Preface

The practice and science of pediatric urology has changed rapidly in the last few years. New surgical techniques, better and more informative prenatal evaluations, and better biomedical substances continue to arrive on the scene. *Pediatric Urology* offers a unique practice-oriented approach to these new developments offered by known experts in the field.

Pediatric Urology updates the reader on the modern evaluation of prenatal hydronephrosis and compares the standard treatment of reflux with newer injectable techniques. The spectrum of treatment for hypospadias is described along with new developments in bladder exstrophy and what's new in the undescended testis. The evaluation of difficult duplication anomalies is described along with their treatment. A unique and very modern approach to voiding dysfunction and neurogenic bladders is presented in a quite logical and straightforward manner. Finally, pediatric stone disease and developmental problems associated with genitourinary defects are described.

We hope *Pediatric Urology* will show the practicing urologist in a clear, concise way where pediatric urology is going and how new discoveries are being applied in our field. Many thanks to our authors for taking time from their families, practices, residents, and research to share their expertise with our readers.

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Pediatric Urologic Emergencies

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INTRODUCTION

Fortunately, few emergencies in pediatric urology result in loss of life. However, these true emergencies, if not recognized and treated quickly, can lead to loss of or severe injury to the affected organ (testicular torsion and severe renal trauma) or to possible loss of life (adrenal hemorrhage and urosepsis). Table 1 lists the emergencies that a urologist may be called on to evaluate and treat. In addition, there are a number of urologic conditions that require expeditious evaluation and treatment. These conditions are referred to as "urgencies" and are listed in Table 2. Although many of these conditions are not immediate threats to life or to end organs, failure to properly diagnose and treat them can have catastrophic consequences.

PEDIATRIC EMERGENCIES

Abdominal Masses

Hydronephrosis

Most abdominal masses in children are found in the retroperitoneum or genitourinary tract and should, therefore, be evaluated by a pediatric urologist (1,2) (Table 3). The most common abdominal mass in the

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Table 1 Pediatric Emergencies

Acute scrotum Ambiguous genitalia Renal vein thrombosis Renal artery thrombosis Adrenal hemorrhage Posterior urethral valves Priapism Trauma Child abuse Abdominal mass Urolithiasis Paraphimosis

Table 2 Pediatric "Urgencies"

Exstrophy Prune-belly syndrome Hematuria Imperforate anus Urethral prolapse Urinary tract infections Myelodysplasia

newborn remains hydronephrosis (3,4) (Fig. 1 A,B), particularly as a result of ureteropelvic junction (UPJ) obstruction (5,6) (Fig. 2). In most cases, the diagnosis is made by prenatal ultrasonography. In most fetuses, hydronephrosis is unilateral and should be followed by a pediatric urologist until birth (7). The newborn should be placed on antibiotic prophylaxis and sonography should be performed. Beware the ultrasound done within the first 24 to 72 h postdelivery because the relative dehydration that occurs during the birth process may mask true hydronephrosis (8). The ultrasound should be repeated 1 wk to 1 mo after birth to confirm prenatal findings. A voiding cystourethrogram (VCUG) is performed to look for vesicoureteral reflux (VUR), and a well-tempered renogram is done to evaluate for obstruction (9, 10). If obstruction or reflux is not found, the patient can be taken off antibiotic prophylaxis. Treatment for a confirmed UPJ obstruction should be based on the degree of obstruction and whether the obstruction is unilateral or bilateral (7,11). Clearly, severe bilateral obstruction, as demonstrated on ultrasound and furosemide renal scan, must be corrected

Table 3 Abdominal Masses in Neonates and Children

Hydronephrosis MCDK Polycystic kidney Distended bladder Hydrometrocolpos Ovarian cyst Neuroblastoma Wilms' tumor Mesoblastic nephroma Adrenal hemorrhage Renal vessel thrombosis Pyloric stenosis Germ cell tumors Hepatic lesions Sacrococcygeal teratoma

MCDK, multicystic dysplastic kidney



Fig. 1. (A) Hydronephrotic right kidney in a newborn.

early (12). Many children who have unilateral UPJ obstruction with function greater than 40% in the ipsilateral kidney can be followed with ultrasound and renal scintigraphy (13). Ransley et al., Koff and



Fig. 1. (B) Marked right hydronephrosis in another child with prenatal diagnosis of hydronephrosis. There is marked pelviectasis and caliectasis.



Fig. 2. Severely hydronephrotic left kidney, with thin parenchyma. At the bottom right of the ultrasound is the bladder.

Campbell, and others have demonstrated that careful follow-up of children with stable hydronephrosis and obstruction is safe. In most children, the obstruction resolves spontaneously (13-17).



Fig. 3. (**A**) Right hydroureter entering the bladder and ending into an ureterocele. (**B**) Bilateral single-system ureteroceles.

