PERSPECTIVE

This chapter discusses some of the more common causes of scrotal, testicular, and penile pain in children that precipitate a visit to the emergency department (ED). Also discussed are other renal or genitourinary tract disorders in this age group for which emergent treatment may be sought, including acute renal failure, urinary tract infections, and hypertension.

SPECIFIC DISORDERS

Penile Disorders

Priapism

Principles of Disease. Priapism is the engorgement of the dorsal corpora cavernosa, resulting in dorsal penile erection and ventral penile flaccidity. The more common low-flow priapism is secondary to decreased venous outflow and is characterized by prolonged painful erection. Sickle cell disease and leukemia are responsible for the majority of cases in children. Immunosuppressive disorders, anticoagulation, and intracavernosal injection of agents such as papaverine, phentolamine, and prostaglandin E, also can result in priapism. Other drugs, such as phenothiazines, sedative-hypnotics, selective serotonin reuptake inhibitors, antihypertensives, anticoagulants, and drugs of abuse (e.g., cocaine, alcohol, marijuana), may be causal. Priapism complications include penile fibrosis, urinary retention, and impotence.

High-flow priapism usually is painless and typically is associated with penile arterial laceration and excessive inflow of arterial blood or spinal trauma resulting in corporal engorgement. With prolonged engorgement of the corpora cavernosa, the resultant stagnation and hypoxia of the blood lead to the development of thrombosis and ischemia.

Clinical Features. Priapism is a clinical diagnosis and can be distinguished from other causes of erection by careful history and complete physical examination. The history should include past medical history with attention to previous treatment of anemia, leukemia, sickle cell disease, or drug abuse and current history of trauma or symptoms of immunosuppressive disease. Although priapism is a clinical diagnosis, laboratory studies may assist in isolation of the cause of the condition. A complete blood count (CBC), urinalysis, and coagulation studies may be useful in some cases. Diagnostic studies of penile blood flow may be indicated in cases for which the cause is unclear. Studies that the urologist may request include magnetic resonance imaging, color Doppler cavernosography, and technetium Tc-99m penile scanning. Angiography is helpful in localization of the arterial bleeding site in high-flow priapism.

Differential Considerations. The differential diagnosis for priapism in children differs from that in adults. In adults, penile erection from sexual arousal, urethral foreign bodies, Peyronie’s disease, spinal cord injury, and penile implants may occur, but these etiologic factors are rare in children. Anticoagulation, drugs of abuse, medications, trauma (including spinal trauma), Kawasaki disease, leukemia, and sickle cell disease are associated with priapism in children.

Management and Disposition. Management centers on hydration, pain control, relief of urinary obstruction, and treatment of any other underlying conditions. Local anesthesia by a dorsal nerve block with 1% lidocaine without epinephrine may be beneficial. In patients with sickle cell disease, treatment with oxygen, hydration at 1.5 times maintenance volume, and analgesics should be initiated. Low-flow priapism may respond to sitz baths or hot compresses because the heat increases blood flow, thereby potentially relieving the obstruction. Oral terbutaline (adult dose 5-10 mg, followed by another 5-10 mg 15 minutes later) produces resolution in about one third of patients. If no resolution occurs within 30 minutes of treatment, aspiration and irrigation therapy is required.

Cavernosal aspiration plus irrigation has been effective in patients with low-flow priapism. This procedure should be performed within the first few hours of symptom onset and rarely is beneficial after 48 hours. Vasoactive substances such as phentolamine and phenylephrine may be used for irrigation therapy; however, these are best administered with urologic consultation. Parenteral vasodilators, such as methylene blue, hydralazine, and terbutaline, also have been tried with variable success, and therefore the treating urologist can help ascertain the best treatment approach.1-4

Exchange transfusion for patients with priapism and sickle cell disease previously has been recommended; however, this treatment may not offer any advantage over conventional therapy and has been associated with serious neurologic sequelae, as in the so-called ASPEN syndrome (association of sickle cell disease, priapism, exchange transfusion, and neurologic events).2,3 Patients with leukemia may respond with detumescence after chemotherapy is begun.

High-flow priapism can be effectively addressed by arterial embolization. Studies have shown success with the use of autologous blood clot embolization.4 If nonsurgical approaches are unsuccessful, referral to a urologist for a corpus cavernosum–glans shunt procedure may be necessary. Ultrasound-guided compression of the perineal arterioca cavernous fistula has been successful.5 Interventions should be initiated within 12 hours of symptom onset to avoid long-term dysfunction and irreversible infarction.6 Patients with persistent priapism or underlying disorders, such as leukemia or sickle cell
disease, may require hospitalization. Pediatric urologic consultation should be arranged as soon as possible. If the priapism has been treated successfully, the patient may be discharged home after some period of observation with urologic specialist follow-up.

Phimosis

Principles of Disease. Phimosis is a pathologic condition of the uncircumcised penis in which constriction of the foreskin prevents retraction of the prepuce from the glans. It can result in pain, hematuria, glans ischemia, infarction, and urinary outlet obstruction. Most cases are physiologic, representing normal development, and do not require intervention. Only 4% of newborn boys have a fully retractable foreskin. This percentage increases with age: 25% of 6-month-old, 50% of 1-year-old, 80% of 2-year-old, and 90% of 4-year-old boys have fully retractable foreskins. Phimosis also may result from trauma, infections, chemical irritation, poor hygiene, and congenital abnormality or be a complication of circumcision.

Clinical Features. Phimosis is a clinical diagnosis. History may reveal that the foreskin is unretractable, along with narrowing or diversion of the urinary stream and bulging out of the foreskin with urination. Pain and hematuria may be accompanying features.

Management and Disposition. Because the ability to retract the foreskin fully is age related, parents should be advised not to retract the prepuce forcefully. Gentle retraction with good hygiene should be stressed. If signs of urinary outlet obstruction are present, dilation of the prepuce or the urethral meatus or both can be performed by gentle use of forceps. With vascular compromise of the glans, a dorsal split procedure, circumcision, preputial plasty, or balloon dilation may be necessary. Topical steroid treatment (with betamethasone valerate 0.6% cream) for 6 weeks has been 87% effective in reducing inflammation and treating phimosis. In patients with severe stenosis, obstructive uropathy can result. In such rare cases, blood urea nitrogen (BUN) and serum creatinine levels should be obtained, and a renal ultrasound examination should be performed if signs of obstructive renal failure are present. Patients who are able to urinate and have no evidence of severe infection or ischemia can be discharged from the ED with outpatient urologic follow-up.

Paraphimosis

Principles of Disease. Paraphimosis is another pathologic condition of the uncircumcised penis in which the proximal foreskin cannot be returned to its anatomic position covering the glans penis, resulting in distal venous congestion with the potential for dire consequences. Paraphimosis can be caused by infection, masturbation, trauma, or hair or clothing tourniquets. Iatrogenic causes include failure to reduce the foreskin after a medical examination. Paraphimosis constitutes a true urologic emergency and can result in arterial compression, penile necrosis, and gangrene.

Clinical Features. The patient typically is anxious, and the history often reveals that the parents or the patient retracted the foreskin and then could not replace the foreskin over the glans (Fig. 174-1). The history should include verification that the patient is uncircumcised because hair tourniquet syndrome in a circumcised patient may mimic paraphimosis. Physical examination reveals a flaccid proximal penis with erythema and engorgement distal to the obstruction. The foreskin is retracted, and cellulitis may be present. The diagnosis of paraphimosis is based on clinical findings, but if a penile foreign body is a concern, radiographs can be obtained after relief of the vascular occlusion.

Management and Disposition. Pain can be controlled either parenterally or by a local dorsal penile nerve block. Procedural sedation also may be necessary. Placement of a finger of a rubber glove filled with ice water over the glans and foreskin can reduce the edema. Circumferential compression of the penis starting at the glans also can reduce the edema. Compression may need to be held for several minutes to achieve adequate reduction of edema. Manual reduction may be necessary, with application of gentle, steady pressure on the glans with both thumbs while the shaft is pulled straight (Fig. 174-2). Another method is to puncture the edematous foreskin with an 18- or 21-gauge needle. The puncture may be followed by squeezing of the glans penis to further facilitate fluid drainage. Dorsal band traction has been performed in some cases with Adson forceps applied directly to the band formed by the retracted foreskin and application of traction and countertraction to loosen the constriction. If all such attempts fail, urologic consultation proceeding to circumcision or a dorsal slit procedure may be necessary. Patients can be discharged home after reduction if they are able to void spontaneously and urologic follow-up care can be arranged. Any evidence of cellulitis or necrosis warrants hospital admission, intravenous antibiotics, and urologic consultation.

Balanoposthitis

Principles of Disease. Balanoposthitis, an inflammation that involves the glans and the foreskin, occurs in up to 6% of
uncircumcised males. Balanitis involves the glans penis only. The primary cause of balanoposthitis is infection; however, chemical irritation, trauma, fixed drug rash, and contact dermatitis also can be contributory. Infectious organisms are gram negative and gram positive, including group A beta-hemolytic streptococci and rarely Neisseria gonorrhoeae and Chlamydia. Recurrent episodes of Candida albicans balanoposthitis should raise suspicion for diabetes mellitus.

Clinical Features. Physical examination reveals penile erythema, edema, and possibly a discharge (Fig. 174-3). Systemic symptoms and signs such as fever, vomiting, and diarrhea are unusual. Diagnosis is clinical and treatment is based on clinical findings; however, if symptoms are severe, the offending organism can be identified by culture of any discharge.

Management and Disposition. Management includes emphasis on adequate hygiene with sitz baths to reduce inflammation. Painful micturition can be addressed by having the child urinate while in a bath of warm water. In patients with cellulitis, antibiotic treatment with agents effective against Staphylococcus aureus and Streptococcus pyogenes is recommended. Seven days of a first-generation cephalosporin is an appropriate regimen. If group A beta-hemolytic streptococci are identified, a streptococcus-specific antibiotic is required. Severe inflammation may be treated with 0.5% hydrocortisone cream applied sparingly to the area. Candidal infections should be treated topically with antifungals. A blood glucose determination should be considered in patients presenting with recurrent candidal balanoposthitis. Patients with a toxic appearance or those who exhibit evidence of cellulitis or constricting phimosis with uropathy should be hospitalized for appropriate management. Circumcision may be required for patients with recurrent disease.

Complications of Circumcision

Principles of Disease. Although there remains societal controversy about circumcision, it is usually advocated for prevention of phimosis, paraphimosis, recurrent balanoposthitis, urinary tract infections, and penile cancer. Any of three techniques can be used: application of a Plastibell or Gomco clamp, excision, or dorsal slit procedure. The choice of procedure depends on the preference of the operator. The most common complication of these procedures is hemorrhage, which usually is minor and can be controlled by direct pressure, silver nitrate application, or suture placement. Significant bleeding may be a sign of a blood dyscrasia. Localized, systemic, or urinary tract infection also can occur.

