10-30%)
- Difficult catheterization / False passage → Emergency if BN closed! leave catheter
  - Glide wire with Council tip catheter; Urgent needle decompression and retry catheter
- Incontinence --> treat bladder pressures first; bulking agent; revision to tunnel channel into bladder further
- Polyps and granulation tissue → silver nitrate



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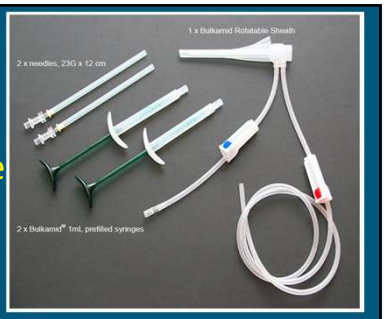
Outlet Procedures

- Offered to select patients with stress incontinence and acceptable bladder storage parameters
- Bulking agents
  - Limited efficacy; consider channel
- AUS
  - Option; not done as much due to risk of infection (augment), erosion (CIC)
- Sling alone
  - Pubovaginal in females; decr efficacy if ambulatory



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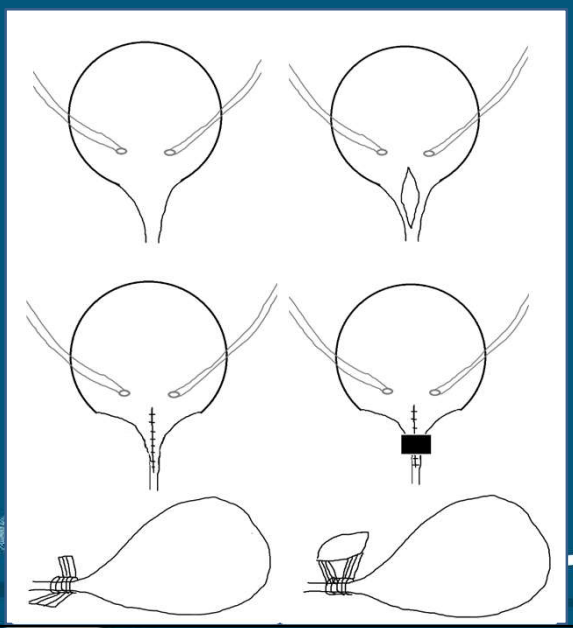
Bladder neck reconstruction



- Many eponyms
- Tighten bladder neck area
  - -Excise tissue and close
  - -Wrap something around it
    - -Rectus fascia, SIS
- May not be able to continue to catheterize through urethra
  - +/- Catheterizable channel
  - +/- Augment
- *If done without augment, 30% will go on to need one*

77

Spina bifida post augment

- New urinary leakage
  - Workup: rule out bowels as problem; UDS to assess pressures, rule out "hourglass"; imaging for stones
  - If pressures high, capacity low: re-start medications or perform Botox injections to the bladder plate
  - Irrigate irrigate irrigate

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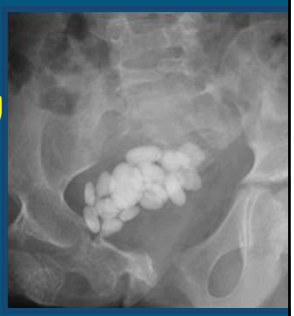
Spina bifida – bladder stones



- *Common, up to 30% at 10 years after augment*
  - *Mucous can be nidus (gentamicin irrigations may help)*
- More common if channel with augment
  - cathing per urethra likely empties better
- Symptoms: pain, hematuria, incontinence, UTIs
- Treatment = cystoscopic, percutaneous (if BNR), open
- Prevention: hydration, ensure emptying
- *Irrigation with 240cc DAILY*

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Spina bifida – long term issues