In addition, hydronephrosis may be the result of obstruction at the ureterovesical junction by ureteroceles (Fig. 3 A,B). In fact, an ectopic ureterocele is the most common cause of bladder outlet obstruction in females (10% of cases) (18) and the second most common cause in males (second to posterior urethral valves) (19,20). These patients also have hydroureter down to the level of the bladder, and classic ultrasound



Fig. 4. Large introital mass, which was a prolapsed ureterocele in a female.

finding of an echogenic rim of tissue that may protrude into the bladder neck (21). Prolapse of the ureterocele may develop in some patients (Fig. 4), and older patients may have calculi within the ureterocele resulting from urinary stasis. Evaluation includes renal and bladder ultrasound, VCUG, and if indicated, a furosemide renal scan. Treatment for obstructing ureteroceles is somewhat controversial. Some authors advocate transurethral incision (22-24). However, there is a high incidence of VUR as a consequence of incision, possibly necessitating reimplant and bladder floor reconstruction (25). Resection with ureteral re-implant and reconstruction of the bladder floor has also been recommended. Obstructing ureteroceles associated with complete ureteral duplication are more common and need to be evaluated in a similar manner to single systems (26). The upper pole moiety is almost always affected and may have minimal function. Consideration should be given to performing a heminephroureterectomy in children with obstructed upper pole moieties and poor function. Many children with complete duplication and an upper pole ureterocele also have reflux into the lower pole moiety and should be followed over time (18).

Other lesions that may be misinterpreted as hydronephrosis include multicystic dysplastic kidney (MCDK), cystic nephroma, and autosomal-dominant and autosomal-recessive polycystic kidney disease. MCDK is the second most common abdominal mass found in infants (27). Both sexes are equally affected. Children found to have MCDK should have a VCUG because these patients have a high incidence of contralateral vesicoureteric reflux (28). The diagnosis can be made by ultrasound, which demonstrates a multiloculated mass with little or no parenchyma. The cysts do not communicate. If there is any question whether the child has MCDK or severe UPJ obstruction, a renal scan will confirm the diagnosis (29). Treatment has changed since the late 1980s. Whereas many children with MCDK underwent nephrectomy in the past, they are now being followed with serial renal ultrasounds. MCDKs generally regress over time. Surgery is reserved for those patients in whom MCDK does not regress or causes symptoms (30,31).

MIDLINE ABDOMINAL MASS

A child with a midline abdominal mass should also be evaluated with ultrasonography. In the emergency department, the most common cause of a midline lower abdominal mass is a distended bladder (32). In males, the presence of posterior urethral valves must be considered and evaluation undertaken to confirm or refute the diagnosis (33). In addition, neurologic causes of urinary retention need to be addressed, such as spinal dysraphism or spinal cord tumor (34).

Hydrocolpos is distension of the vagina that occurs as a result of a build up of cervical mucus behind an imperforate hymen (most commonly) or as the result of some other vaginal abnormality (35). In 1984, Han-Pedersen et al. demonstrated that cervical mucus secretion was a result of maternal estrogen (36). Most neonates will have a lower midline abdominal mass, a bulging mass in the perineum, and hydronephrosis resulting from bladder outlet obstruction caused by the distended vagina. An incision in the hymen is usually all that is needed to completely drain the vagina and relieve the bladder obstruction (35).

Other causes of abdominal mass include ovarian cysts, sacrococcygeal teratomas, and neonatal ascites. Ovarian cysts can be the result of ovarian torsion followed by cystic degeneration of the ovary. More commonly, ovarian cysts are the result of maternal hormonal stimulation and regress quickly in the absence of any stimulus (37). Sacrococcygeal teratoma occurs in 1 in 40,000 births and occurs predominantly in females (38). Like hydrocolpos, sacrococcygeal teratomas can cause outlet obstruction and hydronephrosis (39). Excision of the teratoma can lead to neurogenic bladder (in up to 12% of patients); therefore, the child must be followed to assess for any neurologic sequelae (40).

Urinary ascites can be diagnosed in the fetus or in the newborn period. Ascites is commonly caused by urinary extravasation resulting from urinary tract obstruction or trauma (41). Sahdev et al., in 1997, reported that extravasation of urine occurred most commonly as a result of forniceal rupture in the kidney (42). Some authors believe that extrava-



Fig. 5. Young female with large abdominal mass. Evaluation revealed a large renal mass, which was found to be Wilms' tumor.

sation of urine is a protective mechanism, decreasing the amount of damage to the kidneys from obstruction (43, 44). Bladder perforation as a result of umbilical artery catheterization can also lead to urinary ascites (45). The diagnosis can be made by ultrasonography and VCUG in most cases, and the bladder should be surgically repaired (46).

Solid Abdominal Masses

Solid abdominal masses are now being diagnosed with increasing frequency in the prenatal period. However, many solid abdominal masses are still discovered by physical examination after birth (Fig. 5). The most common solid abdominal mass in children is neuroblastoma (47). Neuroblastoma is a firm, fixed mass that can cross the midline. It can arise from the adrenal gland or from anywhere along the sympathetic ganglia. Although these lesions can be diagnosed with ultrasound, computed tomography (CT) or magnetic resonance imaging (MRI) are usually used to confirm the diagnosis. On KUB, these lesions can have fine calcifications, which suggest the diagnosis. In addition, this tumor is of neural crest origin, and therefore laboratory testing for increased levels of catecholamines (vanillymandelic acid and homovanillic acid) in the urine is an essential part of the diagnosis of neuroblastoma. Bone marrow biopsy and lymph node sampling are done at the time of surgery because of the very high (90%) incidence of metastatic disease. Patients receive chemotherapy with or without radiation therapy based on the stage of disease (48).

