

Pediatric Urological Emergencies for the General Urologist

Learning Objective: At the conclusion of this continuing medical education activity, the participant will be able to diagnose and appropriately manage the urological conditions common to pediatric patients that warrant prompt management.

This AUA Update aligns with the American Board of Urology Module on Core/General Urology. Additional information on this topic can be found in the AUA Core Curriculum sections on Consults & Emergencies and Pediatric Urology.

Cyrus M. Adams, MD, MS¹ and Richard C. Rink, MD, FAAP, FACS¹

¹Riley Children's Hospital at IU Health, Indianapolis, Indiana

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INTRODUCTION

As all surgeons have learned during the COVID-19 pandemic, emergency cases are those that threaten life, limb or organ. Fortunately, these are infrequent in pediatric urology. In this Update, we describe the urological emergencies that occur in pediatric patients which general urologists may encounter and must be prepared to appropriately manage. We review the urological problems seen in infancy and childhood with a description of the typical presenting signs or symptoms, the differential diagnoses, the proper evaluation and diagnostics, and the recommended course of management.

ACUTE SCROTUM

Scrotal swelling is one of the most common acute urological problems seen in children. **The possibility of testicular torsion must be immediately considered, as this is a condition that requires emergent surgery and is time-sensitive.** The cumulative incidence of testicular torsion was historically reported as 1 per 4,000 males by age 25 years.¹ However, recent national database work showed approximately 1 in 1,500 males will require surgery for testicular torsion by age 18 years with an orchiectomy rate of 42%.² Testicular torsion presents in a bimodal age presentation with peaks in the first year of life and early adolescence. Extravaginal torsion occurs during testicular descent in the perinatal period, while intravaginal torsion peaks near puberty, although it may be seen at any age. The estimated incidence is 6.1 per 100,000 births,³ and it rarely may occur bilaterally either concurrently or metachronously. Other processes may mimic testicular torsion and include torsion of a testicular or epididymal appendage, epididymitis, incarcerated hernia, trauma, tumor and Henoch-Schönlein purpura. Other potential etiologies include disorders resulting from a patent processus vaginalis, including trapped meconium or a hematoma from intra-abdominal hemorrhage.

The history including the age of the child, the timing of presentation relative to the onset of symptoms and physical examination are key to making an expedient diagnosis. However, the diagnosis of testicular torsion is not always straightforward. A scrotal ultrasound with Doppler should be emergently completed to evaluate the acute scrotum.

The management of extravaginal torsion remains controversial as the rate of salvage of the affected testis is quite poor. However, the risk of a rare but serious bilateral asynchronous perinatal torsion warrants a discussion with the parents regarding the relative risks of exploration and anesthesia in the neonatal period versus observation.⁴ Therefore, we recommend a consultation with a pediatric urologist. Some pediatric urologists recommend no surgical intervention for extravaginal torsion, but our bias is to recommend immediate surgical exploration in an otherwise healthy child primarily for fixation of the contralateral testicle to eliminate the risk of metachronous torsion. However, all would agree it is important that infants who have normal testes at birth who subsequently develop an acute scrotum secondary to torsion undergo emergent exploration.

Intravaginal torsion is believed to be due to excess mobility of the testis from a “bell-clapper deformity” due to abnormal

fixation of the testis within the tunica vaginalis. This allows the spermatic cord to potentially twist on itself, compromising blood flow to the testis. This is thought to be most common during periods of testicular growth, such as puberty, explaining the peak during this time period. Cryptorchid testes are at increased risk for torsion and are difficult to assess because of the high position. Patients typically present with acute onset of pain associated with nausea and vomiting. They typically have extreme tenderness to palpation. The testis may be noted on physical examination to have a transverse lie and ride higher in the scrotum than the contralateral testis. Over time, the hemiscrotum often becomes edematous, erythematous and may develop a reactive hydrocele, which in addition to the significant pain with palpation may make examination quite difficult. A cremasteric reflex may be absent in cases of torsion. One may occasionally elicit a history consistent with prior similar episodes that resolved spontaneously, referred to as intermittent torsion, which is associated with subsequent testicular torsion. With acute torsion, testicular salvage is directly related to duration of symptoms before exploration, with excellent salvage expected with less than 6 hours of symptoms but progressively worse thereafter. Beyond 48 hours, the salvage rate is poor, and therefore immediate exploration under optimal anesthetic conditions is favored.

