

## **CRACKCast E174 – Genitourinary and Renal Tract Disorders**

### Key concepts:

#### Priapism

• *"In low-flow priapism,* cavernosal aspiration plus irrigation has been effective when performed within the first 48 hours, and preferably within a few hours, of symptom onset. Phentolamine, phenylephrine, ephedrine, or 1:1,000,000 epinephrine can be added to the irrigation solution used in performing corporal aspiration."

#### Phimosis and Paraphimosis

• "Steroid cream is first-line therapy for phimosis. In paraphimosis, most can be reduced utilizing a number of techniques, only in severe cases involving vascular compromise of the glans penis, may a dorsal slit procedure be necessary."

#### **Testicular Torsion**

• Delay in diagnosis and treatment can result in loss of spermatogenesis and, in severe cases, a necrotic, gangrenous testis.

• Color Doppler ultrasonography is the test of choice, but false negatives due occur.

• Testicular salvage rates are 96% if detorsion is performed less than 4 hours after symptom onset; with more than a 24-hour delay, the salvage rate falls to less than 10%.

#### Varicoceles

• Left-sided varicoceLes account for 85 to 95% of the cases.

• Right-sided varicoceles are often caused by inferior vena cava thrombosis or compression by tumors.

#### Urinary Tract Infections

• In children younger than 2 years, a urinalysis alone is inadequate to rule out a urinary tract infection; urinalysis yields false-negative results in 10%–50% of patients. Urine cultures should be sent in children less than 2 years of age. (Cultures can also have false negatives - up to 25%)

• Girls less than 2 years old, and boys, uncircumcised, less than 1 year, and circumcised less than 6 months are at higher risk for UTIs.

• Children less than 2 years old should be considered to have upper tract disease and receive antibiotic treatment for 7–14 days.

#### **Renal Stones**

• Renal stones are more common than ureteral stones in younger children.

• Older children can present with classic renal stones signs and symptoms; younger children may present with more non-specific symptoms, such as malaise and non-tender abdominal pain. Ultrasound should be the first–line imaging modality used in children with suspected renal stones.

#### Poststreptococcal Glomerulonephritis

• Patients will present with a history of a pharyngeal or skin infection in the previous 2–6 weeks; clinical findings are usually limited to the urinary tract including hematuria, flank pain or sometimes generalized edema.

• Diagnostic testing will show blood, protein, and RBC casts in the urine; evidence of renal dysfunction (elevated BUN) and low complement levels will also be found.

• Treatment includes restricting fluid and diuretics for more significant disease.



#### Nephrotic Syndrome

Albumin and immunoglobulin levels are typically both decreased in nephrotic syndrome.
Clinical signs include periorbital edema, weight gain, and more serious signs such as pulmonary edema or ascites.

• Children with nephrotic syndrome are at increased risk for thrombosis and bacterial infections, especially Streptococcus and E.coli.

• Treatment includes corticosteroids and diuretics.

#### Henoch-Schönlein Purpura (HSP)

• HSP is an immunoglobulin A-mediated systemic vasculitis that involves the skin, GI tract, joints, and kidneys.

• Urinalysis may be positive for blood and RBC casts.

• Corticosteroids can be useful for severe abdominal pain.

#### Hemolytic-Uremic Syndrome (HUS)

• HUS is a microangiopathic hemolytic anemia found in young children. In the U.S., it is typically related to Shiga-toxin producing E.coli (STEC), presenting with abdominal pain and bloody diarrhea. Streptococcus pneumoniae has also been implicated.

• Renal vascular endothelium injury results in renal insufficiency and damage to RBCs; glomerular damage occurs due to platelet, complement, and fibrin deposits.

• Peripheral smear will show damaged RBCs and typically thrombocytopenia; renal involvement ranges from hematuria to elevated BUN and creatinine.

• Stool cultures and Shiga toxin testing should be sent in diarrhea-associated cases.

• Antibiotics are not indicated in HUS unless a concurrent presumptive pneumococcal infection such as pneumonia is present.

### **Core questions:**

- 1. List a DDx for priapism and describe treatment
- 2. Describe the management of a paraphimosis.
- 3. Describe the management of a phimosis.
- 4. What is the pathophysiology of balanoposthitis? What is the most common bug in balanoposthitis?
- 5. How is balanoposthitis managed?
- 6. Describe the diagnosis and management of pediatric epididymitis and Orchitis
- 7. Describe the diagnosis and management of testicular torsion
- 8. List common bacteria in peds UTI and describe treatment <2 months, 2M-2Y, 2Y+.
- 9. List 6 ddx renal mass how should these be imaged first?
- 10. Give a differential diagnosis of proteinuria in children.
- 11. What is the most common cause of proteinuria in children?
- 12. Provide a differential diagnosis of acute renal failure in children. What is the most common cause?
- 13. Describe the clinical presentation of glomerulonephritis in a child.
- 14. Define hypertension in a child.
- 15. List causes of hypertension in a child.
- 16. Llist 2 medical treatments for acute HTN crisis in children
- 17. What are the two types of HUS? What are the typical presenting features?
- 18. Describe the management of HUS and HSP in kids

### Wisecracks:

- 1. List three complications of circumcision.
- 2. Describe approaches to penile entrapment and tourniquet states.
- 3. Which side is a varicocele concerning and why?
- 4. Other than cystitis and pyelonephritis, list 6 other causes of pediatric dysuria
- 5. List 10 ddx hematuria in peds
- 6. List 5 false indicators of hematuria (review!)
- 7. List 6 causes of nephrotic syndrome in peds
- 8. List 5 complications of acute renal failure in children.
- 9. How does e. coli cause HUS? Which strain of e. coli is the concerning one?