Management and Disposition. Postoperative pain usually resolves within 12 to 24 hours. Occlusive dressings can contribute to urinary retention and edema and should be removed. Stenosis of the now exposed urethral meatus may result from prolonged exposure to the ammonia in urine. Application of a Plastibell that is too small also can lead to meatal stenosis in 8 to 31% of cases. Signs and symptoms include pain with urination, bloody discharge resulting from an inflamed meatus, high-velocity stream, and the need to sit while voiding. Postcircumcision phimosis may result if excess foreskin remains. If the constriction is severe and urinary outlet obstruction occurs, dilation of the stenosis can be performed with a hemostat. Surgical revision usually is necessary.

Skin bridges are small fibrotic bands that attach the glans to the penile shaft and may form after circumcision. Inclusion cysts result from smegma retained in the wound or from epidermis involution along the circumcision site. Skin bridges and inclusion cysts are treated by surgical resection. These complications may be prevented by proper hygiene and application of an antimicrobial ointment to the circumcision site for 7 to 10 days after the procedure.

Penile Entrapment and Tourniquet Injuries

Penile rings, string, wire, and human hair tourniquets can result in penile venous and arterial occlusion. The patient presents with swelling of the glans, wherein the offending agent may be difficult to visualize because of edema of the coronal sulcus. In addition to the vascular supply, the dorsal penile nerve supply can be occluded. The foreign body can be cut with small surgical scissors. Hair tourniquets have also been successfully removed with Nair. Once the foreign body is removed, urethral obstruction can be evaluated by means of retrograde urethrogram, and Doppler ultrasonography can be performed to assess penile arterial blood flow. Once it is identified, the constriction is relieved, and the patient should not be discharged until spontaneous voiding is ensured. Urologic consultation may be necessary emergently if penile arterial flow is disrupted and the constricting object cannot be removed rapidly or signs of necrosis are present.

Zipper entrapment of the foreskin also can occur in children, typically those between 2 and 6 years of age. The zipper can be removed with bone or metal cutters or a mini-hacksaw to cut the median bar of the zipper (Fig. 174-4). The zipper falls apart, and the foreskin is freed. Although local anesthesia is not necessary, the patient may be anxious and require sedation before its
removal. Success has been reported with soaking of the penis in mineral oil before zipper removal. Additional methods to release the foreskin under the zipper mechanism include cutting of the zipper below the entrapment and pulling the two halves of the zipper apart; cutting of the zipper teeth above and below the entrapment and, with pliers, squeezing the median bar to allow more room to disengage the trapped prepuce; and insertion of a flat screwdriver between the faceplates of the zipper mechanism to pry open the faceplates and allow the prepucce to be released.

Parents should be instructed to encourage their children to wear underwear to decrease the risk of entrapment.

Scrotal Masses and Swelling

Epididymis

Principles of Disease. Epididymitis is inflammation of the epididymis, which is located along the posterior aspect of the testicle. The most common cause is infectious, and the etiology varies by age. Adolescents should be evaluated for possible sexually transmitted diseases, such as N. gonorrhoeae and Chlamydia trachomatis. Patients may have a history of previous urinary tract infections, anatomic abnormalities, or previous genitourinary instrumentation. Urinary tract infections leading to epididymitis typically are caused by viruses or bacterial agents, such as Escherichia coli, Klebsiella pneumoniae, Proteus, or Pseudomonas aeruginosa.

Clinical Features. Patients present with a painful, edematous scrotum and tenderness at the epididymis (Fig. 174-5). A urethral discharge may be present, particularly when the condition is secondary to a sexually transmitted disease. Systemic signs and symptoms, such as nausea, vomiting, fever, and lower abdominal, scrotal, or testicular pain, also may be present. Infants and young children may present with fever without other symptoms. Accordingly, in the ED evaluation of all children with fever or other systemic symptoms or signs, a complete examination should include a genitourinary examination.

As the edema increases, obliteration of the sulcus between the testis and epididymis occurs, making differentiation from testicular torsion extremely difficult. Relief of pain with scrotal elevation (Prehn’s sign) is unreliable.

Diagnostic Strategies. Urinalysis and urine culture should be part of the ED evaluation for all infants and children younger than 2 years. In children 2 years of age or older, urine culture may be performed only if the urinalysis results indicate urinary tract infection. Lack of pyuria does not rule out epididymitis because up to 50% of patients may have normal results on such studies. CBC may show leukocytosis, although this finding is nonspecific. Any urethral discharge should be cultured and sent for Gram’s stain and tests for N. gonorrhoeae and C. trachomatis. Color Doppler ultrasonography is not necessarily performed in all children with suspected epididymitis. However, if there is doubt about the diagnosis, it is the preferred diagnostic study because it does not require placement of an intravenous line and is more readily available at all hours of the day. If color flow Doppler ultrasonography or radionuclide scintigraphy is performed, it will reveal a normal testis and preserved or increased vascular flow toward the side of the inflamed epididymis.

Management and Disposition. Scrotal elevation, placement of ice packs on the swollen area as tolerated, and nonsteroidal anti-inflammatory or narcotic medications are useful to control pain and inflammation. If urethral discharge is present, the adolescent patient should be treated presumptively for both N. gonorrhoeae and C. trachomatis. The treatment for sexually acquired epididymitis in children 9 years of age or older consists of ceftriaxone 250 mg intramuscularly, followed by doxycycline 100 mg orally twice a day or tetracycline 500 mg orally four times a day. A single 1-g dose of azithromycin may also be used to increase compliance. Patients younger than 9 years should be treated with erythromycin 50 mg/kg orally per day, divided four times a day. The course of treatment typically ranges from 7 to 14 days, depending on the antibiotic regimen. In children, non–sexually acquired epididymitis without evidence of urinary tract infection may be managed expectantly with analgesics for pain.

Infants with or without positive findings on urinalysis and young children with positive urine analysis findings may be treated with trimethoprim twice a day or cephalaxin three times a day if a bacterial urinary tract infection is suspected. Other antibiotic options include oral erythromycin, clarithromycin, and azithromycin for patients who do not respond to other therapies because Mycoplasma genitalium and Ureaplasma urealyticum may cause a more chronic form of epididymitis. Infants and children with epididymitis may be discharged for follow-up by their primary care physician, and urine cultures should be checked to determine definitively if urinary tract infection is present. Patients with systemic symptoms and toxicity should be admitted for intravenous antibiotic therapy with either ceftriaxone or cefotaxime. Children need close urologic follow-up to rule out any contributing anatomic abnormalities and to ensure that the antibiotic chosen treats the infecting organism.

Orchitis

Orchitis is a result of a bacterial or viral testicular infection leading to diffuse edema, pain, and discoloration of the scrotum. The most common pathogen is paramyxovirus, which can be the causative agent in up to 38% of postpubertal males with mumps-associated orchitis. Other etiologic agents include E. coli, K. pneumoniae, P. aeruginosa, Staphylococcus or Streptococcus species, Epstein–Barr virus, Coxsackievirus, arboviruses, enteroviruses, Brucella, granulomatous disease, and filariae.

During the course of mumps, orchitis usually develops after the first week and is manifested with tenderness and edema of the testis with discoloration of the scrotum. Bilateral cases are relatively rare, occurring in 2 to 5% of affected patients. If epididymitis coexists, a urethral discharge also may be present. Bacterial orchitis can result in scrotal abscess formation. Because orchitis typically is unilateral, fertility usually is maintained.

Doppler ultrasound imaging may be necessary to distinguish orchitis from testicular torsion. For patients with a clearly viral origin such as mumps, treatment is aimed at pain control (scrotal elevation, nonsteroidal anti-inflammatory agents, and possibly narcotics). When the diagnosis is unclear or when concurrent epididymitis is present, empirical treatment includes oral antibiotics with predominantly gram-negative organism coverage. Hospital admission is warranted for patients who have a toxic appearance or exhibit evidence of a scrotal or testicular abscess or in whom...
the skin of the inner thigh downward from the hip toward the knee. This causes the cremaster muscle on the ipsilateral side to contract rapidly and results in elevation of the testicle. Although in one series the cremasteric reflex was absent in 100% of patients with torsion and in only 14% of patients with epididymitis, the presence of the cremasteric reflex does not preclude testicular torsion. Abnormal epididymal and testicular position also may be noted, with left-sided torsions slightly more common than right. Nausea, vomiting, and a low-grade fever also may be seen. In the patient with an undescended testicle who presents with abdominal pain, torsion should be a consideration.31

Diagnostic Strategies. Unless an alternative diagnosis is absolutely secure (e.g., epididymitis), the patient should be evaluated for testicular torsion. Alternatively, with a relatively confident clinical diagnosis of torsion, diagnostic testing should not delay appropriate management. Results of a urinalysis are rarely helpful as pyuria can be seen in cases of testicular torsion and epididymitis. The widely preferred diagnostic study for the prospect of torsion is color flow Doppler ultrasonography. This technique has a sensitivity of 79 to 86%, with a specificity of almost 100%, for detection of testicular torsion. Scintigraphy has a sensitivity ranging from 79 to 100% and a specificity of 89 to 100%.27,32,33 Magnetic resonance imaging has been evaluated as another diagnostic modality, with a sensitivity in one study of 93% and specificity of 100%.34 In cases of indeterminate ultrasound findings, the urology consultant should be notified for disposition decisions; in addition, scintigraphy may be performed as resources allow. When clinical suspicion is strong, surgical exploration should not be delayed for diagnostic studies, especially in patients in whom duration of symptoms is less than 12 hours.

Management. If the patient presents within 12 hours of symptom onset, immediate surgical exploration is indicated.

Testicular Torsion

Principles of Disease. Torsion of the spermatic cord is a common cause of an acutely painful scrotum. Delay in diagnosis and treatment can result in loss of spermatogenesis and, in severe cases, necrotic, gangrenous testes. Testicular salvage rates are time dependent; the success rate is 96% if detorsion is performed less than 4 hours from symptom onset, decreasing to less than 10% if there is more than a 24-hour delay to treatment.29 The overall incidence of testicular torsion is 1 in 4000, with a peak incidence at age 13 years. Testicular torsion has been reported in all age groups, from the developing fetus to the elderly, but is most common in adolescence.

The testis enters the scrotum through the inguinal canal after descent from the abdomen. The peritoneum invaginates through the canal and partially covers the testis and epididymis, forming the tunica vaginalis. Typically, the tunica vaginalis attaches to the posterior wall of the hemiscrotum and superior pole of the testis to achieve testicular fixation. 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 (Fig. 174-6). In intravaginal torsion, the testicle may rotate within the tunica vaginalis, thereby constricting the arterial blood flow. Extravaginal torsion is seen most commonly in neonates who are premature and also can occur antenatally.