Congenital mesoblastic nephroma is the most common solid mass of the kidney in children less than 6 mo of age. The majority of these masses are discovered during routine well-baby examinations within the first month of life. Although locally invasive, these fibroid-appearing tumors rarely metastasize. Treatment is elective nephrectomy (49).

The most common solid renal tumor in children is Wilms' tumor (50). These children also have an abdominal mass. On CT or MRI, these tumors involve the kidney and are expansile in nature. They rarely cross the midline. A genetic mutation on chromosomes 11p13 and 11p15 has been demonstrated in children with Wilms' tumor. These children may also have hemihypertrophy and aniridia. The pathologic findings may be favorable or unfavorable and will dictate, along with surgical stage, the most appropriate course of therapy. Other solid abdominal masses in children include multilocular cystic nephroma, renal cell carcinoma, pheochromocytoma, adrenal adenoma, adrenal carcinoma, and rhabdomyosarcoma (50, 51).

Thrombosis of Renal Vessels

Renal vein thrombosis in neonates is characterized by gross hematuria, abdominal mass, dehydration, or thrombocytopenia (52,53). Renal vein thrombosis is a common cause of gross hematuria in the first month of life. In addition, renal vein thrombosis can be found in children with gross hematuria (54). Low renal artery and venous flow rates appear to predispose children to renal vein thrombosis. Factors that are most commonly associated with renal vein thrombosis include sepsis, dehydration, maternal diabetes, polycythemia, birth asphyxia, and umbilical artery catheterization (55).

Evaluation of children with renal vein thrombosis includes an ultrasound, which demonstrates echogenic streaks and the absence of hydronephrosis. A clot may be seen within the renal vein and may extend to the vena cava. Unilateral renal vein thrombosis is treated with supportive care, namely fluids and correction of any metabolic abnormality. The use of systemic anticoagulation or thrombolytic therapy is usually reserved for children with bilateral renal involvement (56,57). The prognosis for patients with renal vein thrombosis worsens in those with bilateral disease. Children with bilateral renal vein involvement have a higher rate of renal failure. These children require long-term follow-up to manage their renal insufficiency or to identify new-onset hypertension.

The increasing use of umbilical artery catheterization has resulted in an increased incidence of renal artery thrombosis (58). Children with renal artery thrombosis do not typically have an abdominal mass. However, they may have hematuria, hypertension, and congestive heart failure. The diagnosis can be made and confirmed with Doppler ultrasound, which demonstrates no flow to the affected kidney. Treatment is usually supportive but can include intra-arterial thrombolytic therapy or unilateral nephrectomy (59,60).

Adrenal Hemorrhage

Adrenal hemorrhage is most commonly found in newborns who have been through a dramatic or difficult labor, or who have hypoxia, septicemia, or coagulopathy. The right adrenal gland appears to be the most commonly affected, and 10% of adrenal hemorrhages occur bilaterally (20). Felc in 1995 was able to show an incidence of 1.9 per 1000 births (61). The large and hypervascular adrenal glands in neonates appear to be predisposed to bleeding, especially in those neonates described above. Children may have hemodynamic instability and may also have an abdominal mass.

The diagnosis can usually be made by ultrasound but may be difficult to differentiate from neonatal neuroblastoma. Additional radiographic studies using CT and MRI, and measures of urinary vanillymandelic acid and homovanillic acid help to solidify either diagnosis. Adrenal insufficiency is uncommon. Treatment is generally supportive, and occasionally transfusion and phototherapy may be used (62).

Posterior Urethral Valves

Posterior urethral valves were originally described by Young et al. in 1919(63). They described three types of posterior urethral valves. Type I and type III valves were confirmed over time. Type II valves do not appear to exist. Posterior urethral valves are most commonly diagnosed perinatally but can be diagnosed into adulthood (64). Prenatal ultrasound demonstrates a distended, thick-walled bladder, unilateral or bilateral hydronephrosis, and even oligohydramnios in some cases (65). In the nursery, the pediatric urologist may be called to see the newborn who fails to void within the first 24 h of life. On physical examination, a palpable bladder may be found (33). An abnormal urinary stream is found in some neonates. Ultrasound may show unilateral or bilateral hydronephrosis, a thickened bladder wall, and dilation of the posterior urethra. VCUG further identifies the posterior urethral valves and the presence or absence of reflux (66). Some of these children may also have urinary ascites as a result of forniceal rupture caused by bladder outlet obstruction (41,42). The vesicoureteral reflex dysplasia (VURD) syndrome was identified by Rottenberg et al. in 1988 (67). These boys were found to have severe VUR with ipsilateral renal dysplasia. The contralateral kidney did not have reflux. The authors postulated that the severe reflux with dysplasia actually acted as a protective mechanism for the nonrefluxing kidney, and in these boys, renal function after valve ablation surgery was generally normal. Children who are thought to have posterior urethral valves should be placed on antibiotic prophylaxis. The mainstay of surgical therapy for posterior urethral valves is transurethral ablation of the valve leaflets.

This is most commonly performed in a retrograde manner and can be done with electrocautery or laser ablation (68). Antegrade valve ablation has also been described (69). It is important to identify rupture (ablation) of both valve leaflets to assure opening of the posterior urethra. In children whose urethral caliber is too small to accommodate the resectoscope equipment, vesicostomy is a viable alternative. The longterm prognosis depends on several factors, including renal function, before and after valve ablation, and the long-term management of bladder function. Children with serum creatinine level less than 0.7 mg/ dL after valve ablation surgery have a better prognosis (68). The valvebladder syndrome has been described and can lead to dysfunction of the bladder and long-term upper track dilation (70,71). Some form of renal failure develops in many of these children at the time of puberty because of increased muscular mass along with voiding dysfunction.