### Core questions:

### [1] List a DDx for priapism and describe treatment.

Priapism is the engorgement of the dorsal corpora cavernosa, resulting in **dorsal penile** erection lasting more than 4 hours.

#### Three main types:

- Low flow (ischemic, painful!!!, "limb" threatening!)
  - Sickle cell disease (>50% of pts with SSD have at least one episode)
  - Malignancy (lymphoma; leukemia)
  - Medications
  - Cocaine / drugs of abuse
- High flow (non-ischemic, not painful, scary!)
  - Trauma, AVM, congenital disease
- Neurogenic (penis not threatened harbinger of spinal cord injury)

**Low-flow** (also called anoxic or veno-occlusive) priapism is the most common form and occurs secondary to decreased venous outflow.

Stuttering priapism is recurrent episodes of ischemic priapism, most lasting less than 4 hours. Episodes may increase in frequency and duration, with potential to develop into a major episode.

**High-flow** (also called arterial or congenital) priapism is usually painless and is typically associated with trauma to the cavernosal artery, congenital anomaly, or fistula, resulting in excessive inflow of arterial blood and corporal engorgement. <u>Oxygenation is maintained</u> and emergent intervention is not typically necessary.

Neurogenic priapism is not related to blood flow occlusion.

Other causes of priapism include immunosuppressive disorders, medications (SSRIs) drugs of abuse, and toxin exposure.

Complications include penile fibrosis, urinary retention, and impotence.



<u>Diagnosis:</u>

- In undifferentiated cases, a corporal cavernosal blood gas may differentiate the type of priapism, ischemic versus nonischemic,
- DDX: The differential diagnosis for priapism in children differs from that in adults. Penile erection from sexual arousal, erectile dysfunction medication, urethral foreign bodies, Peyronie's disease, spinal cord injury, and penile implants occur more commonly in adults than children.

#### MANAGEMENT:

#### LOW FLOW

\*\*\*draw an ABG off the dorsal penis: if pH <7.25, PaCO2> 60, Pa02 < 30, and dark coloured blood = LOW flow\*\*\*

- Akin to COMPARTMENT SYNDROME OF THE PENIS
  - hydration,
  - pain control,
  - relief of urinary obstruction,
  - and treatment of underlying conditions.

Local anesthesia by a dorsal nerve block or ring block should be performed prior to (1) aspiration, followed by (2) intracavernous injection (ICI) of sympathomimetic drugs (phenylephrine preferred).

#### Remember = 2 and 10 o'clock are safe zones!

## If no effect with 1 mL of dilute phenylephrine q 5 min after 1 hour, urology may consider placement of a surgical shunt.

To perform aspiration, place an 18-gauge angiocatheter (smaller in young children) percutaneously into the lateral aspect of the penile shaft entering the corpus cavernosum. Aspirate and evacuate blood from the corpora cavernosa. Next, irrigate with normal saline (NS) or in combination with an ICI of an  $\alpha$ -adrenergic sympathomimetic agent. Instill 1 mL of dilute phenylephrine

(100–500  $\mu$ g phenylephrine/mL of NS) into the corpus cavernosum every 3 to 5 minutes for  $\mu$ p to 1 hour.

If these measures fail to resolve the priapism, emergent urologic consultation should be obtained for **possible surgical shunt placement**.

Prolonged episodes (>48 hours) are associated with a high likelihood of erectile dysfunction, irrespective of clinical management. Treatment in nonischemic priapism is observation, because over two-thirds of cases resolve spontaneously. Refractory cases may require cavernosal artery embolization or arterial ligation, although those procedures have high complication rates. Stuttering priapism has no evidence-based preventive treatment.

#### **HIGH FLOW / NEUROGENIC**

Observation





#### • Refractory cases need cavernosal artery surgery

VARIANT	PENILE BLOOD APPEARANCE	PENILE ARTERIAL BLOOD GAS FINDINGS	COLOR DUPLEX ULTRASONOGRAPHY FINDINGS
Ischemic priapism	Corpus cavernosum testing—blood hypoxic, dark in color	Blood gases 1. • $PaO_2 < 30 \text{ mm Hg}$ 2. • $PaCO_2 > 60 \text{ mm Hg}$ 3. • pH < 7.25	Minimal or absent blood flow
Nonischemic priapism	Corpus cavernosum testing—blood is oxygenated, red	Blood gases $PaO_2 > 90 \text{ mm Hg}$ $PaCO_2 < 40 \text{ mm Hg}$ pH = 7.40  (similar to normal arterial blood)	Blood flow normal to high in velocity
Stuttering (recurrent) priapism	Corpus cavernosum testing—blood hypoxic, dark in color	Blood gases $PaO_2 < 30 \text{ mm Hg}$ $PaCO_2 > 60 \text{ mm Hg}$ pH < 7.25	Minimal or absent blood flow during acute priapism; normal blood flow otherwise

#### **TABLE173.1 Pathophysiologic Criteria of Priapism** (Rosen's 9<sup>th</sup> Ed.)

### [2] Describe the management of a paraphimosis.

#### **ALWAYS A PROBLEM**

• Proximal foreskin cannot be returned to its anatomic position covering the glans penis, resulting in distal venous congestion.