- Symptomatic tethered cord
  - Worsening LE function or bladder function with growth → repeat UDS, consult neurosurgery!
- Male sexual function
  - *Dependent on level, more likely to have erectile/ejaculatory function*
  - Some erectile dysfunction responds to PDE 5 inhibitors
- Female
  - Most can bear children
    - 3-4% risk of spina bifida in child if one parent with it, >10% risk if both parents
  - *Augment/APV vascular pedicles shift and typically not affected*
    - *Avoid vascular supply, but if divided -> observe*
  - *Higher risk of pyelonephritis during pregnancy with bladder reconstruction*
  - *If C-section and prior bladder surgery, coordinate to have urologist present*

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Spina bifida – long term issues

- Transition to adult urology providers
  - Individualized process
  - Insurance status
    - Private insurance vs government
  - Medical complexity
    - CIC/medication alone vs complex reconstruction
  - Urologic care is the cornerstone of adult spina bifida care
    - Congenital issues compounded by adult: prostate health; bladder cancer, etc

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Cerebral Palsy

- Condition: Static injury to the brain at childbirth
- Childhood:
  - Tend to have spastic bladder at first
- Teens / Adult:
  - Usually flaccid , low pressure
    - Often void only 2-3 times per day, large volume
  - 1/3 will need a urologic intervention due to increasing UTIs, hydronephrosis, worsening incontinence
    - SPT or MicKey button; Catheterizable channel (Monti, Mitrofanoff) +/- bladder augmentation

- Annual Renal Bladder Ultrasound
- Basic Metabolic Panel
- Urinalysis

- Urodynamics

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Developmental anomalies of the bladder

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Bladder agenesis and bladder hypoplasia

- Bladder agenesis
  - Very rare 1:600,000
  - Associated with renal dysplasia and other neurologic/orthopedic issues
  - *Survival if ureters drain to mullerian structures or rectum in males*
- Bladder hypoplasia
  - Associated with lack of urine production or bladder filling
  - Bilateral ectopic ureters, renal agenesis, exstrophy
  - Some bladders will grow with urine flowing into, but augmentation maybe needed

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Classic bladder exstrophy

- 1/50,000 births
- Epispadias, open bladder, orthopedic anomalies
- Closure in specialized centers
- VUR common after closure
- Inguinal hernias common
- *Need for reconstructive surgery for continence common*
  - *Augmentation, APV, bladder neck reconstruction or closure*
- *Generally normal spinal cords and brains*

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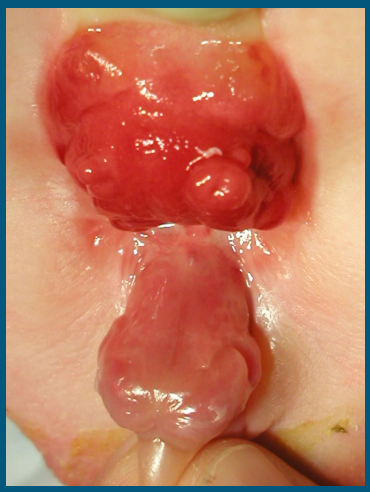
Exstrophy-epispadias complex

- Results from failure of mesodermal ingrowth into the *cloacal membrane, leading to premature rupture*
- Once closed, VUR is common
- Bladder exstrophy ***not*** associated with spinal or upper tract abnormalities… but



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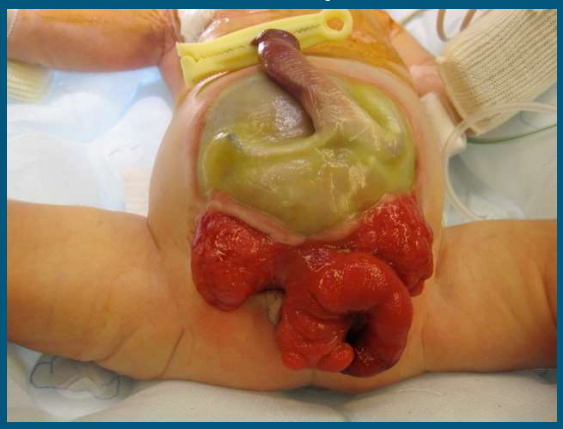
Cloacal extrophy *IS* associated with multiple anomalies