Therefore, it is crucial for the pediatric urologist to maintain longterm follow-up on all boys with posterior urethral valves.

Ambiguous Genitalia (Intersex Disorders)

Intersex disorders are now being identified in the antenatal period. Ultrasound techniques, amniocentesis, and chorionic villae sampling in the prenatal period have assisted in the antenatal diagnosis of intersex disorders (72,73). However, the presence of ambiguous genitalia continues to be the most common means of identifying a child with an intersex disorder (Fig. 6 A,B). The most common cause of ambiguous genitalia is congenital adrenal hyperplasia (74) and 21-hydroxylase deficiency is the leading cause of congenital adrenal hyperplasia. This results in the inability to synthesize cortisol, leading to overproduction of androstenedione as a result of overstimulation of the adrenal gland by the pituitary gland and hypothalamus. Androstenedione is then metabolized to testosterone and dihydrotestosterone, resulting in virilization in females (75). In addition, the lack of mineralocorticoid and glucocorticoid can lead to elevated serum potassium level and decreased serum sodium level, resulting in dehydration or cardiac dysrhythmia (76). Therefore, it is essential that children with any form of ambiguous genitalia have serum chemistry testing to rule out electrolyte abnormalities.



Fig. 6. (A) Toddler with ambiguous genitalia. Note the absence of gonads in the labioscrotal folds. (B) Same child with retraction of phallus. Child was found to have congenital adrenal hyperplasia resulting from to 21-hydroxylase deficiency.

These children should also have a karyotype analysis performed. The family history may include prior neonatal deaths, sterility or amenorrhea, and maternal medication use during pregnancy. In addition to



Fig. 7. Child from Fig. 6 after feminizing genitoplasty.

family history, thorough physical examination and laboratory studies should be performed. These studies, along with the karyotype analysis, will diagnose the majority of children with intersex disorder (77). In addition to potentially life-threatening electrolyte abnormalities, these children are also considered to have a social emergency. It is difficult for parents to understand that the correct sex of rearing cannot be determined at the time of birth. It will be based on the karyotype; phenotype; levels of 17-hydroxyprogesterone, testosterone, and dihydrotestosterone; ultrasound of the pelvis (looking for the presence of female organs); and genitography of the child. Understandably, parents can sometimes be impatient waiting for test results. Therefore, treatment of the child with an intersex disorder must be individualized and should include specialists from other fields. Once gender assignment has taken place, surgical reconstruction can be quite successful (Fig. 7).

Child Abuse

Child abuse is defined by the National Center for Child Abuse and Neglect as "any contact or interaction between a child and an elder where the child is used for sexual stimulation of that individual or another" (78). Perineal examination of both boys and girls should be routinely performed in the physician's office. Examination of the vagina may reveal an enlarged hymenal opening. However, Gardner (79) has described normal hymenal diameter and appearance of the introitis. Most institutions have a mechanism for using a multidisciplinary approach to evaluate any child suspected of being a victim of abuse. For any child seen in the emergency department with what appears to be a sexually transmitted disease, it is important not only to diagnose and treat the infection correctly, but it is also important to ascertain how the disease was acquired. In the prepubertal child, any evidence of sexually transmitted disease must be considered child abuse until evaluated and proven otherwise (78,80). In addition, pregnancy may be the first sign of child abuse. In boys, examination of both the back and perianal region is important in looking for signs of abuse. It is essential to evaluate children for possible sexual abuse in any suspected case.

Trauma

Most genitourinary trauma occurs as a result of blunt injury. The kidney is the most common genitourinary structure injured as a result of trauma. Injuries range from contusions to severe pedicle injuries. A proportionally increased kidney size, paucity of perirenal fat, and persistence of fetal lobulation are all factors that predispose the kidney to injury (81). The presence of renal anomalies is also a predisposing factor to renal injury in up to 25% of cases (82). The kidney is injured most often in deceleration injuries, such as a fall, a motor vehicle accident, or a sports injury (83). Hematuria and flank tenderness are the most common findings associated with renal injury. However, as is seen in the adult population, the degree of hematuria does not necessarily correlate with the severity of the renal injury. In most emergency departments, evaluation of the child with trauma and any degree of hematuria is done with CT to assess the genitourinary tract (84). If renal trauma is found, the treatment is usually conservative, with admission to the hospital for more severe renal injuries (parenchymal lacerations and shattered kidney). As in adults, surgical treatment is reserved for the hemodynamically unstable patient or for instances when there are other associated abdominal injuries that require surgical intervention (85). The ureter is an infrequently injured structure (86). However, disruption can occur at the UPJ in a child who has undergone blunt abdominal trauma and had an unidentified UPJ obstruction (81–84). The bladder is more vulnerable to trauma as a result of its relative intra-abdominal location. Most trauma injuries are associated with pelvic fractures and most are extraperitoneal in location. Surgical exploration is recommended for children with bladder rupture when there is intraperitoneal rupture or associated bony fragments (87).