Early presentation, correct diagnosis and prompt treatment are critical for testicular salvage as the viability is inversely related to duration of torsion. It is important to note that not all boys who have an acute scrotum require an ultrasound. **A clear history and physical examination consistent with testicular torsion is an immediate indication for surgery without the delay of an ultrasound.** Doppler ultrasound is indicated in cases of an equivocal history or physical examination; however, the results are technician dependent. This yields a 1% false-positive rate, 88.9% sensitivity and 98.8% specificity in evaluation of an equivocal acute scrotum.⁵

Detorsion within 4 to 8 hours has generally been accepted as the optimal interval to salvage the affected testis. The risk of orchiectomy was approximately 5%, 20%, 40%, 60%, 80% and 90% at 0 to 6, 7 to 12, 13 to 18, 19 to 24, more than 24, and more than 48 hours after onset of pain, respectively.⁶ **Therefore, once the diagnosis is made, a urologist must perform immediate scrotal exploration without delay. Transfer from a hospital with an available urologist to a tertiary center with a pediatric urologist is unnecessary and is likely to delay surgical intervention, thereby increasing the risk of orchiectomy.**

Surgical exploration can be done through a transverse hemiscrotal or midline raphe incision and should first address the affected side. The testis is delivered, the tunica vaginalis is opened and the testis is untwisted, wrapped in warm soaked gauze and observed while addressing the contralateral side. The contralateral testis is then fixed with nonabsorbable suture to reduce the risk of metachronous torsion. The affected testis is re-examined for potential viability, which is a subjective decision. A Doppler flow probe or incision of the tunica albuginea with assessment of bleeding may document intratesticular flow after detorsion; however, the reliability of each technique lacks validation. If the testis is to be retained, it is fixed to the dartos with nonabsorbable suture. Some have suggested that a compartment syndrome contributes to testicular injury and therefore propose that a patch of vascularized tunica vaginalis be placed in the tunica albuginea defect to maintain lower

intraparenchymal pressure, thereby reducing the likelihood of ongoing ischemia.⁷ This may be considered, although the long-term success of this technique is unclear.

Torsion of an epididymal or testicular appendage typically presents similarly to testicular torsion. However, this is typically not associated with pain as severe as testicular torsion or with systemic symptoms such as nausea and vomiting. On physical examination, the urologist may be able to localize the point of tenderness and/or observe a “blue dot” through the scrotal skin.⁸ Doppler scrotal ultrasound may demonstrate an abnormal or enlarged appendage but frequently reveals hyperperfusion of the epididymis, which may resemble epididymitis.⁹ Torsion of an appendage is typically self-limited and can be managed with reassurance, rest and nonsteroidal anti-inflammatory medications or other analgesics. Providers should inform patients and parents that symptoms will typically resolve over 3 to 5 days. However, rarely, protracted pain occurs. In this situation, exploration and excision of the appendage may be considered on an outpatient basis.

Epididymitis is another common etiology for an acute scrotum in children. The pain typically has a more insidious onset and may be accompanied by a history of fever or new lower urinary tracts symptoms along with pyuria or bacteriuria on urinalysis. This may be seen in both pre- and post-pubertal boys and may mimic testicular torsion. Physical examination may vary from localized epididymal enlargement and tenderness to a massively swollen and erythematous hemiscrotum. However, the cremasteric reflex should be intact. Doppler ultrasound typically reveals an enlargement and increased blood flow to the epididymis in comparison with the contralateral testicle. This hyperemia may also extend into the testis in the case of epididymo-orchitis. The management goal is to relieve inflammation with the use of ice packs, nonsteroidal anti-inflammatory agents, scrotal elevation and rest, and to treat any associated infection in the presence of pyuria or bacteriuria. Testing and treatment for sexually transmitted diseases are warranted in sexually active adolescents. Children found to have epididymitis with a positive urine culture not associated with instrumentation require further diagnostic evaluation with renal-bladder ultrasound and voiding cystourethrography after the inflammation has resolved. There is generally no indication for these further studies when urine cultures are negative.