Abnormalities of the spinal cord/vertebral column in 85-100%

Most patients have lumbar myelodysplasia (80%)

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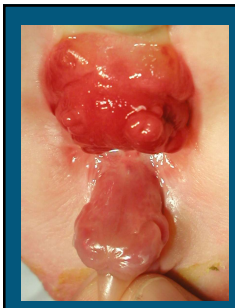

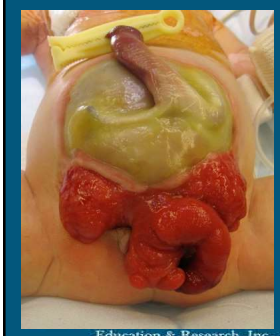

Exstrophy Epispadias complex

- Bladder exstrophy
  - No bowel or limb abn
  - Newborn closure with osteotomies
  - Epispadias repair
  - Long term challenges: sexual and urinary
    - (Male) sexual function due to dorsal curvature and short corporal bodies
    - Normal testes, neurovascular bundles
    - Only 25% will void volitionally → Rest require augment, channel, etc
- Cloacal Exstrophy
  - "OEIS": omphalocele, exstrophy, imperforate anus, spinal abn"
  - Colostomy
  - Spina bifida = lower extremity abn
  - Bladder closure in 2 stages, first to bring halves together, next to close with vesicostomy
  - Unlikely to be continent as adult
    - Almost exclusively diversion

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Male epispadias

- 1/100000 births
- *The more severe, the more likely sphincter affected and continence to be an issue*
- *Normal kidneys*
- *Normal spinal cord and brain*
- Repair using hypospadias like techniques
- May need procedures for incontinence → Bladder neck repair

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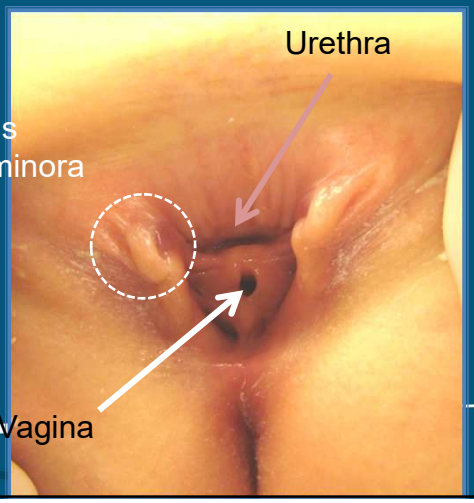
Female epispadias



Bifid clitoris
No hood/minora

- Abnormal external appearance
- Incontinence due to abnormal bladder neck

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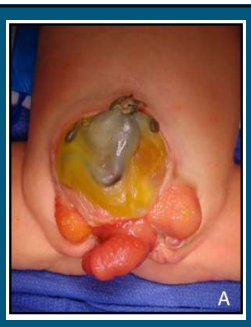
Cloacal exstrophy



- Cloacal exstrophy ≠ persistent cloaca
- Cloacal exstrophy ≠ classic bladder exstrophy
- *Much more severe malformation*
  - *Hind gut open, often 2 phallic structures, omphalocele*
  - *Myelodysplasia*
  - *Renal anomalies*
- Generally will need bladder reconstruction for continence
- Typically have colostomy

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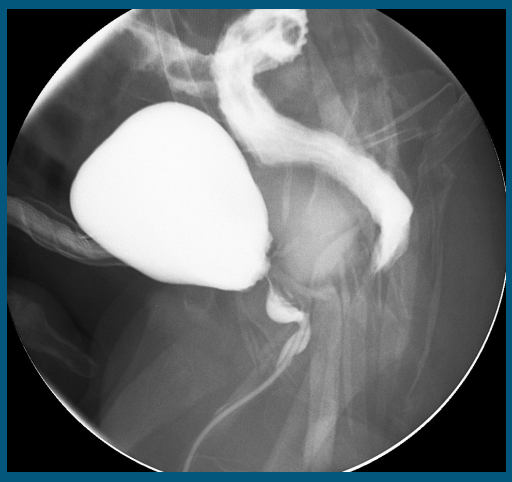