Clinical Features. Patients present with acute scrotal pain and swelling, an elevated testicle, and, typically, absence of the cremasteric reflex.20 This reflex can be demonstrated by lightly stroking the skin of the inner thigh downward from the hip toward the knee. This causes the cremaster muscle on the ipsilateral side to contract rapidly and results in elevation of the testicle. Although in one series the cremasteric reflex was absent in 100% of patients with torsion and in only 14% of patients with epididymitis, the presence of the cremasteric reflex does not preclude testicular torsion. Abnormal epididymal and testicular position also may be noted, with left-sided torsions slightly more common than right. Nausea, vomiting, and a low-grade fever also may be seen. In the patient with an undescended testicle who presents with abdominal pain, torsion should be a consideration.31

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Management. If the patient presents within 12 hours of symptom onset, immediate surgical exploration is indicated,
predicated on clinical findings or confirmatory clinical studies. Detorsion of the affected testicle is performed, followed by an elective orchietomy of the contralateral side to avoid recurrence. Approximately 40% of patients have a bell clapper deformity of the contralateral testicle. Manual detorsion also may be performed by rotation of the testicle in an “open book” fashion as viewed from below, from medial to lateral, until detorsion is complete.

Because the procedure is painful, sedation and pain relief should be considered before this intervention. Manual detorsion should be attempted only if there is a delay in getting a urologist to come in for operative detorsion and the patient has had continuous pain for less than 24 hours or if, in the judgment of the emergency physician, the time zone of opportunity to salvage the testis has not passed.

**Torsion of the Testicular Appendage**

The appendix testis is a remnant of the müllerian duct. The appendix testis is involved in 92% of the cases of testicular appendage torsion, and the appendix epididymis is involved in the remaining cases. The average age at occurrence is 10 years. Patients typically present with moderate pain of sudden onset that is localized to the involved hemiscrotum. The pathognomonic “blue dot” sign, a small area with a bluish hue less than 3 mm across located in the upper lateral portion of the hemiscrotum, is present in less than 25% of cases and represents the cyanotic appendage below the scrotal wall. If any doubt exists about the diagnosis, a nuclear radioisotope scan or color Doppler ultrasound study should be performed. Color Doppler ultrasonography reveals normal or increased flow to the affected testicle. Conservative therapy with analgesics and scrotal support are usually all that is indicated; the involved appendage undergoes autoamputation within 1 week, accompanied by resolution of symptoms.

**Varicocele**

A varicocele is a collection of varous venous collaterals of the spermatic veins in the scrotum caused by incomplete drainage of the pampiniform plexus. Incidence rates of 14 to 16% in adolescent males have been reported, but varicoceles are rare in children younger than 10 years. Left-sided varicoceles account for 85 to 95% of cases; however, bilateral varicoceles may be present in up to 22% of patients. Intra-abdominal disease should be suspected in cases of right-sided varicocele because these usually are caused by inferior vena cava thrombosis or compression of this vessel by tumors. The acute presentation of a left-sided varicocele should raise suspicion for renal cell carcinoma with obstruction of the left renal vein.

The dilated venous collection may be tender on physical examination. It can be palpated superior and posterior to the testis and usually is more pronounced with the patient in the upright position. Therefore, the patient should be examined in both the standing and supine positions. Patients in whom scrotal swelling persists in the supine position should be evaluated with a computed tomography (CT) scan of the abdomen, with oral and intravenous contrast administration for conditions obstructing the renal vein. Varicoceles have been described as a “bag of worms” both in appearance and on palpation. Surgical correction may be required if the patient becomes symptomatic or has bilateral varicoceles.

**Idiopathic Scrotal Edema**

Idiopathic scrotal edema is painless erythema and induration of the scrotum; 77% of cases occur before the age of 10 years. Two thirds of cases are unilateral, and no specific causes have been identified. The condition is characterized by the development of painless erythema and induration of the scrotum, which may be pruritic. There is minimal tenderness on physical examination, but the edema and erythema may extend to the phallus, groin, and abdomen. Examination of the testes and epididymis reveals no palpable masses. Systemic signs and symptoms are rare. Patients can be discharged home with outpatient follow-up after an acute pathologic process has been ruled out. Most cases resolve spontaneously within a few days and do not require any specific treatment. Recurrence rates of up to 21% have been described.

**Hydrocele**

A hydrocele is a collection of fluid that accumulates in the tunica vaginalis. Communicating hydroceles result when the upper processus vaginalis fails to be obliterated, leaving an open tract between the peritoneum and the scrotum. The tract is closed in noncommunicating hydroceles. Most hydroceles are right sided. They may be present at birth, but they usually are painless and worsen with crying or exertion. Hydroceles often resolve spontaneously by the age of 18 months. Examination with transillumination reveals enlargement of the scrotum. Color flow Doppler ultrasonography or radionuclide scintigraphy may be necessary to determine the cause of the hydrocele and to exclude an acute pathologic process in patients with acute symptoms of pain; otherwise, asymptomatic patients can be discharged home with urologic follow-up. Patients with a hydrocele that has persisted for more than 1 year or who are older than 18 months should undergo ultrasonography to ensure that the hydrocele is not a reactive hydrocele caused by testicular tumor or inflammation.

**Inguinal Hernia**

**Principles of Disease.** Inguinal (direct and indirect) hernias are more common in males, with bimodal peaks before 1 year of age and then again after the age of 40 years. An indirect inguinal hernia occurs when the processus vaginalis is not obliterated in infancy and abdominal contents invaginate through this patent sac. Entrapment of mesentery, bowel, intraperitoneal organs, and the hernial sac can occur and is more common with small hernias. If the contents of the hernia can be returned to their anatomic position, the hernia is reducible; if the contents remain entrapped, it is incarcerated or irreducible. Hernias that remain incarcerated can undergo strangulation, with resultant necrosis of bowel or mesentery.

**Clinical Features.** Patients with incarcerated hernias may present with pain, edema extending to the scrotum, nausea, vomiting, and low-grade fever. Physical examination may reveal bowel sounds in the scrotal sac. If the inguinal mass can be palpated separately from the testes, it is possible to diagnose an inguinal hernia on clinical grounds alone. Rarely, the incarcerated and strangulated hernia can be manifested as a tense blue mass in the scrotum.

**Management and Disposition.** The patient should be placed in the Trendelenburg position with an ice pack applied to the groin to reduce swelling; sedation may be necessary before reduction. Slow, gentle pressure should be applied to reduce the hernia. If the pressure technique is not successful in reducing the hernia, the opposite technique can be tried: the hernia mass can be “pulled” to straighten out the entrapped contents, thereby allowing them to slip back into the abdomen.

If the hernia cannot be reduced or strangulation is suspected (as indicated by fever, overlying cellulitis, or signs of peritonitis), the patient should receive fluid resuscitation, broad-spectrum parenteral antibiotics, and an emergent surgical consultation.

Patients who are found to have a hernia on routine examination or who have had the hernia reduced and are without symptoms suggestive of incarceration or strangulation should be referred for surgical repair.
Carcinoma

Principles of Disease. Testicular and scrotal cancer represents approximately 1% of solid tumors in children. An increased incidence of testicular cancer, in both the undescended testicle and the contralateral descended testicle, has been noted in patients with cryptorchidism. Tumor types include teratomas, embryonal carcinomas, yolk sac tumors, choriocarcinomas, Leydig cell tumors, and Sertoli cell tumors. Lymphoma and leukemia can metastasize to the testicle as well.

Patients typically present with a painless unilateral mass palpated separately from the testis or may describe a feeling of fullness, tugging, or increased weight of the scrotum and testicular enlargement. A reactive hydrocele may be present in 7 to 25% of patients and can lead to a delay in diagnosis. Physical examination reveals a firm mass, smooth or nodular, that cannot be tran-illumination. A complete physical examination looking for lymphadenopathy, petechiae, abdominal mass, hepatosplenomegaly, or gynecomastia should be performed.

Diagnostic Strategies. Diagnostic evaluation includes a CBC, urinalysis, urine human chorionic gonadotropin (produced by germ cell tumors) assay, and ultrasonography of the testis.

Management and Disposition. ED management includes prompt referral for urologic and oncologic consultation. Admission to the hospital to facilitate the evaluation may be necessary.

Urinary Tract Infections

Perspective

Sequelae of untreated urinary tract infections may include sepsis and renal scarring, and accurate and early diagnosis is important. At the same time, avoidance of unnecessary evaluation and treatment of children at lower risk for these infections is cost-effective and minimizes the chance for iatrogenic harm. Diagnosis of urinary tract infections in infants and young children can be challenging because the clinical signs often are nonspecific, and it can be difficult to obtain useful urine specimens.

Principles of Disease

The risk for development of urinary tract infections before the age of 12 years is approximately 3% for girls and 1% for boys. Neonatal boys are more susceptible than girls to urinary tract infections, but beyond that period, infections in females prevail. Girls younger than 2 years and uncircumcised boys younger than 6 to 12 months also are especially at risk.43

Approximately 5% of children younger than 2 years with a temperature above 39°C and presenting without a source for the fever have an occult urinary tract infection.44 Urinary tract infections are also significant in the very young infant, with up to 9% prevalence in one study of febrile infants younger than 60 days.45,46 Fever duration also appears to correlate with the prevalence of urinary tract infections. Two days of fever was more likely than one day to be associated with a urinary tract infection.47 Other risk factors for female infants include white race, age younger than 12 months, temperature above 39°C, and no other identifiable source for the fever.48 Risk factors for male infants include nonblack race, similar temperature, and no other identifiable source for the fever.48 Moreover, up to 4% with a significant fever and an associated upper respiratory tract infection or acute otitis media also may have a urinary tract infection.49 By comparison, the background prevalence of asymptomatic bacteriuria is estimated to be 1 to 2% in all children.48

On the basis of renal nuclear scans, it is estimated that 75% of children younger than 5 years with a febrile urinary tract infection have pyelonephritis.49 Vesicoureteral reflux from the bladder into the ureter is a common cause of the pyelonephritis and renal scarring. Urinary tract infection in infants younger than 3 months is associated with bacteremia in up to 50% of cases; in children older than 3 months, the risk drops to 5%. Renal scarring can occur in 27 to 64% of children after pyelonephritis and may lead to renal failure and a risk for hypertension later in life.45

E. coli is the predominant cause of urinary tract infections in children; Klebsiella species are more likely to be the etiologic agents in newborn children. Enterobacter, Proteus, Morganella, Serratia, and Salmonella species also are important pathogens.49 Organisms such as Lactobacillus, coagulase-negative staphylococcus, and Corynebacterium are not considered clinically relevant pathogens in otherwise healthy children.44 In neonates and young infants, bacteremia is considered the route of infection to the urinary tract. In older children, infection in the lower tract often is the source of upper tract infection. A common cause of urinary tract infections in toilet-trained girls is believed to be improper wiping after urination. Young girls should be taught to wipe from anterior to posterior (front to back) after urination.