An incarcerated inguinal hernia may present as a tender or non-tender scrotal swelling that may require emergent management. It is differentiated from the previously mentioned entities by the persistent mass and induration which extends to the inguinal area on examination.¹⁰ The provider may be able to illicit a history of intermittent scrotal swelling suggestive of a patent processus vaginalis. Ultrasonography may demonstrate free fluid in the hernia sac or within the herniated bowel loop, thickening of the incarcerated bowel and dilated bowel loops in the abdomen, while the testicles and epididymis will typically appear normal in size and echotexture.¹¹ General surgery consultation may be warranted. However, manual reduction can usually be performed with sedation, allowing for delayed surgical exploration under optimal anesthetic conditions. Testicular injury can occur from torsion due to the effect of the incarcerated hernia or due to ischemia from cord compression, thus further mandating emergent reduction of the hernia or surgical exploration if reduction is not possible.¹⁰

Scrotal trauma is another relatively common cause of acute presentation and is typically a result of a direct blow or straddle injury in children. Trauma may also lead to torsion, although it more commonly results in a scrotal hematoma and pain limiting physical examination. Again, in these cases Doppler ultrasound can be useful when correlated with the history and examination. Genital trauma in children should be managed as it is in adults.¹² The most specific findings for testicular rupture on ultrasonography are loss of testicular contour and heterogeneous echotexture of parenchyma, which warrant surgical exploration to prevent complications such as ischemic atrophy or infection. Repair of a ruptured testis by debriding non-viable tissue and closing the tunica albuginea is preferred over orchiectomy when possible and may include the use of a tunica vaginalis graft when the tunica albuginea cannot be closed primarily.

Henoch-Schönlein purpura, another mimic of testicular torsion, is a systemic vasculitis affecting the skin, joints, gastrointestinal tract and kidneys, which may also impact the scrotum resulting in acute tenderness, edema or erythema; testicular hematoma, torsion or infarction; or cord thrombosis or epididymitis.¹³ The characteristic purpuric rash of the buttocks, perineum and lower limbs is usually present but may not precede the scrotal findings. It also is usually bilateral in contrast to torsion. Urgent scrotal exploration is only indicated if clinical findings suggest concomitant testicular torsion.

A scrotal hematoma from adrenal hemorrhage may be present in newborns with a patent processus vaginalis, prompting urological consultation for an acute scrotum.¹⁴ Adrenal hemorrhage may range from a small incidental finding without clinical significance to a life-threatening hemorrhage, and a patient should be monitored accordingly. An alternative diagnosis is neuroblastoma, and therefore further evaluation with magnetic resonance imaging may be required. Serial ultrasonography should be done every 1 to 2 weeks to demonstrate decreasing size of an adrenal hemorrhage. A resolving adrenal hemorrhage will display peripheral eggshell calcifications as opposed to the stippled appearance of neonatal neuroblastoma.¹⁵

HEMATURIA

Hematuria in children may result from trauma or from interstitial, glomerular, vascular or urothelial sources. In children, hematuria is more frequently associated with medical problems than with surgical problems. In the setting of trauma, a child should be managed in accordance with the American Urological Association Urotrauma guidelines.¹² Most cases of hematuria in children do not require urgent urological intervention unless the hematuria is so severe as to result in clots or urinary retention, at which point the child should be temporized until transfer to a center with an available pediatric urologist. If the child has associated proteinuria, hypertension, edema or renal insufficiency, then medical etiologies should be investigated. When symptoms of a urinary tract infection are present, urinalysis and culture and appropriate antibiotic treatment are indicated, with outpatient referral to pediatric urology for further evaluation with a renal-bladder ultrasound and voiding cystourethrography if indicated after resolution of the infection. Hematuria is more commonly seen with viral cystitis than with bacterial infection. Just as in adults, if renal colic is present, abdominal imaging should be obtained to rule out a urinary tract stone. In an asymptomatic child without these signs or

symptoms, a urine culture, spot urinary calcium/creatinine ratio or 24-hour urine for calcium, and a renal ultrasound should be considered, which may be done on an outpatient basis.