Urethral injuries occur most commonly at the prostatomembranous junction. These occur as a result of rapid deceleration injuries, as seen in children and adults with pelvic fracture. The urethra may also be injured by a straddle injury, as occurs in boys who fall onto the crossbar of a bicycle (88). The penis can be injured by various mechanisms. In toddlers and young boys, injury to the penis as a result of being crushed between the toilet seat and the commode is a common mechanism of injury. Zipper injuries of the penis and foreskin also occur and can be extremely difficult to treat in the apprehensive child (32). Flowerdew et al. in 1977 described a successful technique to disassemble the zipper in this type of injury (89). Strangulation or tourniquet injuries may also occur as a result of human hair or string being wrapped around the penis. Many also occur as a result of rubber bands, wires, or other structures placed around the penis (32). Treatment of this type of injury depends on identification of the offending agent.

Scrotal and testicular injury most often occur as a result of blunt trauma. These injuries range from scrotal ecchymosis to testicular rupture. Many children are injured during sports activities because they are not wearing proper protective gear. A scrotal ultrasound may be needed to assess the integrity of the testicular parenchyma, and scrotal exploration may be required to repair a ruptured testicle (90). Vulvar trauma commonly occurs as a result of a straddle injury, which can be seen when a girl falls onto the bar of a boy's bicycle (88). These injuries can be treated conservatively in the majority of cases but can be of such severity as to require a general anesthetic for proper diagnosis.

Phimosis, Paraphimosis, and Circumcision Injuries

Few newborns have a fully retractable foreskin at birth. Phimosis, or a stenosed prepuce, can be associated with a forceful retracting of the prepuce over the glans penis and the resultant preputial scarring that occurs. The natural history of the adherent foreskin is to eventually loosen and become fully retractable by the age of 10 or 11 yr. Phimosis may be caused by episodes of balanitis, balanoposthitis, and urinary tract infections (35,91). Treatment is commonly circumcision, but a dorsal or ventral slit can correct the problem and allow the child to keep the prepuce (92,93). Paraphimosis results from retraction of the prepuce proximal to the glans for a prolonged period of time. As result, edema develops and causes a progressive constriction around the penis. Light sedation may be required in younger children to reduce the paraphimosis. To reduce the paraphimosis, manual compression of the glands is done while simultaneously reducing the edematous foreskin. The edema resolves rapidly after reduction of the foreskin (94).

Most neonatal circumcisions are performed in the nursery by either pediatric or gynecologic personnel. There are few indications for neonatal circumcision, and most males in the world are not circumcised. The complication rate associated with circumcision is 0.2–10%. The incidence may be even higher because many patients are treated without referral. The most acute complications are bleeding, infection, partial amputation, and necrosis. Necrosis is most commonly caused by overuse of electrocautery, whereas partial amputation may be the result of incorporation of the frenulum into a Mogen clamp (95). Persistent bleeding can be treated by direct pressure, placement of absorbable suture, or cautery. In the event of penile amputation, the amputated portion can be re-attached in the majority of cases. Long-term, nonacute complications include removal of excess skin, skin bridges, skin chordee, epidermal inclusion cysts, and development of a concealed (buried) penis (95). A concealed penis can result from the penis retracting below the circumcision scar and subsequent scar contraction (96).

Acute Scrotum and Scrotal Masses

A child with an acute scrotum is one of the true emergencies for a urologist. The differential diagnoses include torsion of the spermatic cord, torsion of a testicular or epididymal appendage, and epididymitis. Most patients have a history of severe pain, which is usually unilateral and of sudden onset. The patient is generally uncomfortable and has an enlarged, erythematous hemiscrotum on examination. If the pain is prolonged, there may be skin fixation to the underlying testicle (90). The testis generally lies horizontally, and in many cases, cremasteric reflex will be absent. In addition, pain is severe over the testis and spermatic cord. Many authors suggest surgical exploration for any child with suspected torsion. However, many emergency departments perform color Doppler imaging to assist in the diagnosis. Testicular scintigraphy can also be performed but has been replaced in many institutions by color Doppler ultrasound (97). One must be cognizant of the potential delay in diagnosis by performing ultrasonography or scintigraphy; therefore, surgical exploration is indicated for any patient whose history and physical examination are consistent with testicular torsion. At the time of surgery (Fig. 8 A,B) the testis is detorted and wrapped in a warm, moist sponge to allow for revascularization of the gonad. During this time, the contralateral testis should undergo orchiopexy because the incidence of asynchronous torsion may be as high as 50% (98). On completion of the contralateral orchiopexy, the ipsilateral torsed testis should be examined for viability.



Fig. 8 (A) Exploration of patient with severe hemiscrotal pain and physical examination results consistent with torsion. At exploration, a 720° rotation was noted. (B) Same testis after contralateral orchidopexy. The gonad did not demonstrate any flow and was removed.

If it is deemed viable, it is fixed to the scrotal wall. If it is nonviable, it is removed. Torsion of the appendix testis or the appendix epididymis occurs most often in boys around the time of puberty. The signs and symptoms are similar to those of testicular torsion; however, the pain tends to be more focal in location than in testicular torsion (90,97). Dresner described a "blue dot" sign that is considered to be pathognomonic for torsion of the appendix testis (99). The diagnosis can usually be confirmed by patient history, physical examination, and color Doppler sonography. Epididymitis is much less common in pediatric patients than in adults, but it can occur in all age groups. The onset of testicular pain is more gradual than in testicular torsion but can be just as severe. Color Doppler sonography will demonstrate good testicular blood flow, enlargement, and increased flow to the epididymis (100). The cause of epididymitis can be bacterial or inflammatory (as the result of reflux of sterile urine) (101). Treatment includes antibiotics and anti-inflammatory agents, scrotal support, and restriction of strenuous activity.