Clinical Features

Infants and Children Younger than 2 Years. Signs and symptoms usually are nonspecific and include decreased oral intake, lethargy, jaundice, fever, vomiting, abdominal pain, and irritability. Young children may not be able to verbalize when urination is painful. It is assumed that a urinary tract infection in this age group represents upper tract disease and would therefore arise with more systemic symptoms and signs.

Children Older than 2 Years. Urinary tract infection in children older than 2 years can be either an isolated cystitis or upper tract disease with symptoms and signs of a more systemic nature. Cystitis usually is associated with local symptoms (i.e., suprapubic tenderness and dysuria). Clinical manifestations of pyelonephritis may include fever, costovertebral angle tenderness to palpation, abdominal pain, vomiting, and ill appearance. New-onset bedwetting also may be a sign of a urinary tract infection.

Diagnostic Strategies

Various techniques can be used for collection of urine samples from children. Because of the difficulty in cleaning the perineal area, the bag collection method is associated with an increased risk of contamination by periurethral flora; false-positive results range from 12 to 83%.45 Because urethral catheterization is almost always successful, suprapubic aspiration to obtain a urine sample is rarely needed.46-48 A suprapubic bladder aspiration may be used for young infants because the expanded bladder is located more intra-abdominally. Bladder aspiration is considered more invasive and is more painful than the urethral catheterization method.44 Chances of a full bladder and successful aspiration improve if 45 to 60 minutes have elapsed since the last diaper change. Ultrasonographic guidance has been advocated to enhance the probability of obtaining urine by the suprapubic and catheterization methods. Nursing staff can be trained to use this enhanced method of urinary collection.50,51

Urethral catheterization is relatively simple and poses little risk, although it may be more difficult in uncircumcised boys or in young infants. There is a slight risk of both trauma to the urethra and introduction of bacteria into the urinary tract with this technique. A 5-F feeding tube can be used in young infants. Young male infants sometimes spontaneously urinate when the urethral meatus is cleansed; a midstream clean-catch urine specimen may then be obtained.

For a clean-catch urine sample from a toilet-trained child, the parent can clean the child’s urogenital area with soap and water before urination. Children can be instructed to sit backward on
the toilet to urinate, allowing better access to obtain the urine specimen. A urine sample with more than 10 white blood cells (WBCs) per high-power field and a large number of epithelial cells should be considered contaminated, and either an improved clean-catch method or catheterization should be tried. Females with a vaginal discharge, regardless of age, should be catheterized.

In children younger than 2 years, a urinalysis alone is not considered adequate to rule out urinary tract infections. As many as 10 to 50% of patients with urinary tract infection can have false-negative results on urinalysis.\textsuperscript{52,53} Nitrite and leukocyte esterase urinary markers have the highest combined sensitivity and specificity for detection of infection. If both markers are positive, the false-positive rate is less than 4%. The urine dipstick alone appears to work less well in young infants.\textsuperscript{45,55} Gram's stain of the urine has a sensitivity of 93%,\textsuperscript{45,56} New guidelines have redefined urinary tract infections in children. To establish the diagnosis of urinary tract infection in children, the clinician should require both of the following: (1) urinalysis results suggesting infection (pyuria, bacteriuria, or both) and (2) the presence of at least 50,000 colony-forming units (CFU) per milliliter of a uropathogen cultured from a urine specimen.\textsuperscript{44} Old guidelines of 100,000 CFU/mL were based on studies of adult women and are considered no longer applicable to children.

Renal function tests are rarely abnormal but should be performed in children with hypertension, proteinuria, or signs of dehydration. In general, blood cultures are not indicated in a majority of well-appearing children with urinary tract infection. The true-positive rate for blood culture in the presence of a urinary tract infection in older infants and young children is low, and the organism identified is invariably the same as the organism in the urine culture.\textsuperscript{56}

Differential Considerations

Underlying renal disease or urinary tract abnormality should be considered in a child presenting with hypertension, hematuria, or difficulty with urination; it should be suspected in children with elevated BUN or creatinine concentration, electrolyte abnormalities, or acidosis on laboratory examination. CT of the abdomen may be necessary to delineate the extent of urinary tract abnormalities.

Several other causes of dysuria in children are recognized (Box 174-1). Irritants such as bubble bath and soaps may cause local irritation and dysuria. A retained foreign body in the vagina or penis (such as toilet paper) can cause irritation or bacterial growth with associated dysuria and vaginal discharge. Pinworms in the genitourinary area can cause itching and scratching. Balanitis in uncircumcised boys also can cause dysuria and pyuria.

Accidental injuries to the genital area can cause abrasions or lacerations and subsequent dysuria. Sexual or physical abuse should be considered in any young child with a history of multiple urinary tract infections.

Management

Children who are considered “ill appearing” or “toxic” or who are unable to take oral medications should be treated with parenteral antibiotics until clinical improvement, generally 24 to 48 hours, and they are able to retain oral medications and fluids. If compliance is in question, parenteral antibiotics should also be considered.

Infants Younger than 2 Months. Young infants with urinary tract infections are at risk for associated sepsis. The age at which a urinary tract infection warrants hospital admission has decreased dramatically. Infants younger than 2 months are still considered to be at high risk for the development of sepsis and should therefore be admitted to the hospital for intravenous antibiotic therapy (e.g., with gentamicin and ampicillin). One study, however, showed low adverse events in young infants (29-60 days of age) diagnosed with a urinary tract infection who appeared well in the ED and did not have a high-risk past medical history.\textsuperscript{57-59}

Children 2 Months to 2 Years. Although inpatient treatment of children with suspected pyelonephritis has traditionally been recommended, studies now show that well-appearing children without signs of toxicity may be managed as outpatients.\textsuperscript{60} A meta-analysis of data from both adult and pediatric studies found no significant evidence suggesting that oral antibiotic therapy is less effective than parenteral or initial parenteral therapy for treatment of severe urinary tract infection.\textsuperscript{61} Choices of oral antibiotics include a cephalosporin, amoxicillin with clavulanic acid, and trimethoprim-sulfamethoxazole.\textsuperscript{44} Antibiotics that are excreted in the urine but do not reach sufficient levels in the bloodstream, such as nitrofurantoin, should not be used.\textsuperscript{44} Community antimicrobial resistance patterns are important to consider in deciding on an appropriate antimicrobial. Rates of E. coli resistance to trimethoprim-sulfamethoxazole can be as high as 20 to 30% in some communities.\textsuperscript{54} The first dose of a parenteral antibiotic and a double dose of an oral antibiotic appear to be equivalent. Because urinary tract infections in this age group are considered to be upper tract disease processes, a longer course of antimicrobials is indicated (e.g., 7 to 14 days). Although a review of studies found no difference in the frequency of positive urine cultures between short-course therapy (2 to 4 days) and longer course therapy (7 to 14 days), most of these studies mixed children younger than 2 years with older children, including adolescents. Thus the recommendation for short-course therapy in children younger than 2 years is not supported at this time.\textsuperscript{43,65} Follow-up in 2 to 3 days is essential to evaluate the culture and sensitivity results and to assess clinical status. Older recommendations required both ultrasonography and voiding cystourethrography to be performed on all infants with first-time urinary tract infections. Newer recommendations focus only on a renal and bladder ultrasound examination looking for evidence of renal scarring or anatomic abnormalities that require further evaluation. For children who clinically improve quickly, this ultrasound examination can be done at a later time on an outpatient basis.\textsuperscript{44}

Children Older than 2 Years. In older children with simple cystitis, a short 3-day course of antibiotics, such as trimethoprim-sulfamethoxazole or amoxicillin-clavulanate, is adequate. Shorter courses result in higher failure rates and are not indicated. In older children with pyelonephritis, a longer course of antibiotics (generally 7-14 days) is recommended.\textsuperscript{44} In the adult population, a study

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**Box 174-1 Etiology of Dysuria in Children**

**Infection**
- Urinary tract infection, including cystitis and pyelonephritis
- Vaginitis resulting from *Gardnerella, Trichomonas, Candida*, or sexually transmitted organisms
- Pinworms
- Balanitis

**Irritation**
- Bubble bath, new soaps, or douches
- Vaginal foreign body, such as retained toilet paper

**Trauma**
- Sexual or physical abuse
- Straddle injury (unintentional)
- Self-stimulation or masturbation

**Other**
- Labial adhesions
- Renal stones or hypercalciuria
showed that a 7-day course of a fluoroquinolone is as effective as a 14-day course of trimethoprim-sulfamethoxazole. In children, fluoroquinolones are still contraindicated because of the concern for their effect on skeletal development.

Disposition

Children with signs of toxicity, urinary obstruction, or inability to take oral medications should be hospitalized for intravenous antibiotic therapy. On discharge, parents should be instructed to return if the child is unable to take the oral medication or if the symptoms worsen. The child may remain febrile for 48 hours despite adequate antimicrobial therapy. The child’s clinical status should be checked in 48 to 72 hours, either in person or by telephone. At follow-up, the urine culture results should be checked to ensure appropriate antimicrobial therapy.

Hematuria

Perspective

Hematuria is common in the pediatric population. Hematuria, defined as the presence of red blood cells (RBCs) on examination of two successive urine samples, has a prevalence rate of 1 to 2% in children 6 to 15 years of age. Microscopic hematuria is defined as the presence of more than 5 RBCs/mm<sup>3</sup> and is detected by chemical reagent strips and microscopic urinalysis. Persistent hematuria is defined as positive results on three urinalyses during a 2- to 3-week period. Macroscopic or gross hematuria is presence of blood in the urine visible to the naked eye and may be associated with clots.

Principles of Disease

Hematuria results from the entry of RBCs into the urinary tract. Inflammation, infection, trauma, or anatomic abnormalities can occur anywhere from the glomerulus to the urethra. The endothelium and basement membrane of the glomerulus usually are impervious to large proteins such as RBCs and hemoglobin, but myoglobin may pass freely. Damage to the glomerulus, however, may allow RBCs and hemoglobin to pass into the collecting system.

Urine with lysed RBCs tests positive for hemoglobin and negative for RBCs and may be pink. Urine containing myoglobin from muscle breakdown tests positive for hemoglobin and negative for RBCs. Evaluation of creatine kinase and fractionated bilirubin levels and hematocrit often can help differentiate hemoglobinuria from myoglobinuria. Not all red urine contains blood. Certain drugs or foods, such as phenothiazines, ibuprofen, beets, and blueberries, can cause reddish urine. In neonates, urate crystals can cause red-tinged urine in the diaper. Serratia marcescens, a fecal pathogen, can cause a red pigmentation when left in the diaper. Bleeding from the vagina or rectum can sometimes be mistaken for blood in the urine.

Clinical Features

The history and physical examination should focus on signs of infection, trauma, or bleeding disorders. Signs of renal disease (e.g., hypertension), edema, rales, and cardiac murmurs are important findings. An examination of the genitalia may reveal inflammation or bleeding.

Differential Considerations

The etiology of hematuria comprises a diverse group of disorders and conditions (Box 174-2). Presence of RBCs in the urine as a result of trauma is discussed elsewhere.