If any abnormalities are discovered during this evaluation, an elective referral to a pediatric urologist is warranted.

Hematuria in a newborn is fortunately rare, but when noted it is mandatory to evaluate emergently. **Both renal vein and renal artery thrombosis may present with hematuria, with both disorders threatening the viability of the kidney and potentially even the child.** Renal vein thrombosis is much more common, accounting for 20% of gross hematuria in this age group. This condition may present with the classic “triad” of hematuria, palpable abdominal mass and thrombocytopenia. However, these signs are not always all present. The classic presentation is that of an ill neonate who has a combination of hypertension, proteinuria, an enlarged kidney and hematuria. Risk factors include severe dehydration, hypotension, sepsis, asphyxia, prematurity and maternal diabetes. Renal ultrasound typically demonstrates an enlarged kidney with echogenic streaks radiating peripherally within the parenchyma and poor corticomedullary differentiation. Occasionally, a thrombus can be identified with Doppler. Computerized tomography typically shows an enlarged, edematous kidney with poor perfusion. Treatment of unilateral thrombosis is supportive care with hydration and close monitoring, whereas bilateral renal vein thrombosis requires more aggressive treatment with anticoagulation or systemic thrombolytics. Renal artery thrombosis is almost always associated with umbilical artery catheterization. It should be suspected in a neonate with an umbilical artery catheter who develops hematuria, hypertension, congestive heart failure or cyanotic limbs. The therapeutic options are controversial and based largely on small series or case reports, but they include conservative management with anticoagulation, surgical thrombectomy, systemic thrombolysis and catheter-directed thrombolysis.¹⁶

EXSTROPHY-EPISPADIAS COMPLEX

The exstrophy-epispadias complex is a rare spectrum of defects affecting the genitourinary and gastrointestinal tracts, musculoskeletal system, pelvic floor musculature and bony pelvis. The most common presentations of the spectrum include epispadias, classic bladder exstrophy and cloacal exstrophy. In classic bladder exstrophy, which is the most common, the umbilical cord should be suture-ligated to avoid erosion of the exstrophy plate by a clamp. A non-adherent and non-sterile dressing, such as cellophane, should be applied over the bladder plate rather than gauze, which will denude the mucosa over time. Prophylactic antibiotics should be initiated, and a renal ultrasound is recommended soon after birth. Cloacal exstrophy presents with omphalocele, exstrophy, imperforate anus and, frequently, spinal abnormalities, and gender identification may be difficult. Therefore, these patients should be transferred to a tertiary center with a pediatric general surgeon and pediatric urologist available. Initial management involves decompression of the obstructed genitourinary and intestinal tracts.

CLOACA ANOMALY

Cloacal malformations occur in a spectrum of severity and represent an extremely rare disorder with a varied presentation that demands surgical expertise in managing both anorectal and urogenital malformations. The external genital appearance

varies along a wide spectrum from nearly normal appearing to a perineal opening that exits onto a “doll-like” perineum, to a well masculinized phallic structure. **Urgent evaluation is mandatory. Diagnosis is key, and during the first 24 hours of life it is important to rule out associated congenital malformations that might be life-threatening.** Incorrect newborn management includes clinical misdiagnoses, failure to recognize and manage hydrocolpos appropriately, and pitfalls in colostomy and vesicostomy creation.¹⁷ Children born with a persistent cloaca require urgent clinical and radiological evaluation by an experienced pediatric surgeon, pediatric urologist and radiologist, and therefore require urgent transfer to a tertiary center.