#### Paraphimosis

• can be caused by infection, masturbation, trauma, hair or clothing tourniquets, or failure to reduce the foreskin after a medical examination or procedure.

*Paraphimosis is a urologic emergency with potential for arterial compression, penile necrosis, gangrene, and/or autoamputation.* Hair tourniquet can mimic paraphimosis in the circumcised male.

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#### Management:

- 1. Pain control:
  - a. topically, parenterally, and/or with a local dorsal penile nerve block or ring block or procedural sedation.
- 2. Firm compression / methods to reduce edema
  - a. Firm ACE wrap on the penis to encourage venous outflow
  - b. Parent's hand holding constant pressure on the penis
  - c. placing the finger of a rubber glove filled with ice water over the glans and foreskin, sprinkling granulated sugar over the edematous tip, and injection of hyaluronidase into the edematous prepuce.

#### 3. Reduction

- a. Manual reduction using index fingers and thumbs (see figure in text)
- b. Needle fenestration
  - *i.* edematous foreskin may be punctured several times circumferentially with a 25-gauge needle to decrease edema further.
- c. Dorsal band traction
  - *i.* Dorsal band traction can be performed with Adson forceps applied directly to the band formed by the retracted foreskin and application of traction and countertraction to loosen the constriction.
- d. Dorsal slit procedure
  - i. An emergency dorsal slit should be performed when these measures fail to reduce the paraphimosis. Two straight hemostats are applied to crush the foreskin at the 12 o'clock position perpendicular to the corona. After 2 minutes, the prepuce between the hemostats is sharply incised, releasing the constricting band of tissue. The incisions are approximated with 4-0 absorbable sutures. Circumcision should be performed at a later date.

Follow up - with urology if child is able to urinate. If there are any signs of cellulitis/ischemia/necrosis or the child is unable to urinate they should be admitted.

### [3] Describe the management of a phimosis.

#### **USUALLY NOT A PROBLEM**

#### Inability to retract the foreskin in a patient where it was previously retractable.

Phimosis is a constriction of the foreskin that prevents retraction of the prepuce from the glans. Most cases are physiologic, represent normal development, and do not require intervention.

Four percent of newborns, 25% of 6-month-olds, 50% of 1-year-olds, 80% of 2-year-olds, and **90% of 4-year-old boys have fully retractable foreskins. Trauma, infections, chemical irritation, poor hygiene, congenital abnormality, or a complication of circumcision may contribute to the development of phimosis.** 

On history ask about:

• Pain, "ballooning" of the foreskin, urinary sprinkler/spray

#### Management:

- Without glans ischemia/infarction: (able to urinate, no signs of infection)
  - Gentle retraction and good hygiene are mainstays of management. Retraction of the prepuce should not be forced, because this can lead to



future adhesions and strictures. A 6- to 8-week course of topical corticosteroids (eg, 0.1% triamcinolone topical cream), applied to the outlet twice daily, may expedite the development of retractable foreskin.

- WITH ischemia/infarction:
  - dorsal slit procedure, circumcision, preputial plasty, or balloon dilation may be necessary. Obstructive uropathy can occur secondary to severe stenosis.

## [4] What is the pathophysiology of balanoposthitis? What is the most common bug in balanoposthitis?

Balanoposthitis, an inflammation that involves the glans and foreskin, occurs in approximately 5% of uncircumcised males.

Balanitis involves the glans penis only.

Balanoposthitis is usually secondary to poor hygiene, infection (bacterial and fungal), contact dermatitis, chemical irritation, or local trauma. Less commonly, a drug eruption, scabies infection, sexually transmitted infection (STI; eg, human papillomavirus [HPV], herpes), or nummular eczema may cause inflammation.

Infectious organisms are gram-negative and gram-positive, including group A beta hemolytic streptococci and, rarely, Neisseria gonorrhoeae and Chlamydia.

Canadia - is the most common fungal cause.

Bacterial causes: In a study of uncircumcised children, the most common organisms found by age were *E. coli* (ages 0 to 2 years), enterococci (ages 3 to 6 years), and *Staphylococcus aureus* and group A beta-hemolytic streptococci (ages 7 to 12 years) [17]. (Uptodate)

Physical examination reveals penile erythema, edema, and occasionally discharge. Streptococcal balanoposthitis is characterized by a fiery red surface and moist exudate under the prepuce. The patient may have a concomitant or recent streptococcal infection in other locations. Candidal balanoposthitis is associated with generalized erythema, fissuring, eroded papules, and a whitish discharge. Characteristic satellite lesions may be present.

## [5] How is balanoposthitis managed?

- 1. Avoid irritants
- 2. Warm sitz baths
  - a. May help to have child urinate in the bathtub
- 3. Good hygiene
- 4. Control inflammation/infection
  - a. Topical antibiotics
  - b. Topical antifungals
  - c. Topical corticosteroids for contact dermatitis
    - Topical corticosteroids (eg, hydrocortisone, 0.5%–1%) may help inflammation due to contact irritation. Candidal infections should be treated topically with antifungals (eg, clotrimazole, miconazole, nystatin). A blood glucose test should be considered in patients presenting with recurrent candidal balanoposthitis.



d. Systemic abx for severe infections / STIs

## [6] Describe the diagnosis and management of pediatric epididymitis and orchitis

Epididymitis:

- Etiology varies by age and presence of structural abnormalities
- Pre-adolescence:
  - Usually viral
  - Bacterial causes are rare and occur with structural abnormalities
- Adolescent:
  - Consider STI's as a cause
  - A urethral discharge may be present, particularly when the condition is secondary to an STI.