The list of differential diagnoses of scrotal masses is shown in Table 4. In the neonate, the differential diagnosis is limited and includes torsion, hydrocele, incarcerated hernia, testis tumor, trauma, and lesions resulting from a patent processus vaginalis. Although there is little debate regarding the existence of neonatal testicular torsion, there is controversy regarding the mechanism of torsion, the risk of bilaterality, and the need for exploration and contralateral testis fixation (102). Most cases of neonatal torsion are considered to be extravaginal, involving the testis, epididymis, and tunica vaginalis. The most common findings are a nontender, firm scrotal mass or a nonpalpable testis (in older children) (103). Color Doppler ultrasound can confirm torsion in the former. Meconium, hematoma, and tumor seeding from abdominal or adrenal tumors can be found in the scrotum as a result of a patent processus vaginalis (102,104). Testis tumors are rare but if found, should be explored surgically and resected as indicated. In older children, testicular tumors are rare but need to be considered in the evaluation of scrotal mass (102). Therapy (surgery, chemotherapy, and radiation) is individualized depending on the tumor. There is a role for testis-sparing surgery and for observation only for the more benign tumors (102). Varicoceles occur in up to 15% of adolescents (105). They are most commonly on the left and decompress when the patient is in the supine position. The indication for intervention is testicular growth failure. Treatment includes embolotherapy and surgical vein ligation. Kass demonstrated that when internal spermatic vein ligation was performed in children with testicular growth failure, greater than 75% had "catchup" growth of the affected testis (106).

Table 4 Scrotal Masses

Neonatal torsion Hydrocele Incarcerated hernia Spermatic cord torsion Testis tumor Epididymitis Paratesticular tumor Varicocele Epididymal cyst Epididymal tumor Idiopathic edema Schönlein-Henoch purpura Cavernous hemangioma Funiculitis Patent processus lesions

Priapism

Priapism can occur in all age groups. In most children, a hematologic disorder is the cause of priapism. The most notable cause of priapism in children is sickle cell disease (92). Priapism in this group of children may signal a sickle cell crisis, but more commonly it is an isolated event. This form of priapism is most commonly a low-flow or ischemic condition resulting from decreased venous outflow (107). The patient has a very hard, painful erection with a soft glans and soft corpus spongiosum. Initial treatment of this form of priapism includes oxygen supplementation and intravenous fluid hydration. In addition, analgesia should be provided. Priapism should be treated more aggressively if the erection does not subside. This includes aspiration and irrigation of the corporeal bodies, use of intracorporeal vasodilating agents, hypertransfusion therapy, and surgical treatment. As in adults, prolonged priapism in children may be associated with corporeal fibrosis and erectile dysfunction (108).

Urolithiasis

It is estimated that up to 3% of all urinary calculi occur in children (109). Renal calculus disease in children has a number of causes, including furosemide therapy, glucocorticoid therapy, relative hypophosphatemia, and low birth weight (110). Children with urolithiasis may have gross or microscopic hematuria and pain. Renal stone disease can be treated with observation except in patients who are symptomatic. Modes of therapy include extracorporeal shock wave lithotripsy, endoscopic stone manipulation, and open surgery (111).

PEDIATRIC URGENCIES

Hematuria

There are many causes of hematuria in children (Table 5) (112). Medical processes occur more frequently than surgical processes in this population. Lesions involving the glomerulus, renal interstitium, renal vascular supply, or urinary tract can all result in hematuria. An accurate diagnosis can be made by obtaining a thorough patient history and by performing a physical examination and urinalysis. One of the more common causes of hematuria is an acute postinfectious glomerulonephritis such as poststreptococcal glomerulonephritis. Children with poststreptococcal glomerulonephritis frequently have a history of illness 1 to 3 wk before the onset of hematuria. The patient may be fully recovered or may have malaise, headache, nausea, and vomiting. Urinalysis reveals red blood cell casts and proteinuria. Serum measures of renal function are most commonly normal (112). Antistreptolysin-O and anti-DNAase B titers may be elevated, confirming the diagnosis. Treatment is supportive, however, microscopic hematuria can persist for up to 2 yr. Other glomerular causes of hematuria include Schönlein-Henoch purpura, chronic glomerulonephritis, systemic lupus, Alport's syndrome, and benign familial hematuria. Benign familial hematuria is a very common cause of asymptomatic microscopic hematuria. Family history is helpful, and the physical examination and laboratory investigations are usually normal. Benign familial hematuria is passed by means of an autosomal-dominant route (84,112).