Box 174-2 Etiology of Hematuria in Children

**Extrarenal**
- Trauma
- Meatal stenosis or posterior urethral valves
- Exercise
- Menstruation or rectal bleeding
- Foreign bodies
- Cystitis, urethritis, or epididymitis

**Intrarenal**
- Pyelonephritis
- Renal or bladder stones or tumors
- Poststreptococcal or idiopathic glomerulonephritis
- Acute interstitial nephritis
- Acute tubular necrosis
- Basement membrane glomerular disease
- Renal vein or artery thrombosis
- Recurrent familial hematuria
- Polycystic kidney disease

**Systemic**
- Henoch-Schönlein purpura
- Systemic lupus erythematosus
- Hemolytic-uremic syndrome
- Infectious mononucleosis
- Sickle cell disease or other hemoglobinopathies
- Bacterial endocarditis or artificial cardiac valves
- Bleeding disorders, warfarin, or aspirin
- Medications such as amitriptyline or chlorpromazine, radiocontrast dyes
- Munchausen syndrome or factitious

Extrarenal causes of hematuria include urethral valves and meatal stenosis. Extensive exercise can also cause hematuria or myoglobinuria as a result of both direct renal trauma and ischemic injury.

Intrarenal disorders include pyelonephritis, renal or bladder tumors, poststreptococcal acute glomerulonephritis, and basement membrane disease of the glomerulus.

Systemic illnesses that cause hematuria, such as Henoch-Schönlein purpura and hemolytic-uremic syndrome (HUS), are discussed later in this chapter. Other systemic causes of hematuria include bleeding disorders, aspirin, and anticoagulants.

Diagnostic Strategies

A urinalysis that shows more than 5 RBCs per high-power field indicates hematuria. The presence of WBCs or leukocyte esterase should be noted. RBC casts in the urine or significant proteinuria demonstrates a glomerular origin for the hematuria. If glomerular disease is suspected, a throat culture, antibody test for streptococcus, complement studies, erythrocyte sedimentation rate determination, antinuclear antibody assay, or hepatitis B serology tests should be considered. If the patient has signs of hypertension, edema, or proteinuria, laboratory tests for electrolytes, total protein, and albumin should be ordered.

If the cause of the hematuria is still unclear, follow-up with the patient’s pediatrician for a 24-hour urine collection for creatinine and protein is important. Because hypercalcemia is common in children, urine and plasma calcium levels should also be measured as they will give a better indication of the cause of the hypercalcemia.

Because younger pediatric patients have predominantly renal stones rather than ureteral stones, ultrasonography can be used as the first-line imaging modality for the diagnosis of renal stones in institutions with dedicated pediatric ultrasonographers. Should the ultrasound study be normal in a patient when renal stones are
Renal Stones

Management

Management of the child with hematuria depends on the underlying cause. The management of the child with renal stones and renal tumors is discussed later in the chapter.

Disposition

The disposition of a child with hematuria depends on the underlying cause. Simple cystitis or pyelonephritis can be treated in the ED with follow-up by the child’s primary care physician. A child diagnosed with a renal tumor or stone should be referred to a nephrologist or urologist. Children with acute renal failure, especially those with signs of fluid overload or hyperkalemia, need to be hospitalized for appropriate management. For children with no known cause of the hematuria but who appear well in the ED, follow-up with their own pediatrician in the next 1 to 2 weeks is important. Persistent hematuria with no known cause should be followed up by a nephrologist for further evaluation.

Clinical Features

The signs and symptoms of renal stones in older children typically include colicky flank pain, vomiting, and hematuria. Younger children with renal stones also can present with less specific complaints, such as nontender abdominal pain, vomiting, or malaise. In a study from the United Kingdom, 17% of pediatric patients with renal stones were found to present with hematuria only, without flank or abdominal pain. In the initial evaluation of children with possible renal stones, the history should ascertain these symptoms. Family history also seems to play a role in the formation of kidney stones. In younger pediatric patients, renal stones predominate over ureteral stones.

Perspective

Renal stones result from a complex process of crystallization involving growth inhibitors and promoters and changes in urine pH and flow. Congenital abnormalities, trauma, or infection also can be an inciting cause. An increased risk for hypercalcuria and the formation of renal stones often is inherited.

Principles of Disease

Renal stones are uncommon in children, with approximately 1 case diagnosed for every 1000 pediatric hospital admissions. Calcium-containing stones are responsible for approximately 60% of all cases, followed by struvite, uric acid, and cystine stones. Struvite stones are composed of ammonium, magnesium, and phosphate and are potentiated by urea-splitting organisms such as Proteus, Pseudomonas, and Klebsiella. Renal stones are three to four times more common in white children than in nonwhite children. Family history also seems to play a role in the formation of kidney stones. In younger pediatric patients, renal stones predominate over ureteral stones.

Diagnostic Strategies

In children thought to have renal stones, the urine should be checked for hematuria and evidence of infection, such as positive leukocyte esterase or nitrites, leukocytes, and bacteria. The urine also should be strained and any crystals sent for analysis. Other laboratory tests to consider are CBC, electrolyte panel, and determination of BUN, creatinine, uric acid, total protein, and albumin levels. Ninety percent of renal calculi are visible on a plain abdominal radiograph, although stool can sometimes obscure the view. Other types of calcifications appearing on plain abdominal radiographs are gallstones, phleboliths, vessel or lymph node calcifications, and calcified malignant neoplasms. Plain films of the abdomen have no additional value over CT scan of the abdomen and should not be obtained in lieu of the CT scan or other imaging.

Non–contrast-enhanced helical CT is the current study of choice for the identification of renal and ureteral stones. It has exceptional sensitivity and specificity for detection of calculi as small as 1 mm and can identify associated processes, such as obstruction, hydroureret, hydrocalyx, and renal abscess. In addition, it is superior to intravenous pyelography in determining alternative diagnoses, such as abdominal tumor. In children, however, the risk of ionizing radiation with CT scanning pushes clinicians to look for alternative forms of imaging. Because renal stones predominate over ureteral stones in younger pediatric patients, ultrasonography is a viable alternative for imaging in this population. In one study, sensitivity of ultrasonography was 90% for renal stones and 38% for ureteral stones. Ultrasonography for renal stones in pediatric patients will be of highest utility in institutions with a large pediatric patient volume and dedicated pediatric ultrasonographers. The use of CT scanning in smaller institutions or after a normal ultrasound study in a patient in whom renal stones are still a concern is a reasonable approach.

Differential Considerations

The differential diagnosis for colicky flank or abdominal pain in the pediatric patient generates a long list of possibilities. Gastroenteritis and constipation are more common than kidney stones as causes of colicky abdominal pain in young children. Intussusception most commonly occurs in children younger than 2 years and can be manifested with intermittent abdominal pain. In adolescents, biliary colic and intermittent gonadal torsion also must be considered.

Management

Initial pain management should include a narcotic analgesic in conjunction with a longer-acting nonsteroidal anti-inflammatory drug (NSAID). The analgesic effect of NSAIDs is directly related to the inhibition of prostaglandin-mediated contraction of the ureter and is characteristic of all NSAIDs (e.g., ibuprofen, naproxen, ketorolac). Patients with signs of volume depletion or with a history of fever, vomiting, or poor fluid intake should be adequately hydrated to improve urine flow and to prevent urinary stasis, which may contribute to stone formation. Excessive fluid administration to increase urine flow has not been proved effective, however, to enhance movement of the stone down the urinary tract. Although tamsulosin, an alpha-blocker, has been shown to be effective in adults to increase the passage of kidney stones, there are no significant studies in children to support the practice. One review of hospitalizations of children for renal stones found, however, that the use of tamsulosin had increased in the hospitals.
surveyed from less than 1% to more than 8% in pediatric patients during the years surveyed.74

After culture specimens have been obtained, children with infected urinary tracts with stones should be treated with antibiotics; hospitalization generally will be required. Children with larger renal stones may undergo shockwave lithotripsy as a therapeutic modality. In some centers, ureteroscopy also may be a management option for larger stones.75

Children with normal renal function and adequate pain control, without signs of toxicity or renal infection, can be safely discharged home with good follow-up. Several studies have shown that stones 5 mm or smaller appear to pass safely in pediatric patients.76 Follow-up with a nephrologist or urologist may include parathyroid hormone assay, analysis of fasting urine samples for calcium-to-creatinine ratio, or 24-hour urine collection for determination of calcium, magnesium, phosphorus, uric acid, oxalate, cystine, protein, and creatinine levels.70,77

Renal Tumors

Perspective

Abdominal masses in children are not uncommon, and in infants most are benign renal tumors or cysts.78

Principles of Disease

Renal tumors in children can range from the benign cystic nephroma to the more aggressive malignant rhabdoid tumor. The prognosis will depend on the type and staging of the renal tumor.

Clinical Features

The most frequent presentation for a child with a renal tumor is that of an abdominal mass found by the parent while bathing or dressing the child. Hematuria or pain is a less common presenting manifestation than in the adult population.

Diagnostic Strategies

Because a significant number of abdominal masses are the result of renal cysts, renal ultrasonography is the imaging study of choice. Ultrasonography can define the mass in question and is not associated with exposure to ionizing radiation. Laboratory tests should include CBC, platelet count, BUN and serum creatinine concentrations, urinalysis, and urine catecholamine levels. Urinary catecholamines are increased in 95% of patients with neuroblastoma but are normal in those with Wilms’ tumor. Any solid masses found on ultrasonography will be better defined by CT of the abdomen. If the mass appears malignant, a CT scan of the chest is indicated to look for pulmonary metastases.79

Differential Considerations

Considerations in the differential diagnosis for a renal mass include cystic lesions, such as those of polycystic kidney disease, and severe hydronephrosis resulting from obstruction or severe reflux. Solid masses include Wilms’ tumor, renal cell carcinoma, mesoblastic nephromas, and cystic nephromas.

Management and Disposition

Management depends on the function of the urinary tract. Because the renal mass typically involves only one kidney, renal function usually is maintained.

A preliminary diagnosis of the cause of the mass should be obtained before discharge of the child from the ED. Well-appearing children with normal renal function for whom close follow-up can be ensured may be managed on an outpatient basis. Because of the potential seriousness of a renal tumor, hospital admission should be considered for all children. Consultations with a nephrologist, urologist, and hematologist-oncologist can be coordinated during the admission process.

Proteinuria

Perspective

The normal glomerulus is relatively impervious to albumin, a high-molecular-weight protein. Low-molecular-weight proteins, however, pass through the glomeruli and are reabsorbed in the proximal tubule. Proteinuria can result from either increased passage through the glomeruli or decreased reabsorption by the tubules. In most cases, proteinuria is benign and asymptomatic. If the amount of protein lost is significant, such as in nephrotic syndrome, the resultant hypoalbuminemia (albumin less than 2 g/dL and protein less than 4 g/dL) may cause ascites and generalized edema.