Patients present with a painful edematous scrotum and tenderness at the epididymis. Nausea, vomiting, fever, and lower abdominal, scrotal, or testicular pain may be present. Infants and young children may present with isolated fever.

Diagnosis:

- U/A +/- culture
- Capture first void and wait for results if suspicious of STI
- Consider your ddx:

Lack of pyuria does not rule out epididymitis; **up to 50% of patients have normal urinalysis.** 

Ultrasonography may be indicated in undifferentiated cases to exclude torsion. Increased vascular flow toward the side of the inflamed epididymis may be seen.

#### Management:

Scrotal elevation, ice packs to the scrotal area, and pain medications are useful to control pain and inflammation.

Sexually active:

- If urethral discharge is present, sexually active adolescents should be treated presumptively for both N. gonorrhoeae and C. trachomatis. Ceftriaxone (250 mg intramuscularly) and doxycycline (100 mg bid for 10 days) is preferred treatment. To increase compliance, azithromycin (1 g once) may be used instead of doxycycline, although there are little data on the use of azithromycin for chlamydial epididymitis. Follow-up for resolution of symptoms should occur if azithromycin used.
- In children, non-sexually acquired epididymitis without evidence of a urinary tract infection may be managed expectantly without antibiotics.
- Infants with or without positive findings on urinalysis and young children with positive urinalysis findings may be treated with cephalexin tid if a bacterial urinary tract infection is suspected.



#### Orchitis:

- Usually viral or bacterial testicular infection
- Sx:
  - Tenderness, edema, swelling, discolouration of the scrotum
  - If there is urethral discharge think of epididymo-orchitis
  - Causes:
    - Paramyxovirus is most common, associated with mumps. Other causes include Escherichia coli, Klebsiella pneumoniae, Pseudomonas aeruginosa, Staphylococcus or Streptococcus spp., Epstein-Barr virus, coxsackievirus, arbovirus, enterovirus, Brucella, granulomatous disease, and filariae.
- Treatment:
  - Treatment of viral orchitis is aimed at pain control (scrotal elevation, nonsteroidal antiinflammatory drugs [NSAIDs], and possibly narcotics).
     Bacterial:
    - Cover for gram-negative bacteria (eg, a third-generation cephalosporin).
      - urologist should be urgently consulted for a scrotal or testicular abscess.

## [7] Describe the diagnosis and management of testicular torsion

Key points:

- Can occur at any age most commonly 12-18 yrs
- Time sensitive dx! 10% salvage rate at 24 hrs!

Diagnosis:

- Hx: acute constant scrotal pain, swelling, high riding testicle, with nausea and vomiting.
  - acute scrotal pain and swelling, an elevated testicle and, typically, absence of the cremasteric reflex. This reflex can be demonstrated by lightly stroking the skin of the inner thigh downward from the hip toward the knee.
    - The cremaster muscle on the ipsilateral side rapidly contracts and elevates the testicle.Although absent in the vast majority of cases, the presence of the cremasteric reflex does not preclude testicular torsion.
  - **Abnormal (high-riding and transverse) epididymal and testicular position may also be noted**, with left-sided torsions slightly more common than right.
  - Pain is typically constant, and the patient may have a history of a similar prior episode. Up to 90% of patients have associated nausea and vomiting.
  - Cryptorchidism (one or both testes are undescended) increases the risk of torsion and may present with abdominal or inguinal pain. Familial history and recent trauma may increase the risk of torsion
- Imaging:
  - Emergent doppler ultrasonography of both testicles and spermatic cords.
    - 85-95% sensitivity

#### Management:



- Immediate surgery ideally no more than 12 hrs from onset of symptoms
   Surgery should not be delayed while waiting for ultrasound
  - Followed by elective orchiopexy of the contralateral side to avoid recurrence.
- Bedside manual detorsion may be performed with analgesia if operative repair is delayed. The testicle is rotated in an open book fashion as viewed from below, from medial to lateral, until detorsion is complete.
  - Torsion may cause ischemia at 180 degrees of rotation but typically occurs with 360 degrees of twisting. Up to one-third of patients may torse laterally; in these cases, manual reduction worsens torsion and its symptoms, and the procedure should be halted.
- In causes where the testicle is no longer viable an elective orchiectomy may be performed

Note: Both the appendix of the testis and appendix of the epididymis can torse, although the former is much more common. Patients with appendiceal torsion present with moderate pain of sudden onset localized to the involved hemiscrotum. The pathognomonic blue dot sign (<3 mm bluish hue in the upper lateral hemiscrotum—a cyanotic appendage) is present in less than 25% of cases. <u>Color Doppler ultrasound should be</u> performed when the diagnosis is uncertain and will reveal normal or increased flow to the affected testicle.

Conservative therapy with analgesics and scrotal support is indicated; the involved appendage undergoes autoamputation within 1 week, accompanied by resolution of symptoms.

Testicular salvage rates are time-dependent. Nearly 100% of testes can be saved if detorsed within 4 hours of symptom onset. This decreases to less than 10% if there is more than a 24-hour delay to treatment.

If the tunica completely covers the testis and attaches higher up on the spermatic cord (bell clapper deformity), proper testicular fixation does not occur, and there is a predisposition to torsion. See Figure 173.9.