AMBIGUOUS GENITALIA

A neonate born with atypical genitalia often results in severe parental anxiety. **It is essential in the care of these patients to involve a multidisciplinary team of clinicians who are familiar with evaluating and managing children with genital atypia and differences of sexual differentiation.** The most common cause of ambiguous genitalia at birth is congenital adrenal hyperplasia, which can affect up to 1 in 16,000 births. Initial evaluation should focus on identification of classic salt-wasting congenital adrenal hyperplasia since an elevated serum potassium and low serum sodium level can lead to dehydration and cardiac arrhythmias, and subsequent death. Laboratory testing should include karyotyping and serum studies, including electrolytes, 17-hydroxyprogesterone, testosterone and dihydrotestosterone levels. **These children should not be discharged until congenital adrenal hyperplasia has been ruled out.**

FEMALE GENITAL ABNORMALITIES

A urologist may be consulted for an interlabial mass in a pediatric female patient. These masses may range from having minimal significance, to threatening the urinary tract, to even threatening the child's life. Therefore, it is imperative to make a prompt and accurate diagnosis. The differential diagnosis includes a paraurethral cyst, imperforate hymen, prolapsed urethra, prolapsed ureterocele and a malignant vaginal botryoid rhabdomyosarcoma. The diagnosis is typically confirmed on physical examination. Examination of interlabial masses is best achieved by the “pull-down maneuver,” where the labia majora are gently grasped and pulled down and out from the midline. Use of a cotton-tipped swab to evaluate the entire contour of the mass and to look for important landmarks such as the urethral meatus and vaginal opening and their relationship to the mass may be helpful.

Both an imperforate hymen and a prolapsed urethra will be midline. An imperforate hymen appears as a white, bulging membrane at the vaginal introitus. The urethral meatus is normal and non-obstructed. Ultrasonography will demonstrate the hydrometrocolpos or vaginal cavity distension as a fluid collection behind the bladder. Hydronephrosis resulting from the distended vagina is not uncommon. Treatment involves a horizontal incision of the hymen to allow the vaginal secretions to drain. Urethral prolapse is a circumferential collar of edematous and often ecchymotic urethral mucosa encircling the urethral meatus. Treatment is symptomatic with warm compresses, sitz baths and topical moisturizers, but persistence may require excision of the prolapsed tissue.

Prolapsed ureterocele is an emergency, whereas paraurethral cysts are not. Both are often offset from the midline meatus and typically have an intact hymenal ring posterior to the lesion. A prolapsed ureterocele appears as a smooth, round, dark red or purple cystic structure from within the urethral meatus. **Ureteroceles not only may obstruct the associated ureter, but when prolapsed may result in urinary retention affecting the entire urinary tract.**¹⁸ At times they are large enough to obscure all of the anatomical landmarks. Renal-bladder ultrasonography and voiding cystourethrography (when possible) will confirm the diagnosis, and treatment is emergent endoscopic decompression, often with incision of the ureterocele. A parametatal or paraurethral cyst (Skene's duct cyst) is an ovoid structure that will sometimes displace the urethral meatus and may deflect the urinary stream. The large majority of these cysts resolve spontaneously within a few weeks or rupture without intervention.¹⁹ If the diagnosis is certain, a small needle may be used to puncture the cyst. If the diagnosis is uncertain or the cysts fail to resolve, the child should be evaluated by a pediatric urologist to assess for other etiologies including urethral diverticulum, ectopic ureterocele or obstructed hemivagina.

Rhabdomyosarcoma is the most common soft-tissue tumor of childhood and young adults, accounting for 4%–6% of all malignancies in this age group, and the genitourinary tract is the second most common site after the head and neck.²⁰ In the urinary tract, these may present with gross hematuria, urinary retention, vaginal bleeding and a visible or palpable mass. Sarcoma botryoides occurs in the vagina during infancy and early childhood, and typically presents as soft nodules protruding from the vagina, resembling a bunch of grapes. **A child with suspected rhabdomyosarcoma of a genitourinary site requires prompt evaluation by a pediatric urologist as these are life-threatening.**

ABDOMINAL DISTENSION

Pediatric abdominal distension is most commonly of urological etiology, including hydronephrosis, cystic mass, solid mass, and urinary retention and ascites due to obstruction or extravasation. Initial assessment should include abdominal ultrasound evaluation of the upper and lower urinary tract.

Severe hydronephrosis due to upper urinary tract obstruction may result in a palpable abdominal mass. The most common site of obstruction is the ureteropelvic junction, but obstruction may also occur at the ureterovesical junction. Percutaneous drainage may be required in rare scenarios due to respiratory compromise or feeding intolerance, and these patients should be managed at centers with pediatric subspecialty care.