Principles of Disease

Proteinuria is a common finding in children. Trace to mild proteinuria (1+ to 2+) can be seen in up to 85% of children and adolescents screened, especially during the summer months.80,81

Clinical Features

Clinical features seen in a proteinuric child depend on the cause of the proteinuria. Recent pharyngitis, presence of hematuria, changes in weight or urine output, or family history of proteinuria should be investigated. Abnormal findings may include hypertension, edema, ascites, or palpable kidneys in infants. A butterfly rash of systemic lupus or the purpuric rash of Henoch-Schönlein purpura may be evident.

Differential Diagnosis

Causes of proteinuria can be divided into glomerular and tubular. Glomerular causes include nephrotic syndrome, glomerulonephritis, and post-transplantation rejection. Transient causes of altered glomerular function include exercise, fever, and seizures. Tubular causes of proteinuria include heavy metal poisoning, urinary tract infections, and diabetes; an asymptomatic tubular proteinuria also has been described.80,81 False-positive results on urinary dipstick testing for proteinuria can be obtained when the urine is alkaline or when it contains mucus, blood, vaginal secretions, semen, or a significant number of inflammatory cells.

Orthostatic proteinuria is a benign condition characterized by the presence of protein in the urine collected with the patient in an upright position but not in samples collected from a supine child. Proteinuria that is persistent or associated with hematuria or other signs of renal disease usually is a sign of a more serious condition.

Diagnostic Strategies

Mild proteinuria (2+ or less; equivalent to 100 mg/dL or less) requires no further investigation unless there are signs of infection. Moderate proteinuria (3+ or more; equivalent to 300 mg/dL or more) necessitates additional evaluation with laboratory tests for serum total protein and albumin, serum electrolyte, BUN, and serum creatinine levels and urine culture. The child should be referred to a nephrologist, who can initiate a 24-hour urine collection for protein. The urine protein-to-creatinine ratio (urine
Poststreptococcal Glomerulonephritis

Perspective

Poststreptococcal glomerulonephritis (PSGN) is one of the sequelae of streptococcal pharyngitis and, less commonly, infections of the skin. Treatment of streptococcal pharyngitis with antibiotics has not been shown definitively to decrease the incidence of PSGN, as opposed to acute rheumatic fever, another sequel of streptococcal pharyngitis. PSGN remains more of a cause of morbidity and mortality in developing countries and among impoverished populations.

Principles of Disease

PSGN is not well understood but probably results from deposition of circulating immune complexes in the kidney. This results in decreased glomerular filtration, allowing proteins to flow freely into the urine.

Clinical Features

PSGN most commonly occurs in children 3 to 7 years of age, usually with a history of pharyngitis with fever 2 weeks before the onset of glomerulonephritis. Symptoms can be localized to the urinary tract, manifested as hematuria or flank pain, or may be less specific, such as lethargy or generalized edema. Some children may present with more significant clinical signs, such as pulmonary edema, cardiac arrhythmias, or significant hypertension. Renal failure is found in 2% of these patients. In one study, delay in diagnosis of PSGN was associated with a negative infection history and absence of gross hematuria.

Diagnostic Strategies

Urinalysis shows significant blood and protein, with RBC casts in 60% of cases. Pyuria with granular or hyaline casts also may be found.

The ASO titer and immunoglobulin G levels are elevated in PSGN. Total complement levels, especially C3, are decreased in most patients during the first 2 weeks of the illness. The complement levels should return to normal within 3 to 4 weeks. The BUN concentration is elevated; hyponatremia and hyperkalemia also may be present.

Differential Considerations

Considerations in the differential diagnosis include the entities previously mentioned regarding proteinuria, including nephrotic syndrome and urinary tract infection.

Management and Disposition

Management includes restriction of fluid and salt intake and sometimes use of diuretics. A child with significant hypertension, congestive heart failure, or uremia requires hospitalization for inpatient management. Significant hypertension should be treated as outlined in the section on hypertension.

Children with mild symptoms in whom good follow-up can be ensured can be discharged home.

Nephrotic Syndrome

Perspective

Nephrotic syndrome is characterized by hypoproteinemia, proteinuria, and edema. Primary nephrotic syndrome applies to diseases limited to the kidney; renal biopsy is used to stratify patients and to determine therapeutic and prognostic decisions. Secondary nephrotic syndrome results from systemic illnesses such as PSGN.

Principles of Disease

From 2 to 7 cases of nephrotic syndrome per 100,000 children are discovered each year. Boys are affected twice as often as girls are, but the distribution equalizes by adulthood. Primary nephrotic syndrome occurs more commonly in children younger than 5 years, and secondary nephrotic syndrome occurs more often in older children. Ninety percent of affected children have the primary disease; 85% have minimal change nephrotic syndrome, 10% focal sclerosis, and 5% mesangial proliferation (referring to the thin membrane-supporting capillaries surrounding the tubule of nephrons).

The etiology of primary nephrotic syndrome is thought to be idiopathic, but various theories involving bacterial or viral infections, allergic reactions (pollens, poison ivy), or drug ingestions (heroin, mercury) have been investigated.

Clinical Features

Characteristics of nephrotic syndrome include edema, hypoalbuminemia, proteinuria, and hyperlipidemia. The onset of edema may be insidious, beginning with periorbital edema. As weight increases, pants and shoes may not fit. The edema progresses, but the child usually does not appear ill unless pulmonary edema or ascites is present. Other features may include anorexia, nausea, and vomiting secondary to edema of the intestine. Hypertension, hematuria, or oliguria may be present. Acute renal failure is rare in primary nephrotic syndrome.
Nephrotic children also are at risk for thrombosis, with a reported rate of thromboembolic complications of 2%. Renal veins are particularly vulnerable to thrombosis, as characterized by flank pain, hematuria, and worsening renal function. Nephrotic children should not undergo punctures to deep vessels because of this risk of thrombosis.

Children with nephrotic syndrome taking corticosteroids also are at greater risk for side effects of corticosteroid use even at dosages as low as 1 mg/kg per 48 hours. Acute mood changes, from depression to mania, are associated with use of steroids. Irritability, excessive crying, and sleeping difficulties can result.

Because of steroid therapy and decreased levels of immunoglobulins, nephrotic children are at risk for bacterial infections, such as from *E. coli* and *Streptococcus pneumoniae*.

**Diagnostic Strategies**

Proteinuria in nephrotic syndrome is defined as excretion of more than 3.5 g of protein per 1.73 m² per 24 hours or more than 50 mg/kg per 24 hours. This corresponds to 3+ or 4+ on the dipstick reading. Specific gravity may be high because of the proteinuria. Microscopic hematuria also may be present. Total serum protein usually is low at 4.5 to 5.5 g/dL, and serum albumin is less than 2 g/dL.

Hyperlipidemia may occur because of the increased cholesterol. Hyponatremia may be present, but other electrolytes usually are normal. If an elevated cholesterol level is present, the lowered sodium level may be a combination of true hyponatremia and pseudohyponatremia. BUN and creatinine concentrations usually also are normal, and hemoglobin and hematocrit levels may be elevated because of hemoconcentration.

Chest radiographs sometimes show pleural effusions or pulmonary edema. The heart appears normal or small because of hypovolemia. An abdominal radiograph may reveal ascites, and ultrasonography may show a renal abnormality.

Renal biopsy is important for diagnostic and therapeutic decisions and should be performed in older children or in patients with evidence of hematuria, elevated BUN, or persistent hypertension or in whom the renal dysfunction fails to respond to steroids.

**Differential Considerations**

Other renal diseases that cause edema include glomerulonephritis and renal failure. A vasculitis or acute thrombosis of the renal vessels also must be considered. Gastrointestinal disorders that produce hypoproteinemia include cirrhosis, cystic fibrosis, and protein-losing enteritides.

**Management**

Despite the edema, children with signs of hypovolemia or shock need to be resuscitated with crystalloid. Hypertension may result from the intrinsic disease; prompt recognition and treatment of this condition are important.

After consultation with a pediatric nephrologist, patients between 12 months and 5 years of age with no gross hematuria and no large loss of protein or complement can be treated with corticosteroids. After the initial evaluation is completed, including a tuberculin test, prednisone at 2 mg/kg per 24 hours orally, divided two or three times a day, can be initiated. Relapses or steroid resistance may necessitate a second course of steroids.

Diuretics such as furosemide, 1 to 2 mg/kg per 24 hours orally or intravenously in divided doses, may be necessary if respiratory distress or significant ascites is present. Salt restriction may be required. Fluid intake should be restricted only if edema is present despite salt restriction or if the child exhibits hyponatremia because of an impaired ability to excrete excess water.

The relatively immunocompromised status of nephrotic children increases the risk of infection. A fever or signs of peritonitis must be investigated thoroughly. A paracentesis should be performed and fluid sent for cell and differential counts, Gram’s stain, and culture. Hospital admission and empirical antibiotic therapy with agents with activity against *S. pneumoniae* and *E. coli* are appropriate.

**Disposition**

Hospitalization should be considered for newly diagnosed patients for initial evaluation, treatment, and education of both the child and the parents. Patients with signs of shock or respiratory distress should be admitted to the hospital after initial stabilization. Other patients with suspected bacterial infections, peritonitis, edema refractory to therapy, or evidence of renal insufficiency also should be hospitalized.

**Acute Renal Failure**

**Perspective**

Acute renal failure results from impaired glomerular filtration rate. Blood pressure, acid-base balance, removal of nitrogen waste, and fluid management may be affected. The incidence of acute renal failure is unknown, but large children’s hospitals may see 30 to 50 new cases each year.

**Principles of Disease**

Acute renal failure can be divided into three categories: prerenal, which involves decreased renal perfusion; renal (intrarenal), which is caused by parenchymal damage; and postrenal, which involves obstruction of the urinary tract (Box 174-3).

Causes of prerenal failure include hypovolemia resulting from a variety of factors (e.g., dehydration, burns, hemorrhage), shock (e.g., sepsis, anaphylaxis), and congestive heart failure (e.g., decreased cardiac output). Obstruction of the renal artery or thrombosis of the renal vein also can cause acute renal failure.

Intrarenal causes involve damage to the nephron. Glomerular damage most commonly results from PGN. Systemic lupus erythematosus, HUS, and sepsis with hypoperfusion are systemic causes of renal failure. Tubular damage can result from heavy metal poisonings or hemoglobin-myoglobin in the tubules from a crush injury, burn, or hemolytic crisis. Cases of acute renal failure in dehydrated children after NSAID use have been reported.

Postrenal failure resulting from an obstruction in the urinary tract may be caused by infection, tumor, renal stones, or posterior urethral valves. Bilateral obstruction of the kidneys usually is necessary for renal failure to occur.