A hydrocele is a collection of fluid that accumulates in the tunica vaginalis. **Communicating** hydroceles have an open tract between the peritoneum and scrotum. Infants - likely primary hydrocele. Older kids and teens - look for a secondary cause.

In older children and adolescents, they may occur secondary to epididymitis, orchitis, testicular torsion, appendix testis or epididymis torsion, trauma, or tumor. Examination with transillumination reveals enlargement of the scrotum. Color flow Doppler ultrasonography may be necessary to determine the cause of the hydrocele and exclude an acute pathologic process in patients with acute symptoms of pain.

Girls (esp. Infants that are premature) are more likely to have inguinal hernias that become incarcerated. Concomitant ovarian torsion may occur with inguinal hernias and should be suspected with continued irritability after successful hernia reduction.

## [8] List common bacteria in peds UTI and describe treatment <2 months, 2M - 2 Yrs, > 2 Yrs.

Neonatal boys are more susceptible to UTI's than girls but, beyond that, females are at higher risk.



< 2 months 2 months - 2	E coli Klebsiella Other pathogens E. coli!	<ul> <li>Infants younger than 2 months are more at risk for sepsis.</li> <li>Most require admission and broad spectrum IV abx (ampicillin and cefotaxime)</li> <li>Rosen's suggests that some 29-60 day olds may be managed as outpatients if well - but this doesn't seem common in our practice</li> <li>The CPS statement doesn't comment on this specifically</li> <li>PO abx for 7-10 days.</li> </ul>	***up to 50% of UTI's < 3 months are associated with bacteremia! These kids should be considered for a FSWU
2 months - 2 yrs	E. COII! Other pathogens include Enterobacter, Proteus, Morganella, Serratia, and Salmonella spp.	<ul> <li>PO abx for 7-10 days.</li> <li>PO options: <ul> <li>Cefixime (drug of choice for first febrile UTI)</li> <li>Amoxicillin</li> <li>Clavulin</li> <li>Cephalexin</li> </ul> </li> <li>See list below for more.</li> <li>IV options: <ul> <li>Ampicillin and ceftriaxone</li> </ul> </li> <li>UTIs in this age group are considered to be upper tract disease processes, a 7- to 10-day course is recommended</li> </ul>	option for well appearing children who are able to receive and tolerate every dose and have good follow up (tough in children < 6 months of age) No macrobid Need an outpatient bladder, ureters, kidney ultrasound to exclude structural abnormalities
> 2 yrs	E coli, and others (see above)	<ul> <li>3-day course of antibiotics, such as cephalexin or amoxicillin-clavulanate for cystitis.</li> <li>7-14 days for pyelonephritis.</li> <li>Avoid fluoroquinolones</li> </ul>	



Amoxicillin	50 mg/kg/day (divided in three doses)
Amoxicillin/clavula nate	(7:1 formulation) 40 mg/kg/day (divided in three doses)
Co-trimoxazole	8 mg/kg/day of the trimethoprim component, divided in two doses (0.5 mL/kg/dose)
Cefixime	8 mg/kg/day (given as a single dose)
Cefprozil	30 mg/kg/day (divided in two doses)
Cephalexin	50 mg/kg/day (divided in four doses)
Ciprofloxacin*	30 mg/g/day (divided in two doses) (only for post-pubertal kids)

#### Check out for more:

- <u>https://pedscases.com/cps-statement-urinary-tract-infections</u>
- https://www.cps.ca/en/documents/position/urinary-tract-infections-in-children

#### **Complicated UTI (CPS Statement)**

- Hemodynamically unstable
- Elevated Creatinine
- Bladder or abdominal mass
- Poor Urine Flow
- Not improving clinically within 24h of starting treatment
- Fever after 48h of antibiotics.
- **Treatment** should include IV antibiotics and a renal and bladder ultrasound should be performed to look for obstruction or abscess.



## [9] List 6 ddx renal mass – how should these be imaged first?

Three things: hydronephrosis; cystic masses; solid masses

- cystic lesions, such as those of polycystic kidney disease, or benign cystic nephroma
- severe hydronephrosis resulting from obstruction or severe reflux.
- Solid masses:
  - Wilms' tumor, neuroblastoma, renal cell carcinoma, mesoblastic nephroma, and cystic neuromas, malignant rhabdoid tumour

Start with ultrasound!

### [10] Give a differential diagnosis of proteinuria in children.

First determine (using dipstick or U/A) whether it's mild or moderate.

Patients with moderate proteinuria ( $\geq$ 3+; equivalent to  $\geq$ 300 mg/dL) should have additional testing, including the total serum protein, albumin, electrolyte, BUN, and creatinine levels, and urine culture.

#### Also, try to add on a random urine protein-to-creatinine ratio.

A urine Pr/Cr more than 3.0 mg/dL correlates to nephrotic syndrome. Children with elevated urine Pr/Cr levels should be referred to a nephrologist for a 24-hour urine collection for protein.

Consider adding on an antistreptolysin O (ASO) titer can identify a previous streptococcal infection as the cause of the proteinuria. (PSGN)

Causes outside of trace/mild proteinuria: think glomerular vs. tubular.

Glomerular causes include nephrotic syndrome, glomerulonephritis, and posttransplantation rejection. Transient causes of altered glomerular function include exercise, extreme cold or heat, fever, seizures, and stress.

Tubular causes of proteinuria include heavy metal poisoning, urinary tract infections, and diabetes, as well as an asymptomatic tubular proteinuria.