Urinary retention due to bladder outlet obstruction most commonly occurs in males and may be caused by posterior urethral valves and urethral atresia. Bladder outlet obstruction is rare in females, but may occur with prolapsed ureteroceles. Catheter placement into the bladder is indicated for diagnosis and management. However, this may be difficult in select scenarios. Males with posterior urethral valves often have a high bladder neck, requiring the urologist to attempt placing a finger in the rectum to deflect the catheter above the bladder neck. Females may also develop urinary retention in the case of a very large or prolapsed ureterocele, or persistent urogenital sinus or cloaca. The catheter should be directed anteriorly toward the bladder in females; however, drainage of the vagina often is adequate to temporarily decompress the lower urinary tract. If attempts to decompress the bladder with catheter

placement are unsuccessful, then percutaneous drainage may be required and aided with the use of ultrasonography. The Seldinger technique using a 16-gauge angiocatheter will accommodate passage of a 3.5Fr feeding tube, while a 14-gauge angiocatheter will accommodate passage of a 5Fr feeding tube.²¹ At times, in posterior urethral valves or urogenital sinus anomalies, a cutaneous vesicostomy may be required.

Cystic and solid renal lesions in children that may prompt urological consultation include renal cysts, multicystic dysplastic kidney, autosomal recessive or dominant polycystic kidney disease, cystic nephroma and solid renal masses. Renal lesions can often be distinguished on ultrasonography along with a careful family history to detect autosomal dominant or autosomal recessive polycystic kidney disease. Further evaluation is then determined based on sonographic findings. **A multicystic dysplastic kidney will appear as a multiloculated mass with little to no renal parenchyma on ultrasonography and no functional renal tissue on nuclear medicine renal scan.** Solid renal masses in children include pseudotumors, congenital mesoblastic nephroma, Wilms tumor, clear cell sarcoma, renal cell carcinoma, ossifying renal tumor, rhabdoid tumor and neuroblastoma. A child found to have a solid renal tumor should be promptly evaluated by a pediatric urologist.

TRAUMA

The American Urological Association Urotrauma guidelines provide detailed recommendations for evaluation and management of genitourinary trauma with no change in management for pediatric patients, and therefore we have not spent significant time discussing it in this Update.¹² **However, it is important to note that the sympathetic tone in children may maintain a normal blood pressure despite significant blood loss, making hypotension a less reliable sign of acute blood loss in children.**²²

Pediatric penile trauma may be iatrogenic, occurring during circumcision, or commonly caused by domestic animal attack, child abuse, motor vehicle accidents, trapping in a zipper and penile strangulation by hair, thread or Plastibell® ring.²³ Hair or thread at the coronal margin or a retained Plastibell ring may result in ischemic damage and should be promptly removed. Circumcision injuries are usually minor, such as minor skin loss, and generally heal spontaneously with local wound care. However, more significant injuries, especially those with glans amputation, may necessitate emergent care by a pediatric urologist for possible re-anastomosis of the amputated organ.

PRIAPISM

Priapism in children is a urological emergency as it is in adults. In children, sickle cell disease is the most common cause, followed by trauma, leukemia, medication-induced and idiopathic causes.²⁴ Evaluation and management are similar to adults, with goals to prevent erectile dysfunction, penile disfigurement and psychological sequelae. The physician must assess the type of priapism (ischemic versus non-ischemic) and aim to achieve detumescence. Conservative measures include physical exercise such as running up a flight of stairs, urination, a cold bath, ejaculation and fluids. A complete blood panel should be performed to exclude hemoglobinopathies and leukemia. If priapism persists, the clinical features and identification of an underlying cause will guide management. Children are generally more likely to require general anesthesia or dissociative

sedation for painful procedures. **The urologist must consider the risk of lasting psychological damage, aspiration complications, anesthetic risk and risk of an anesthetic delay when choosing the optimal anesthetic.** In ischemic priapism, potential interventions include intracorporal injection, corporal aspiration and lavage, or more invasive interventions including shunt procedures, whereas management of non-ischemic priapism is not urgent and is typically conservative.