**Clinical Features**

A child with acute renal failure accumulates nitrogen wastes shown by an increase in BUN and serum creatinine levels. A decrease in urine output is sometimes seen. A urinary output of 1 mL/kg per hour usually is considered adequate, but the output may be lower if the patient is dehydrated. Nephrotic agents such as certain aminoglycosides can actually increase the urinary output because of renal tubular damage, but BUN and serum creatinine levels are still elevated.

Acute renal failure can result in life-threatening complications that must be recognized and treated promptly. These include severe hyperkalemia, pulmonary edema or fluid overload,
Etiology of Acute Renal Failure in Children

**Pre-renal**
- Decreased intravascular volume or dehydration
  - Burns or hemorrhage
  - Third spacing
  - Sepsis
- Decreased cardiac output
- Cardiac shock
- Decreased renal artery blood flow

**Intra-renal**
- Glomerular disease
  - Poststreptococcal and other glomerulonephritis
  - Pyelonephritis
- Systemic causes
  - Hemolytic-uremic syndrome
  - Henoch-Schönlein purpura or other vasculitides
  - Systemic lupus erythematosus
  - Sepsis or other causes of prolonged decreased perfusion

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

**Post-renal**
- Obstructive lesions
  - Nephrolithiasis or tumor
  - Posturethral valves
  - Intra-abdominal tumor obstructing urinary flow
  - Infection
- Renal vein thrombosis

Management

The initial management of a child with acute renal failure depends on the clinical evaluation. If hypovolemia resulting from dehydration or blood loss is thought to be the cause of the renal failure, immediate rehydration is necessary. A bolus of 20 mL/kg of crystalloid should be given to prevent possible progression to acute tubular necrosis. If no urinary response is obtained after two boluses of crystalloid, diuretics may be useful in the euvolemic patient. Furosemide, 1 mg/kg per dose every 2 to 6 hours intravenously, may be tried if there is no evidence of obstruction. Bumetanide, 0.015 to 0.1 mg/kg per dose every 6 to 24 hours intravenously (maximum of 10 mg per 24 hours), can be useful if furosemide has no effect. Mannitol, 0.75 g/kg per dose every 6 hours intravenously, also may be helpful but because of the resultant increased urine flow is contraindicated if there is evidence of obstruction.

If the child is considered euvolemic and has no urine output despite diuretic therapy, renal-dose dopamine should be initiated (2 to 5 µg/kg per minute). Consultation with a nephrologist should be obtained.91

If hypertension with encephalopathy develops, a controlled 10 to 20% reduction in blood pressure should be achieved by use of nitropusside or other intravenous blood pressure agents. Oral medications, such as nifedipine and captopril, are useful but are associated with the risk of a precipitous drop in blood pressure. Overaggressive reduction in blood pressure can result in hypoperfusion of central target organs, such as the brain, heart, and kidneys, that have become accustomed to higher perfusion pressures. Further discussion of hypertensive emergencies and treatment can be found in Chapter 84.

Hyperkalemia can result in cardiac dysrhythmias. Potassium levels higher than 6.5 mEq/L may cause changes on the electrocardiogram such as peaked T waves and, if significant, widened QRS complexes. Hyperkalemia with resultant electrophysiologic changes, such as loss of the p wave or widened QRS complex, should be treated with calcium gluconate, 10% solution, 0.5 mL/kg per dose given intravenously over 5 minutes (maximum dose, 20 mL [2 g]). Calcium alters the action potentials of the cardiac cells, thereby decreasing the risk of arrhythmias. The calcium dose may be repeated in 5 minutes.

Sodium bicarbonate, 1 to 2 mEq/kg per dose intravenously every 4 hours, also can be administered. The resultant alkalosis helps move the potassium intracellularly by exchanging H+ for K+. Continued therapy with sodium bicarbonate should be administered in conjunction with pediatric nephrology services because of the risk of increasing volume load with repetitive dosages of sodium. Potassium-binding agents such as sodium polystyrene sulfonate (Kayexalate) are administered orally or rectally and exchange Na+ for K+ in a 1:1 ratio.

Glucose (50% dextrose in water for ≥3 years old and 25% dextrose in water for ≤2 years old), 0.5 to 1 g/kg, should be given along with regular insulin (1 unit for every 4 g of glucose administered). The insulin temporarily shifts the potassium intracellularly.

Nebulized albuterol also can be used to shift potassium temporarily into the cells and lowers serum potassium by 1 to 1.5 mEq/L during 30 minutes.99 Severe cases and those associated with renal failure should be immediately referred to a pediatric nephrologist for emergent dialysis.91

Acute renal failure may lead to seizures caused by either hypertensive encephalopathy or a metabolic derangement, most commonly hyponatremia resulting from dilution of the sodium by free water. Intractable hyponatremic seizures may rarely necessitate the use of hypertonic saline (3% sodium chloride). Each 1 mL/kg of 3% sodium chloride increases the serum sodium by approximately 1 mEq/L. A child with hyponatremic symptoms, such as seizures, will often improve after receiving 3 to 5 mL/kg of 3% sodium.
chloride. Once the seizures have ceased, sodium correction should then continue more slowly to avoid central pontine myelinolysis. Recommendations are to correct the sodium no faster than 10 to 12 mEq/L in the first 24 hours and not more than 18 mEq/L in the first 48 hours.

Normal saline (0.9%) is relatively hypertonic compared with serum and therefore also may be helpful in correcting the hypotremia. Water restriction along with the administration of saline usually is only a temporary treatment because many of these hyponatremic patients ultimately require dialysis.

Sodium bicarbonate will correct a persistent metabolic acidosis and maintain pH 7.1 or higher and a serum bicarbonate level of 15 mEq/L. The base deficit can determine the amount of bicarbonate needed:

\[
\text{Base deficit} = [0.6 \times \text{(body weight in kg)} \times (\text{desired bicarbonate level} - \text{observed level})] + 2
\]

Half the replacement is given in the first 3 hours, with the remainder given during the next 24 hours.

Hemodialysis or peritoneal dialysis may be indicated for refractory fluid overload associated with hypertension, congestive heart failure, pulmonary edema, severe hyperkalemia, hyponatremia or hypernatremia, metabolic acidosis, myoglobinuria resulting from burn or crush injuries, or HUS with hemoglobinuria or encephalopathy.91

Disposition

All children with acute renal failure should be admitted to the hospital. Any child with signs of congestive heart failure, pulmonary edema, significant hyperkalemia, or acidosis should be admitted to a monitored bed.

Hypertension

Perspective

Hypertension is defined as a systolic or diastolic blood pressure higher than 2 SDs above the mean for the age and sex of the patient (Table 174-1). This diagnosis requires three or more accurate blood pressure measurements during the course of several weeks. A correct cuff size should be chosen (i.e., the air bladder should cover 80 to 100% of the circumference and two thirds of the length of the upper arm).85 It is recommended that all children receive yearly blood pressure measurements starting at the age of 3 years.85 A child who is in pain or agitated may have falsely elevated blood pressure readings.

Principles of Disease

Hypertension occurs throughout childhood in both boys and girls. It probably occurs more often in African American children than in white children, just as in the adult population. Predisposing factors include obesity, physical inactivity, and a strong family history. Metabolic syndrome, a combination of insulin resistance, hypertension, and hyperlipidemia, may affect up to 50% of overweight adolescents.85

Just as in adulthood, primary or essential hypertension is unrelated to a second systemic disease. Children diagnosed with primary hypertension are more likely to become adults with hypertension. Secondary hypertension results from endocrinologic, cardiac, neurologic, or other factors, such as exposure to certain drugs or poisons (Box 174-4). In children with significant hypertension, the underlying cause usually is renal (as in glomerulonephritis) or renovascular.

### Table 174-1 Blood Pressure Limits in Children

<table>
<thead>
<tr>
<th>AGE</th>
<th>BLOOD PRESSURE: UPPER LIMIT</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Systolic</td>
</tr>
<tr>
<td>0-2 years</td>
<td>110 mm Hg</td>
</tr>
<tr>
<td>3-6 years</td>
<td>120 mm Hg</td>
</tr>
<tr>
<td>7-10 years</td>
<td>130 mm Hg</td>
</tr>
<tr>
<td>11-15 years</td>
<td>140 mm Hg</td>
</tr>
</tbody>
</table>


### Box 174-4 Etiology of Hypertension in Children

**Primary**
- Essential hypertension

**Secondary Renal**
- Glomerulonephritis
- Henoch-Schönlein purpura
- Pyelonephritis
- Obstruction or reflux
- Polycystic kidney disease
- Diabetic nephropathy
- Trauma
- Renal transplant or hemodialysis
- Tuberous sclerosis
- Systemic lupus nephritis

**Endocrine**
- Pheochromocytoma
- Cushing’s syndrome
- Congenital adrenal hyperplasia
- Corticosteroid treatment
- Hyperthyroidism
- Neuroblastoma
- Ovarian tumor

**Cardiac**
- Congestive heart failure
- Coarctation of the aorta

**Vascular**
- Hemolytic-uremic syndrome
- Kawasaki syndrome
- Renal artery thrombosis or stenosis

**Neurologic**
- Central nervous system tumor or infection
- Central nervous system trauma or abuse
- Increased intracranial pressure
- Guillain-Barré syndrome

**Neoplastic**
- Neuroblastoma
- Wilms’ tumor
- Pheochromocytoma
- Adrenal carcinoma

**Drugs**
- Corticosteroids
- Cocaine
- Sympathomimetics
- Oral contraceptives
- Phencyclidine
- Beta-blocker or clonidine withdrawal
- Lead, mercury

**Others**
- Iatrogenic fluid overload
- Volume overload from end-stage renal disease
Clinical Features

There are a variety of clinical presentations of hypertension in children. First, asymptomatic or mildly symptomatic hypertension may show up in routine vital signs measured in children evaluated in the ED for unrelated illnesses. When asked, these children may complain of headaches, abdominal pain, irritability, or nosebleeds. Personality changes and difficulties in school are sometimes noted.

Hypertension in young children may also be manifested as severe elevations in systolic or diastolic blood pressures (younger than 10 years: systolic blood pressure 160 mm Hg or higher, diastolic blood pressure 105 mm Hg or higher; older than 10 years: systolic blood pressure 170 mm Hg or higher, diastolic blood pressure 110 mm Hg or higher) but without signs of end-organ damage. These cases are also important to recognize as close follow-up with a primary care physician will be important.

In children experiencing a hypertensive emergency, clinical signs of end-organ damage will be present. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage. A severe elevation in blood pressure is associated with acute neurologic changes or signs of end-organ damage.

Hypertensive encephalopathy symptoms include headache, vomiting, altered mental status, visual disturbances (including blurry vision and diplopia), and seizures or stroke. Papilledema, decreased retinal venous pulsations, and cranial nerve palsies may be found on examination. The diagnosis is confirmed when the symptoms and signs subside rapidly after the blood pressure is lowered. Headache alone, without any other associated symptoms or signs, generally is not considered to represent a hypertensive emergency.