## [11] What is the most common cause of proteinuria in children?

Usually we see trace or mild proteinuria (< 100 mg/dL):

Trace to mild proteinuria (1+ to 2+) is a common laboratory finding in young children and can represent benign conditions, such as <u>exercise or mild dehydration or urinary</u> <u>tract inflammation/infection (cystitis)</u>

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Mild proteinuria ( $\leq$ 2+; equivalent to  $\leq$ 100 mg/dL) requires no further investigation. Evidence of a UTI, such as WBCs or a positive leukocyte esterase or nitrite level, should be treated.

Causes outside of trace/mild proteinuria: think glomerular vs. tubular.

#### The #1 cause would be post-infectious (IgA nephropathy or PSGN)

Glomerular causes include nephrotic syndrome, glomerulonephritis, and posttransplantation rejection. Transient causes of altered glomerular function include exercise, extreme cold or heat, fever, seizures, and stress.

Tubular causes of proteinuria include heavy metal poisoning, urinary tract infections, and diabetes, as well as an asymptomatic tubular proteinuria.

## [12] Provide a differential diagnosis of acute renal failure in children. What is the most common cause?

Most common: prerenal AKI secondary to volume depletion.

#### Box 173.3: Cause of Acute Renal Failure in Children

#### Prerenal

- 1. Decreased intravascular volume or dehydration
  - Burns or hemorrhage
  - Third spacing
  - Sepsis
- 2. Decreased cardiac output
  - Cardiac shock

3. Decreased renal artery blood flow

#### Intrarenal

#### 1. Glomerular disease

- Poststreptococcal and other glomerulonephritis
- Pyelonephritis

#### 2. Systemic causes

- Hemolytic-uremic syndrome
- Henoch-Schönlein purpura or other vasculitides
- Systemic lupus erythematosus
- Sepsis or other causes of prolonged decreased perfusion

#### 3. Toxins

- Heavy metal poisonings, such as lead and gold
- Myoglobin or hemoglobin deposits
- Antibiotics such as aminoglycosides
- Anticonvulsants such as phenytoin
- Radiocontrast dyes

#### Postrenal

- 1. Obstructive lesions
- 2. Nephrolithiasis or tumor
- 3. Posturethral valves
- 4. Intra-abdominal tumor obstructing urinary flow
- 5. Infection
- 6. Renal vein thrombosis



## [13] Describe the clinical presentation of glomerulonephritis in a child.

Here we are classically thinking about "PSGN"

Poststreptococcal glomerulonephritis (PSGN) is one of the most common glomerulonephritides. PSGN is a sequelae of streptococcal pharyngitis and, less commonly, streptococcal skin infections.

**PSGN is** believed to result from the **deposition of circulating immune complexes in the kidney.** How those immune complexes develop is not completely understood. These immune complexes result in decreased glomerular filtration, allowing proteins to flow freely into the urine.

But taking a step back and considering the host of other causes of glomerulonephritis - think about the GU system, and then don't forget to consider systemic diseases that could lead to GN (e.g. lupus rash, Goodpasture's presenting with chest pain and hemoptysis)

Urinalysis will show significant blood and protein, with RBC casts in 60% of cases. Other findings may include pyuria, with granular or hyaline casts.

## [14] Define hypertension in a child.

systolic or diastolic blood pressure higher than 2 standard deviations (SDs) above the mean for age and gender

	BLOOD PRESSURE: UPPER LIMIT (mm Hg)		
AGE (yr)	Systolic	Diastolic	
0–2	110	65	
3–6	120	70	
7–10	130	75	
1–15	140	80	

 Table 173.2: Blood Pressure Limits in Children

### [15] List causes of hypertension in a child.

Big list!

- Primary (dx of exclusion in kids!)
- Secondary
  - Think through broad categories:
    - Cardiac
      - Vascular
    - Endocrine
    - Renal



#### Drug related

Let's go through a few for each.

#### Box 173.4: Causes of Hypertension in Children

#### Primary

1. Essential hypertension

#### Secondary

#### Renal

- 1. Glomerulonephritis
- 2. Henoch-Schönlein purpura
- 3. Pyelonephritis
- 4. Obstruction or reflux
- 5. Polycystic kidney disease
- 6. Diabetic nephropathy
- 7. Trauma
- 8. Renal transplant or hemodialysis
- 9. Tuberous sclerosis
- 10. Systemic lupus nephritis

#### Endocrine

- 1. Pheochromocytoma
- 2. Cushing's syndrome
- 3. Congenital adrenal hyperplasia
- 4. Corticosteroid treatment
- 5. Hyperthyroidism
- 6. Neuroblastoma
- 7. Ovarian tumor

#### Cardiac

- 1. Congestive heart failure
- 2. Coarctation of the aorta

#### Vascular

- 1. Hemolytic-uremic syndrome
- 2. Kawasaki syndrome
- 3. Renal artery thrombosis or stenosis

#### Neurologic

- 1. Central nervous system tumor or infection
- 2. Central nervous system trauma or abuse
- 3. Increased intracranial pressure
- 4. Guillain-Barré syndrome

#### Neoplastic

- 1. Neuroblastoma
- 2. Wilms' tumor
- 3. Pheochromocytoma
- 4. Adrenal carcinoma

#### Drugs

- 1. Corticosteroids
- 2. Cocaine
- 3. Sympathomimetics
- 4. Oral contraceptives
- 5. Phencyclidine
- 6. Beta-blocker or clonidine withdrawal

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7. Lead, mercury

#### Others

- 1. latrogenic fluid overload
- 2. Volume overload from end-stage renal disease

## [16] Llist 2 medical treatments for acute HTN crisis in children

Rare!