CONCLUSIONS

Emergency cases are those that threaten life, limb or organ. Fortunately, these are infrequent in pediatric urology. In this Update, we have described the urological emergencies that occur in pediatric patients which general urologists may encounter and must be prepared to evaluate and manage appropriately. We have reviewed the urgent urological problems seen in infancy and childhood with a description of typical presenting signs or symptoms, differential diagnoses, proper evaluation and diagnostics, and recommended courses of management.

REFERENCES

- Williamson RC: Torsion of the testis and allied conditions. *Br J Surg* 1976; **63**: 465.
- Zhao LC, Lautz TB, Meeks JJ et al: Pediatric testicular torsion epidemiology using a national database: incidence, risk of orchiectomy and possible measures toward improving the quality of care. *J Urol* 2011; **186**: 2009.
- John CM, Kooner G, Mathew DE et al: Neonatal testicular torsion—a lost cause? *Acta Paediatr* 2008; **97**: 502.
- Yerkes EB, Robertson FM, Gitlin J et al: Management of perinatal torsion: today, tomorrow or never? *J Urol* 2005; **174**: 1579.
- Baker LA, Sigman D, Mathews RI et al: An analysis of clinical outcomes using color doppler testicular ultrasound for testicular torsion. *Pediatrics* 2000; **105**: 604.
- Visser AJ and Heyns CF: Testicular function after torsion of the spermatic cord. *BJU Int* 2003; **92**: 200.
- Kutikov A, Casale P, White MA et al: Testicular compartment syndrome: a new approach to conceptualizing and managing testicular torsion. *Urology* 2008; **72**: 786.
- Dresner ML: Torsed appendage. Diagnosis and management: blue dot sign. *Urology* 1973; **1**: 63.
- Baldisserotto M, de Souza JC, Pertence AP et al: Color Doppler sonography of normal and torsed testicular appendages in children. *AJR Am J Roentgenol* 2005; **184**: 1287.
- Leslie JA and Cain MP: Pediatric urologic emergencies and urgencies. *Pediatr Clin North Am* 2006; **53**: 513.
- Rettenbacher T, Hollerweger A, Macheiner P et al: Abdominal wall hernias: cross-sectional imaging signs of incarceration determined with sonography. *AJR Am J Roentgenol* 2001; **177**: 1061.
- Morey AF, Brandes S, Dugi DD et al: Urotrauma: AUA Guideline. *J Urol* 2014; **192**: 327.
- Ioannides AS and Turnock R: An audit of the management of the acute scrotum in children with

DID YOU KNOW?

- The pediatric acute scrotum is a common complaint with a wide array of etiologies that must be promptly evaluated to facilitate the potentially time-sensitive interventions required.
- Hematuria in children may result from trauma or interstitial, glomerular, vascular or urothelial sources and is more frequently associated with medical problems than with surgical problems in children. However, proper evaluation is essential to diagnose potentially life-threatening etiologies and allow for appropriate management.
- The general urologist must be familiar with congenital genitourinary anomalies and any required acute management in order to appropriately triage the care of these patients.