Renal interstitial hematuria may be caused by infection, medications, or toxins, or it may be metabolic, anatomic, or neoplastic. Pyelonephritis is a common cause of interstitial hematuria and resolves as the infection clears. Nephrocalcinosis resulting from idiopathic hypercalciuria is one of the most common metabolic causes of interstitial hematuria. Ultrasound evaluation of these patients demonstrates increased echogenicity, particularly in the medulla. Treatment is based on the underlying cause. The most common medications that cause interstitial hematuria are nonsteroidal anti-inflammatory drugs, and hematuria usually resolves after discontinuing the medication. The most common anatomic abnormality associated with hematuria is renal cyst disease. Treatment for renal cyst disease is, for the most part, supportive. Renal tumors were discussed previously in this section. The two main vascular causes of hematuria are sickle cell trait and disease and trauma. Children

Causes of Hematuria
Glomerular
Interstitial
Vascular
Genitourinary
Acute GN
Pyelonephritis
Trauma
UTI
Schönlein-Henoch purpura
Nephrocalcinosis
Renal vessel thrombosis
Urethrorrhagia
Systemic lupus
Nephrotoxins
Sickle cell trait/disease
Nephrolithiasis
Amyloidosis
Acute tubular necrosis
Nutcracker syndrome
Tumors

Table 5

GN, glomerulonephritis; UTI, urinary tract infection.

with sickle cell trait or disease have a 1% incidence of gross hematuria and approximately a 15% incidence of microscopic hematuria. Of note, the hematuria associated with sickle cell disease is usually painless and episodic. Sickle cell disease can lead to glomerular scarring and interstitial fibrosis, otherwise known as sickle cell nephropathy. Idiopathic urethrorrhagia, manifested by postvoid dripping of blood from the urethral meatus, blood spots in the underwear, and occasionally dysuria, are a particular concern to peripubertal males. Although the cause is unclear, the condition resolves spontaneously. Hematuria is generally a symptom that needs to be investigated but is generally self-limiting with no long-term sequelae (112).

Urinary Tract Infection

Children commonly are seen by pediatricians or in the emergency department with signs or symptoms or urinary tract infection. These include failure to thrive, fever, irritability, dysuria, enuresis, foul-smelling urine, and hematuria. It is estimated that up to 60% of neonates and up to 30% of older children who have a febrile urinary tract infection have some underlying genitourinary abnormality (113,114).

Therefore, development of a urinary tract infection requires treatment and follow-up in all children. Initial treatment will depend up the results of the urine culture. The child should continue antibiotic prophylaxis pending radiologic evaluation. This evaluation should include a renal/bladder ultrasound, VCUG, and renal scan (114). These radiographic studies should be deferred until the child is afebrile and has a normal urine culture. In the sick child, collection of urine via midstream or bag specimen should be avoided because it is unreliable. In these children, catheterization with a small-caliber catheter is the preferred manner of urine collection (115). Treatment of any genitourinary abnormality is based on the specific anomaly. The child should remain on antibiotic prophylaxis throughout the evaluation. Prophylaxis should be amoxicillin for the neonate less than 3 mo of age and preferably trimethoprim-sulfamethoxazole or nitrofurantoin for those children older than 3 mo.

Bladder Exstrophy and Cloacal Exstrophy

Bladder exstrophy (Fig. 9) occurs in approx 1 in every 10,000 to 50,000 live births (116). Most children with bladder exstrophy are diagnosed at birth, despite knowledge of antenatal sonographic findings (117). Associated findings include a wide pubic diastasis, a high incidence of inguinal hernia, and an approx 90% incidence of VUR after bladder closure (Fig. 10). With the exception of the exstrophied bladder, these children are healthy. Before the 1970s, treatment was usually urinary diversion. However, beginning in the 1970s, Jeffs and others began to perform staged reconstruction for these children (118). Today, the bladder can remain intact in most of these children. These patients should be cared for in a center of excellence, where exstrophy reconstruction is common. Bladder reconstruction is highly successful, as measured by continence interval and the ability of these children to later have satisfactory sexual relations. The most popular method is a staged reconstructive strategy (119). This involves bladder and abdominal wall closure, with osteotomy in children over 72 hr old (Fig. 11). In males, the epispadias is usually repaired at about 1 yr of age. When the child is ready to achieve continence, bladder neck reconstruction with ureteral reimplantation is performed. An alternative method of reconstruction combines the first two stages into one operation, as described by Grady and Mitchell (120). If adequate bladder capacity fails to develop either after primary or secondary reconstruction, bladder augmentation



Fig. 9. Classic bladder exstrophy in newborn male.

with or without bladder closure and a catheterizable abdominal stoma is the best alternative (121).

Cloacal exstrophy (Fig. 12) occurs in approx 1 in every 250,000 to 400,000 live births (77,122). In addition to the abdominal wall and bladder defects seen in classic bladder exstrophy, omphalocele (95%) and exstrophy of the large bowel, which is sandwiched between two hemibladders, are also present. In addition, there is usually prolapse of the ileum. Up to 75% of these children can also have associated neural tube defects (Fig. 13) (122). Reconstruction is extremely difficult and requires a multidisciplinary plan; however, many children can achieve continence with this approach (123). An attempt should be made to save as much bowel as possible at the time of bladder and omphalocele closure. Males may also need to undergo gender reassignment as a result of the markedly shortened and separated erectile bodies (122).