Kids < 10 yrs: BP > 160/105 - hunt for the signs of end organ damage! Kids > 10 yrs: BP > 170/110 - go looking!

Big list to choose from; here's two. (you may have other drugs easily accessible at your hospital)

Goal is a 10-20% reduction in hours.

- Labetalol
- Hydralazine

Children with hypertensive emergencies have evidence of end organ damage, including acute neurologic changes or encephalopathy, pulmonary edema, myocardial ischemia, and proteinuria. The electrocardiogram may show signs of ischemia or ventricular hypertrophy. Chest radiography may reveal cardiomegaly or pulmonary edema.

Isolated headache is not considered a sign of hypertensive emergency.

See Table 173.3 For additional info on treatment of hypertensive emergency.

## [17] What are the two types of HUS? What are the typical presenting features?

- 1. Primary atypical HUS
- 2. Secondary HUS

Primary results from complement dysregulation.

Secondary is a post-infectious/drug/idiopathic phenomenon.

(eg, STEC, Shigella organisms, S. pneumoniae, Aeromonas, or HIV), drugs (eg, chemotherapeutic or transplant anti-rejection drugs), or other idiopathic causes (eg, pregnancy, lupus). UTIs have also been implicated.

#### Presentation:

Secondary HUS due to STEC presents with watery diarrhea, crampy abdominal pain, and occasionally fever. Within days of symptom onset, patients experience increasing abdominal pain, with 50% to 85% developing bloody stools.

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Patients may also develop toxic megacolon, ischemic colitis, intussusception, perforation, or delayed colonic stricture.

Patients with a pneumococcal cause for the HUS typically present with pneumonia or, less commonly, meningitis, bacteremia, sinusitis, or otitis media.

Up to 10% of patients will experience the triad of sudden onset hemolytic anemia, thrombocytopenia, and acute renal insufficiency, with possible progression to renal failure. Up to 60% of diarrhea-associated HUS cases require dialysis, and death or end-stage renal disease was found to occur in 12%.

Remember this is a hemolytic disease: The peripheral blood smear shows microangiopathic changes such as schistocytes, teardrop cells, helmet cells, microspherocytes, and burr cells.

## [18] Describe the management of HUS and HSP in kids

HUS	HSP	
a microangiopathic hemolytic anemia, is one of the most common causes of acute kidney injury in children. HUS is rare after 5 years of age,	Hypersensitivity small vessel vasculitis affecting the joints, GI system and skin. Usually post URTI or group a strep infection (can be provoked by any virus) classic HSP rash is usually diagnostic. Watch for renal or abdominal complications.	
<ul> <li>These folks can become sick fast! <ul> <li>Aggressive supportive care</li> <li>Correct electrolytes</li> <li>Treat pancreatic failure</li> <li>Treat seizures</li> <li>Treat hypertension</li> </ul> </li> <li>No role for antibiotics (enhance release of verotoxin)</li> <li>Except: Pneumococcal-associated HUS often presents with pneumonia and should be treated</li> <li>Early consideration of dialysis</li> <li>PRBCs if Hgb &lt; 60</li> <li>?consideration of plasmapheresis if presenting with stroke</li> <li>?eculizumab if primary HUS</li> </ul>	<ul> <li>Controversial</li> <li>NSAIDs for arthralgias (assuming no renal involvement)</li> <li>Corticosteroids may help abdominal pain resolve; but do NOT prevent abdominal or renal complications</li> <li>IVIG if severe renal impairment</li> </ul>	



## Wisecracks:

## [1] List three complications of circumcision.

Circumcision prevents phimosis, paraphimosis, recurrent balanoposthitis, and decreases urinary tract infections, sexually transmitted disease transmission (including human immunodeficiency virus [HIV]), and penile cancer. An American Academy of Pediatrics task force has concluded that the benefits of circumcision outweigh the risks.

- Infection
  - Local, systemic
  - UTI
- Bleeding
  - Minor
  - Major bleeding may raise suspicion for a blood dyscrasia
- Pain
- Urinary problems:
  - Stricture formation
  - Urinary retention
- Post-circumcision phimosis

## [2] Describe approaches to penile entrapment and tourniquet states.

- Depend on the object
  - Rings:
    - Mineral oil; string wrap technique; ring cutters
  - Zippers:
    - Mineral oil; cutting median bar of zipper

### [3] Which side is a varicocele concerning and why?

A varicocele is a collection of venous varicosities of the spermatic veins in the scrotum caused by incomplete drainage of the pampiniform plexus.

Varicoceles are more pronounced when upright. Varicoceles are described as a "bag of worms" in appearance and on palpation.

- Normally occurs on the left side
- Any RIGHT sided varicocele should raise suspicion for neoplasm and patient should be considered for imaging to investigate for retroperitoneal neoplasm (U/S, CT, MRI)

Concerning features for varicoceles:

- 1. Sudden onset
- 2. Right sided
- 3. Do not decrease in size when supine.



# [4] Other than cystitis and pyelonephritis, list 6 other causes of pediatric dysuria

- Stones kidney
- Strictures labial adhesions
- STI's balanitis or vaginitis
- Stuff retained foreign bodies
- Stuck labial adhesions, phimosis
- Sad situation abuse
- Self stimulation
- Sticky tape pin worms
  - **Or Scratching while sleeping** (just treat it empirically in real life if the dx is highly suspected).