Cloaca is a complex ARM found in girls

The GI, reproductive, and urinary tracts join as a common channel

Empties onto the perineum as a single orifice

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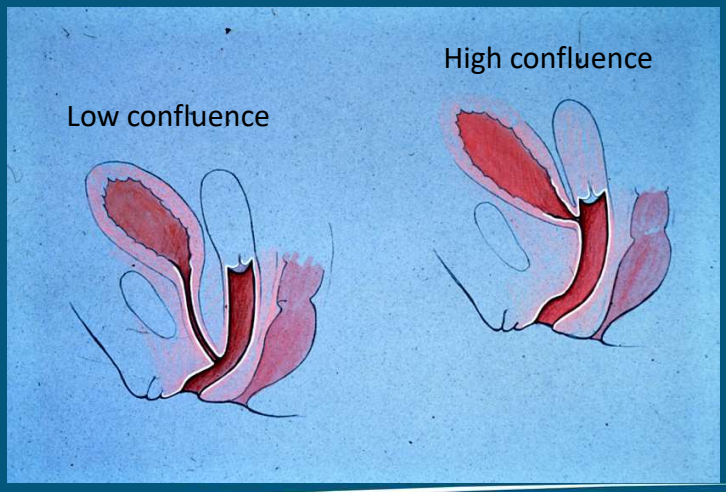
UG Sinus

- Urinary and reproductive tract form together
  - Rectum is separate
- Isolated; Congenital Adrenal Hyperplasia (CAH); other ARMs
- Diagnosis: Exam, ultrasound (hydrocolpos?), cystoscopy possible genitogram
- Tx: surgery to separate



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Ano-rectal malformations (ARMs)

- 25% of children have findings consistent with NGB
- NGB is more common with concomitant spinal anomalies
- Iatrogenic neuropraxial injury may occur during primary repair, and is more likely in severe ARM
-
- ~30–40% of children with ARM will have VUR found on VCUG
- CKD and ESRD are the greatest cause of decreased life expectancy in patients with ARM. Patients with cloacal anomalies are at the highest risk

	Krickenbeck (2005)[4]
Male	Rectoperineal fistula
	Rectourethral bulbar fistula
	Rectourethral prostatic fistula
	Rectobladder neck fistula
	Imperforate anus without fistula
Female	Rectoperineal fistula
	Rectovestibular fistula
	Cloaca with short common channel (<3 cm)
	Cloaca with long common channel (>3 cm)
	Imperforate anus without fistula
Complex	Rectal atresia/stenosis
	Posterior cloaca
	Cloacal exstrophy
	Associated presacral mass
	H-type fistula
	Rectovaginal fistula
	Pouch colon

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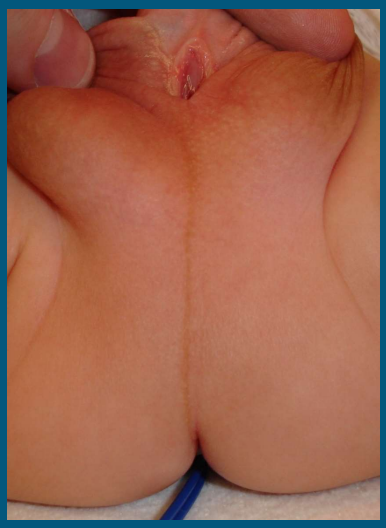

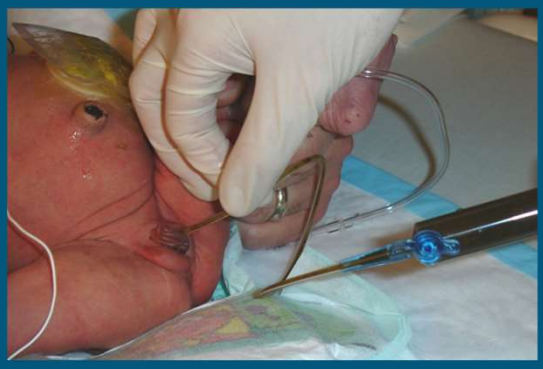

Recto-urethral and rectovesical fistula in boys, recto-vestibular fistula in girls (70%–80%)
Rectovaginal fistula or a cloacal anomaly (1-5% of females)