Myelodysplasia

One of the most fascinating groups of children to follow is that group born with myelodysplasia (Fig. 13). These children can demonstrate a full spectrum of neurogenic bladder dysfunction (124). With the advent of routine prenatal ultrasound and maternal folic acid supplementation, the incidence of children born with myelodysplasia has decreased. Myelodysplasia was once one of the more common causes of neuro-



Fig. 10. Sleep cystogram in child who underwent prior bladder closure. There is high-grade reflux bilaterally.

genic bladder. Once the spinal cord defect has been closed, the genitourinary tract should be evaluated. This includes the use of renal/bladder ultrasound, VCUG, and urodynamics testing. Up to 5% of children with myelomeningocele have VUR. Urodynamics testing is important to rule out sphincter dyssynergia and uninhibited bladder contractions. This may allow the urologist to predict which patients may have upper tract deterioration in the future (124). In addition, postvoid urine should be measured; the results may prompt voiding by Crede's method or by clean intermittent catheterization.



Fig. 11. Child after bladder exstrophy repair and bilateral osteotomies. Use of a fixation device has replaced the use of modified Buck's traction for these children.



Fig. 12. Newborn female with cloacal exstrophy. Note the omphalocele and disproportionately sized hemi-bladders situated on either side of a portion of bowel.



Fig. 13. KUB of child with cloacal exstrophy. There is a wide pubic diastasis. In addition, the child has spinal dysraphism.

This population of patients is interesting because of the changes that occur in bladder dynamics as the child grows. Spinal cord growth and possible tethering can determine changes in bladder function. Normal reflex voiding, dyssynergic voiding, flaccid bladder, and a spastic bladder may occur in the same child over time (125). It is crucial to follow these patients throughout their childhood. Management of these patients can include observation, clean intermittent catheterization, and the use of anticholinergic medications. Patients with lipomeningocele or sacral agenesis should undergo similar evaluation and follow-up.

Imperforate Anus

This group of children has a form of a cloacal lesion that can be as mild as anal stenosis, requiring only dilation, or as serious as supralevator lesions with rectovaginal or rectocloacal fistulae in females and rectourethral or rectovesical fistulae in males (122).

In 1987, McLorie et al. presented their experience with 484 patients with imperforate anus. They found that the incidence of genitourinary abnormalities was 60% in high lesions and 20% in low lesions (126). The most common genitourinary abnormalities were VUR, renal agenesis, renal dysplasia, neurogenic bladder (as a result of either a concomitant vertebral anomaly or bladder denervation that occurred during rectal pull-through), and cryptorchidism (127,128). The evaluation of these children should always include a renal and bladder ultrasound to evalu-

ate the upper and lower tracts, VCUG looking for the location of the fistula and for the presence of VUR, and urodynamic testing for any patient with a supralevator lesion or evidence of vertebral anomalies. Many of these children are treated emergently with fecal diversion, with reconstruction occurring later. DeVries and Pena originally described the treatment of the imperforate anus in 1982 (129). These authors pioneered a posterior sagittal approach that is still the preferred method of rectal pull-through. In addition, patients undergo fistulectomy at the time of the pull-through procedure.

Prune-Belly Syndrome

Prune-belly syndrome, also known as Eagle-Barrett syndrome and triad syndrome, is manifested by deficient or absent abdominal wall musculature, urinary tract dilation, and cryptorchidism (Fig. 14) (130). This syndrome occurs almost exclusively in males, with a female variety being described by Hirose in 1995 (131).

There is a wide spectrum of this syndrome, ranging from normal renal function to in utero death. This syndrome has three broad categories. Children with urethral obstruction, oligohydramnios, and Potter's syndrome-type facies comprise the first group. Death in this group is usually the result of pulmonary hypoplasia. The second group is characterized by mild renal impairment, although renal failure may develop in the future. The last group has normal renal function (132). Urologic evaluation of children with prune-belly syndrome should include renal and bladder ultrasound, VCUG, and furosemide renal scan to assess differential function and adequacy of upper tract drainage. About 50% of patients have a patent urachus. These children should be started on antibiotic prophylaxis at birth. Treatment is individualized; however, there is little debate regarding the need for bilateral orchiopexy. This can be accomplished either laparoscopically or by an open procedure (133). Despite early orchiopexy, these children are unable to father children. However, there is evidence to suggest that there are spermatogonia in the testes of these children, opening the possibility of assisted reproductive techniques for this population (134). Other surgical treatments described include abdominoplasty, reduction cystoplasty, and bilateral ureteral reimplantation.

Urethral Prolapse

Urethral prolapse is found predominantly in young African-American females. Often treatment is sought because of vaginal bleeding or pain. Occasionally these children will be referred to the gynecologist. On examination, these girls have protrusion of the urethral mucosa, and



Fig. 14. VCUG of a child with prune-belly syndrome. Note the diffuse dilation of the lower genitourinary tract, which is part of the classic triad.

the mucosa will most often be hyperemic and friable. Conservative treatment with local estrogen cream and sitz baths usually allows complete resolution. However, for those girls who do not respond to conservative therapy, surgical excision of the prolapsed mucosa is curative. The differential diagnosis of an introital mass includes ureterocele prolapse and sarcoma botryoides; therefore, careful inspection is warranted to confirm the diagnosis of urethral prolapse (*35,101*).

SUMMARY

This section has been devoted to an overview of conditions that require emergent or urgent evaluation and treatment. Often pediatric urologists are called on to evaluate these children because emergency medicine physicians, pediatricians, or parents perceive an emergency. It behooves us to see these children, begin the appropriate evaluation, reassure the consulting physicians, and allay the fears of the parents and children. The majority of the conditions discussed will be found in greater detail in other sections.

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