#### See Box 173.1: Cause of Dysuria in Children

#### Infection

- 1. Urinary tract infection, including cystitis and pyelonephritis
- 2. Vaginitis resulting from *Gardnerella*, *Trichomonas*, *Candida*, or sexually transmitted organisms
- 3. Pinworms
- 4. Balanitis

#### Irritation

- 1. Bubble bath, new soaps, or douches
- 2. Vaginal foreign body, such as retained toilet paper

#### Trauma

- 1. Sexual or physical abuse
- 2. Straddle injury (unintentional)
- 3. Self-stimulation or masturbation

#### Other

- 1. Labial adhesions
- 2. Renal stones or hypercalciuria

## [5] List 10 ddx hematuria in peds

Red blood cells (RBCs) can enter the urinary tract from inflammation, infection, trauma, or anatomic abnormalities, anywhere from the glomerulus to the urethra. Microscopic hematuria is defined as more than 5 RBCs/mm3; macroscopic or gross hematuria is the presence of blood or clots visible to the naked eye.

#### From glomerulus to ureters! Think structurally or think:

- Renal
- Extra-renal (including mimickers)
- Systemic

## CRECK The rest of the rest of

#### Box 173.2: Cause of Hematuria in Children

#### Extrarenal

- 1. Trauma
- 2. Meatal stenosis or posterior urethral valves
- 3. Exercise
- 4. Menstruation or rectal bleeding
- 5. Foreign bodies
- 6. Cystitis, urethritis, or epididymitis

#### Intrarenal

- 1. Pyelonephritis
- 2. Renal or bladder stones or tumors
- 3. Poststreptococcal or idiopathic glomerulonephritis
- 4. Acute interstitial nephritis
- 5. Acute tubular necrosis
- 6. Basement membrane glomerular disease
- 7. Renal vein or artery thrombosis
- 8. Recurrent familial hematuria
- 9. Polycystic kidney disease

#### Systemic

- 1. Alport syndrome nephritis
- 2. Henoch-Schönlein purpura
- 3. Systemic lupus erythematosus
- 4. Hemolytic-uremic syndrome
- 5. Infectious mononucleosis
- 6. Sickle cell disease or other hemoglobinopathies
- 7. Bacterial endocarditis or artificial cardiac valves
- 8. Bleeding disorders, warfarin, or aspirin
- 9. Medications such as amitriptyline or chlorpromazine, radiocontrast dyes
- 10. Munchausen syndrome or factitious

look for signs of renal disease (eg, glomerulonephritis), including hypertension, facial edema, rales, and cardiac murmurs.

### [6] List 5 false indicators of hematuria (review!)

Urine with lysed RBCs or myoglobin from muscle breakdown tests positive for hemoglobin, yet will not contain RBCs.

Certain drugs or foods, such as phenothiazines, ibuprofen, beets, and blueberries, can cause reddish urine.

In neonates, urate crystals can cause a benign red-tingedurine in the diaper, also termed brick dust urine.



## [7] List 6 causes of nephrotic syndrome in peds

Nephrotic syndromes are kidney diseases that result in significant proteinuria, hypoproteinemia, and edema.

#### Although hypoalbu-

minemia defines the diagnosis of nephrotic syndrome, levels of other important proteins such as immunoglobulins can also be reduced by the disease.

#### Usually broken down by two age groups:

- < 5 yrs likely primary nephrotic syndrome due to minimal change disease
- > 5 yrs usually due to a secondary/systemic disease
  - PSGN
  - Lupus

#### Here's that list of causes:

- Primary proteinurea due to minimal change disease
  - primary nephrotic syndrome (no gross hematuria or elevated creatinine level, normal complement levels, and no evidence of extrarenal causes such as a malar rash)
- Secondary causes:
  - IgA nephropathy
  - PSGN
  - HUS
  - **HSP**
  - Lupus
  - Goodpasture's disease
  - Wegener's granulomatosis
  - Alport syndrome

#### What is "nephrotic range" proteinuria?

• Nephrotic-range proteinuria is a daily excretion of more than 3.5 g of protein/1.73 m2 or more than 50 mg/kg, corresponding to 3+ or 4+ on the urine dipstick.

### [8] List 5 complications of acute renal failure in children.

- edema
- ascites,
- significant hypertension (>99th percentile for age and height) resulting from glomerulonephritis,
- marked impairment of renal function (>50% over normal) should be hospitalized.
- pulmonary edema
- cardiac arrhythmias

Let's go through the life threats again: severe hyperkalemia, hyponatremia, pulmonary edema or fluid overload, hypertensive encephalopathy, septic shock from renal obstruction and infection, and seizures from metabolic abnormalities or encephalopathy.



## [9] How does e. coli cause HUS? Which strain of e. coli is the concerning one?

The BIG concern with HUS is acute renal failure!

Renal compromise is due to renal vascular endothelial injury, often induced by viral or bacterial agents.

RBCs are injured within narrowed blood vessels, resulting in a microangiopathic hemolytic anemia. Platelets, complement, and fibrin are deposited in the glomerular lumen, resulting in thrombocytopenia, a decrease in the glomerular filtration rate, and renal failure.

Most common cause is Shiga toxin (verotoxin)–producing E. coli (STEC), specifically serotype O157:H7.

Spread fecal orally or from contaminated foods.