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Duplicated bladder

- Very rare
- Complete = usually 2 separate bladder and 2 urethras
- Partial = usually 2 bladders and 1 urethra
- Duplication anomalies of genitalia and hindgut common
- VUR, renal dysplasia, renal ectopia common
- *Presentation is widely variable and treatment individualized*

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Megacystis Microcolon Intestinal Hypoperistalsis Syndrome

- Prenatal diagnosis suspected
  - Large bladder with normal to high amniotic fluid levels
- Postnatal diagnosis suspected
  - Large bladder, failure to pass meconium, abdominal distention, vomiting
- Treatment
  - Ileostomy, G-tube, +/- TPN, +/- intestinal transplant
  - CIC, vesicostomy, timed voiding in minority

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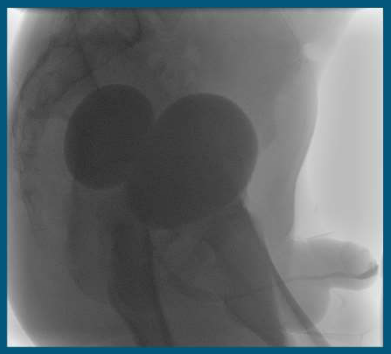
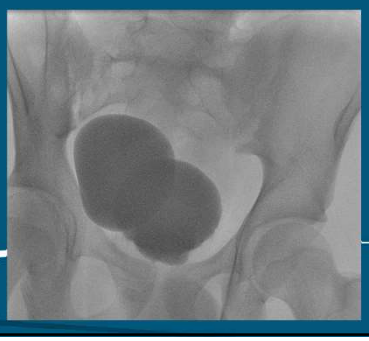
Bladder Diverticula

- Can be found for evaluation for UTI, incontinence
- Diagnosed by VCUG
- *Paraureteral diverticula most common*
  - Less likely to resolve in males
  - Bladder otherwise normal
- Indications for surgery
  - Hydonephrosis/hydroureter, UTIs, VUR associated
  - Excision of diverticula, +/- ureteral reimplant





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Urachal anomalies

- *Patent urachus*
  - *Can be associated with obstruction (PUV, urethral atresia, etc)*
  - *Generally surgery needed*
- Urachal cyst
  - If infected -> treatment is ABX, +/- drainage, followed by excision
- Urachal diverticulum
  - Open to bladder, rarely leads to issues unless obstruction
- Urachal sinus
  - open at umbilicus
- Asymptomatic urachal remnant
  - Some advocate for removal (undefined cancer risk), some only if symptoms, choose your adventure

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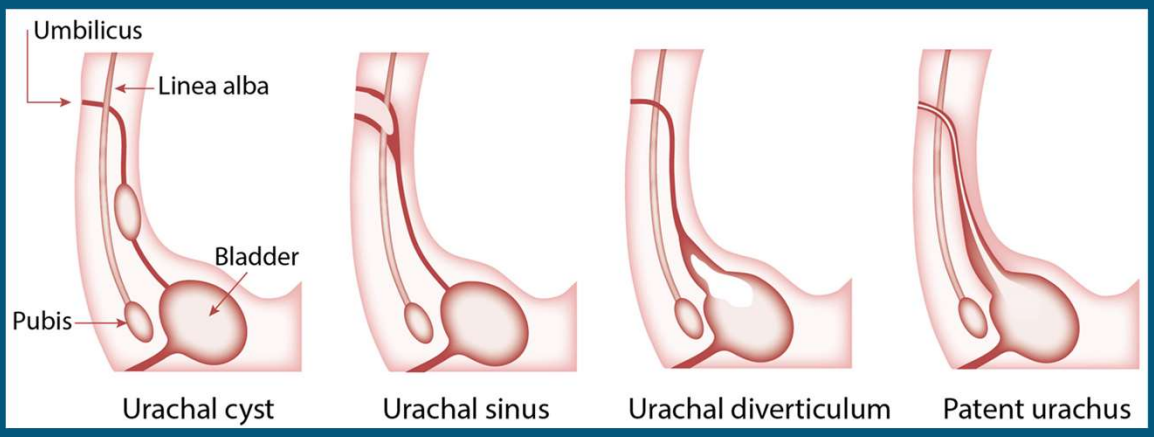
Urachal anomalies