- Henoch-Schonlein purpura. *J R Coll Surg Edinb* 2001; **46**: 98.
- Avolio L, Fusillo M, Ferrari G et al: Neonatal adrenal hemorrhage manifesting as acute scrotum: timely diagnosis prevents unnecessary surgery. *Urology* 2002; **59**: 601.
- Cain MP, Casale AJ, King SJ et al: Appendicovesicostomy and newer alternatives for the Mitrofanoff procedure: results in the last 100 patients at Riley Children's Hospital. *J Urol* 1999; **162**: 1749.
- Silverberg D, Menes T, Rimon U et al: Acute renal artery occlusion: presentation, treatment, and outcome. *J Vasc Surg* 2016; **64**: 1026.
- Levitt MA and Pena A: Pitfalls in the management of newborn cloacas. *Pediatr Surg Int* 2005; **21**: 264.
- Badalyan V, Burgula S and Schwartz RH: Congenital paraurethral cysts in two newborn girls: differential diagnosis, management strategies, and spontaneous resolution. *J Pediatr Adolesc Gynecol* 2012; **25**: e1.
- Herek O, Ergin H, Karaduman D et al: Paraurethral cysts in newborn: a case report and review of literature. *Eur J Pediatr Surg* 2000; **10**: 65.
- Nasioudis D, Alevizakos M, Chapman-Davis E et al: Rhabdomyosarcoma of the lower female genital tract: an analysis of 144 cases. *Arch Gynecol Obstet* 2017; **296**: 327.
- Herndon A and Zee RS: Perinatal urology. In: Campbell-Walsh-Wein Urology, 12th ed. Edited by AW Partin, RR Dmochowski, LR Kavoussi et al. Philadelphia: Elsevier 2021; chapt 22.
- Quinlan DM and Gearhart JP: Blunt renal trauma in childhood. Features indicating severe injury. *Br J Urol* 1990; **66**: 526.
- El-Bahnasawy M and El-Sherbiny M: Paediatric penile trauma. *BJU Int* 2002; **90**: 92.
- Donaldson JF, Rees RW and Steinbrecher HA: Priapism in children: a comprehensive review and clinical guideline. *J Pediatr Urol* 2014; **10**: 11.

Study Questions Volume 40 Lesson 29

1. A 1-week-old girl is referred to the emergency department after the pediatrician noted an intralabial mass. Physical examination reveals a round and smooth red cyst from within the urethral meatus and a palpable suprapubic mass. The next step in management is
 - a. reassurance
 - b. percutaneous puncture with a small needle
 - c. Foley catheter placement with reduction of the mass into the bladder
 - d. renal-bladder ultrasound
2. A newborn boy whose mother had no prenatal care is born at 35 weeks with acute respiratory failure at birth requiring intubation and significant respiratory support. He has not voided in 24 hours and ultrasound reveals a distended bladder, left grade 3 hydroureteronephrosis and right grade 4 hydroureteronephrosis. You were unable to place a urethral catheter despite several attempts and maneuvers due to proximal resistance. The next step in management is
 - a. reassurance
 - b. percutaneous suprapubic tube placement with ultrasound guidance
 - c. cystoscopy and catheter placement in the operating room
 - d. vesicostomy
3. A newborn boy is noted to have a right scrotal swelling after prolonged labor. Physical examination reveals an enlarged right scrotum with purple discoloration and a palpably normal left testicle; however, you are unable to palpate the right testicle due to the swelling. Scrotal ultrasound demonstrates normal appearing testicles with adequate vascular flow bilaterally. The next step is
 - a. reassurance
 - b. abdominal ultrasound
 - c. right scrotal exploration
 - d. general surgery consultation
4. A neonate born with atypical genitalia is found to have a single perineal opening and a cystic pelvic mass. The cystic mass likely represents
 - a. urinary ascites
 - b. distended rectum
 - c. distended vagina
 - d. distended bladder
5. A 4-year-old boy with right lower quadrant abdominal pain is found to have an incidental left multiloculated mass with no apparent renal parenchyma on ultrasonography. The next step is
 - a. outpatient observation
 - b. computerized tomography of abdomen and pelvis with and without contrast
 - c. genetic testing
 - d. left nephrectomy

Informed Consent

Learning Objective: At the conclusion of this continuing medical education activity, the participant will be able to describe the components required for adequate informed consent, define standards for informed consent of minor age patients and list important elements for informed consent of human subjects involved in clinical research.

This AUA Update aligns with the American Board of Urology Module on Core/General Urology. Additional information on this topic can be found in the AUA Core Curriculum section on Ethics.

C. Adam Lorentz, MD,¹ Adam C. Webb, MD² and Chad W. M. Ritenour, MD³

¹Assistant Professor, Department of Urology, Emory University, Atlanta, Georgia

²Associate Professor, Departments of Neurology, Neurosurgery and Pediatrics, Emory University, Chief Quality Officer, Emory University Hospital, Emory Healthcare, Atlanta, Georgia

³James C. Kennedy Professor in Prostate Health, Department of Urology, Chief Medical Officer, Emory University Hospital, Emory Healthcare, Atlanta, Georgia

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