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Pediatric Metabolic Stones

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Pediatric stone disease

- Increasing incidence
- Etiology – more likely to be genetic than adults
  - Dietary, genetic, metabolic, anatomical
- Diagnosis
  - *Ultrasound used initially*, selective use of CT scan
- Treatment, similar to adults
  - ESWL
  - *Ureteroscopy – may need passive dilation with stent, but routine prestenting should be avoided (AUA guideline)*
  - PCNL – mini perc options

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Metabolic evaluation

- 1st time stone former
  - Electrolytes, serum calcium, +/- PTH, UA
  - *24 hour urine collection = AUA guidelines state for "all" children*
    - *Most peds uro will get for prepubertal, multiple stones*
    - *If teenager with solitary stone, some advocate no 24 hr urine collection*
- Recurrent stone former: same + nephrology referral
- Most common: Hypercalciuria

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General prevention & treatment

- Similar to adults
  - Hydration, lower salt, increase citrate
  - *However, should not restrict protein intake especially younger child*
- Medical management similar to adults
  - Alpha blocker -> possible promote passage
  - Thiazide, potassium citrate
  - Off label usage

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Kidney stones in premature infants

- *Often related to use of Furosemide diuretic*
  - *Used to treat respiratory disease from bronchopulmonary dysplasia*
- *Most resolve spontaneously*
  - ~15% progress to have surgery
  - Urine Ca/Cr ratio > 0.4 predictive
- Monitor with serial renal US

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Cystinuria

- Autosomal recessive
  - Amino acid transporter of cysteine, ornithine, lysine, argenine (COLA)
- Only cysteine crystalizes in urine -> stones
- Diagnosis
  - *Hexagonal shaped crystals* or positive nitroprusside test
- 70% develop CKD
- *Treatment: hydration, alkalization of urine, cysteine binding medications*
  - Binders: Tiopronin aka alpha-mercaptopropionyl glycine (1st line), d-penicillimine (2nd line). FDA approved.

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Primary hyperoxaluria

- Autosomal recessive
  - Increased oxalate production by liver
- Rare, but accounts for 1-2% of pediatric ESRD patients
- Treatment
  - Pyridoxime (vit B6), Lumasiran (RNA inhibitor, FDA approved)
- *Kidney-Liver transplant needed for ESRD*
  - *Liver is main source of hyperoxaluria*

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Lesch-Nyhan syndrome

- X-linked
- Mutation in HPRT1 gene -> high uric acid levels
- Associated with developmental delay, *self harming behaviors*
- *Treatment*
  - *Allopurinol -> reduces production of urine acid, FDA approved*
    - *Xanthine oxidase inhibitor*
  - *Overtreatment -> xanthine stones*

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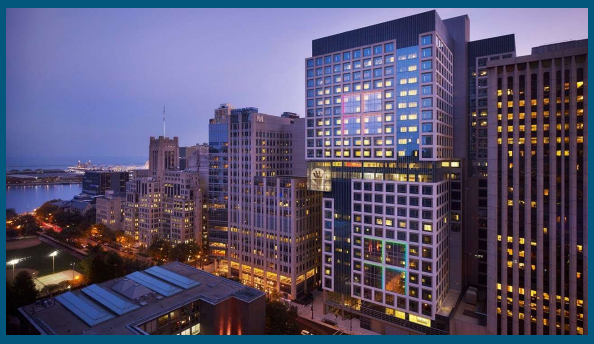
Thank you



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109