Diagnostic Strategies

In addition to a thorough history and physical examination, laboratory and radiologic studies performed in the ED often can determine both the cause of the hypertension and whether a hypertensive emergency exists (Box 174-5).

Differential Considerations

Any situation that causes undue agitation or pain in a child can cause a transient rise in blood pressure. Therefore, the diagnosis of hypertension in a pediatric patient should be based only on carefully measured blood pressures in a nonagitated child during several weeks. Secondary hypertension is more likely in symptomatic younger children, especially those without a strong family history.

Other disorders with presentations similar to that of hypertensive encephalopathy include meningitis, brain tumor, intracerebral hemorrhage, stroke, and uremia. These conditions, however, generally produce only a mild increase in the systolic blood pressure; CT or lumbar puncture also can help identify these other disorders.

Management

The management of hypertensive emergencies also is discussed in Chapter 84. When a child with severe hypertension is seen in the ED, questions should focus on a history of hypertension, urinary tract infections, hematuria, edema, or umbilical artery catheterization. A history of joint pain or swelling, palpitations, weight loss, flushing of the skin, or drug ingestion or a family history is important to ascertain.

A physical examination emphasizing the central nervous system and cardiopulmonary system is indicated. Examination of the fundus may reveal papilledema or hemorrhages. Signs of congestive heart failure or a difference in the upper and lower extremity blood pressures should be noted. Coarctation of the aorta may be manifested with a difference in either blood pressure or palpable pulse between the upper and lower extremities. A renal cause of the hypertension may be revealed by the presence of peripheral edema or palpable kidneys. An abdominal or flank bruit suggests renovascular hypertension. Initial laboratory tests should include a CBC, electrolyte values, BUN and creatinine concentrations, urinalysis, urine culture, chest radiograph, and electrocardiogram.

A child in whom clinical findings are consistent with a hypertensive emergency (e.g., acute end-organ damage as found by physical examination, laboratory, or radiologic results) should have intravenous access and be monitored with continuous blood pressure readings; an arterial catheter is preferable. The goal of therapy is to reduce the mean arterial blood pressure by 10 to 20% during several minutes to hours, depending on the nature of the emergency. Headache and vomiting require blood pressure control during several hours, whereas intracranial bleeding or herniation requires reduction in several minutes. Beta-blockers are contraindicated in patients with decreased cardiac output and clinical signs of congestive heart failure. Oral nifedipine is contraindicated in patients with signs of end-organ damage, such as intracerebral bleeding, because of the inability to control the amount of blood pressure reduction. For avoidance of overaggressive treatment and resultant relative hypotension, medications that can be controlled by intravenous infusions are preferred (Table 174-2). Severe hypertension without evidence of end-organ damage, as stated earlier, is also important to recognize. Antihypertensives should be started or restarted for these patients to prevent end-organ sequelae. Angiotensin-converting enzyme inhibitors or calcium channel blockers are useful as first-line agents and usually are well tolerated. The child may be observed for a few hours after

**Box 174-5**  Diagnostic Evaluation in Hypertensive Emergencies* in Children

| History of medications or drugs, or family history of cardiovascular disorders |
| Symptoms of severe headache or chest pain |
| Physical examination focused on acute neurologic changes, funduscopic abnormalities, pulmonary edema |
| Urinalysis for significant protein |
| Chest radiograph for cardiomegaly or congestive heart failure |
| Electrocardiogram for ventricular hypertrophy |

**Primary or Secondary Cause? Laboratory and Radiologic Tests to Consider**

| Urinalysis and urine culture |
| Urine catecholamine assay |
| Complete blood count with platelet count |
| Blood smear |
| Sodium, potassium, chloride, carbon dioxide, calcium, phosphorus, magnesium, uric acid measurement |
| Blood urea nitrogen and creatinine determinations |
| Serum C3 complement assay, antistreptolysin O titer, antinuclear antibody assay |
| Plasma renin level |
| Computed tomography urography or intravenous pyelography |
| Voiding cystourethrography |
| Renal ultrasound scan |
| Renal arteriography |

*Evidence of end-organ damage.
administration of the medication for evaluation of effectiveness or complications. With no evidence of acute end-organ damage, the child can be safely discharged home, assuming that good follow-up can be ensured.

Children with mildly elevated blood pressures (5 to 10 mm Hg above normal) unrelated to the ED visit require repeated blood pressure measurements before treatment of hypertension is begun. If the blood pressure is moderately elevated and the patient is asymptomatic, appropriate management consists of discharge home for outpatient workup of the hypertension and follow-up with the patient’s primary care physician for monitoring of blood pressure values. Thiazide diuretics or beta-blockers may be started at low doses.

**Disposition**

A child with evidence of a hypertensive emergency (i.e., acute end-organ damage) must be hospitalized for evaluation and care in a monitored bed. A child with significantly elevated blood pressure values. Thiazide diuretics or beta-blockers may be started at low doses.

**Henoch-Schönlein Purpura**

**Perspective**

Henoch-Schönlein purpura is an immunoglobulin A–mediated systemic vasculitis involving the small blood vessels supplying the skin, gastrointestinal tract, and joints. The peak incidence is between 4 and 7 years of age, with an overall occurrence rate of 13.5 episodes per 100,000 children annually.

**Principles of Disease**

Immune complex deposition results in a systemic vasculitis, with up to 33% of patients experiencing recurrences. Approximately 50% of affected children have a history of previous upper respiratory tract infection, and as many as 75% have group A beta-hemolytic streptococci cultured from the oropharynx. Other theorized predisposing factors include exposure to cold weather, certain foods, drugs, and insect bites. Varicella-zoster virus, *Mycoplasma* species, parvovirus, *Campylobacter enteritis*, parvovirus B19, and Epstein-Barr virus also have been implicated.

**Clinical Features**

The hallmark of Henoch-Schönlein purpura is a palpable, purpuric, or petechial rash most prominent on the lower extremities, starting at the lateral malleoli and extending to the buttocks. The cutaneous manifestations are the initial presenting complaint in 50% of patients. Arthralgia or arthritis occurs commonly and usually involves the knee and ankle joints. Gastrointestinal manifestations are present in up to 65% of patients, with the most common symptom being dull periumbilical pain resulting from bleeding into the intestinal wall. The abdominal pain typically occurs concurrently with or immediately after onset of the rash; however, in up to 25 to 50% of patients, abdominal pain or arthritis can be the initial complaint. A self-limited glomerulonephritis manifested by hematuria develops in 25 to 50% of children. This condition progresses to chronic renal insufficiency in less than 1% of these patients. Children presenting with acute renal failure, nephrotic syndrome, or hypertension are more likely to have unfavorable outcomes, such as chronic renal failure.

**Diagnostic Strategies**

There are no specific tests to confirm the diagnosis of Henoch-Schönlein purpura, and recognition of the disorder can be difficult if the classic rash is absent at the time of presentation. Screening tests such as urinalysis, BUN and serum creatinine determinations, CBC, and coagulation studies may be needed to rule out other pathologic conditions. Ultrasound findings in patients with abdominal pain typically include evidence of intraluminal hematomas and duodenal wall thickening. Intussusception also may complicate the disease.

**Differential Considerations**

Considerations in the differential diagnosis include meningococcemia, Rocky Mountain spotted fever, intussusception, trauma,
applicative forms are recognized as well. Immunodeficiency virus, and drugs; hereditary, familial, and idiopathic, kidney stones, and primary renal disease.

Management

Treatment of Henoch-Schönlein purpura remains controversial because most cases resolve spontaneously and do not require therapy. NSAIDs can be used to treat joint pain, but close attention must be paid to renal function. Corticosteroids have been used to treat severe renal or gastrointestinal involvement. A meta-analysis of data on the use of steroids for Henoch-Schönlein purpura found these agents to be effective in reducing the time to resolution of abdominal pain and to reduce the odds for development of persistent renal disease. Prednisone or methylprednisolone pulse therapy has shown some benefit, but an acute surgical process should be excluded before initiation of steroids. Therapy for patients with severe renal involvement also includes intravenous immune globulin, although promising results also have been seen in patients with severe abdominal pain. Other treatment options for patients with severe Henoch-Schönlein purpura–related nephropathy include early treatment with oral immunosuppressants and possibly the use of methylprednisolone and urokinase pulse therapy. A nephrologist should be consulted for appropriate management of patients with renal involvement, and good follow-up must be ensured.

Disposition

Patients with only the skin manifestations of Henoch-Schönlein purpura usually can be discharged home with symptomatic therapy for the joint pain and malaise. An NSAID or acetaminophen usually is sufficient, but close follow-up should be ensured. Patients with abdominal pain or renal involvement should be admitted for further evaluation and treatment.

Hemolytic-Uremic Syndrome

Perspective

HUS continues to be one of the most common causes of acute renal failure in children. It most commonly affects infants and children, with a mean age at presentation of 3 years, but is rare after 5 years of age. There is no sex predilection, and outbreaks can be sporadic or epidemic, especially when the disorder is related to the most common offending agent, verotoxin produced by E. coli serotype O157:H7. Transmission is through person-to-person contact and also exposure to contaminated food, such as unpasteurized dairy products or beef. Other causes of HUS include Shigella organisms, S. pneumoniae, Aeromonas, human immunodeficiency virus, and drugs; hereditary, familial, and idiopathic forms are recognized as well.

Principles of Disease

Renal compromise is the result of injury to the renal vascular endothelium induced by viral or bacterial agents or the toxins released. Microangiopathic hemolytic anemia then results from injury to the RBCs by fibrin strands along the narrowed blood vessels. Platelets, complement, and fibrin also are deposited in the glomerular lumen, leading to a decrease in glomerular filtration rate and renal failure.

Clinical Features

Patients with HUS present with watery diarrhea, crampy abdominal pain, and occasionally fever. From 2 to 3 days after onset of symptoms, patients experience increased abdominal pain with bloody stools, the latter developing in up to 89% of patients by day 5. Other pathologic features may include toxic megacolon, ischemic colitis, intussusception, perforation, or delayed colonic stricture. After the prodromal gastroenteritis, patients experience sudden onset of hemolytic anemia, thrombocytopenia, and acute renal insufficiency, with possible progression to renal failure. In a meta-analysis published in 2003, death or end-stage renal disease occurred in 12% of cases of diarrhea-associated HUS, and 25% of survivors demonstrated long-term renal sequelae. Pancreatic insufficiency resulting in insulin-dependent diabetes mellitus also has been reported. There are reports of HUS occurring in association with urinary tract infection as well.

Central nervous system irritability may develop and may result in seizures in 40% of patients. Hypertension occurs in up to 50% of patients and may contribute to the development of encephalopathy. HUS recurrences have been associated with a 30% mortality rate.

Diagnostic Strategies

Leukocyte counts and C-reactive protein levels were found to be significantly higher in patients with toxin-producing E. coli O157:H7. The peripheral blood smear shows microangiopathic changes such as teardrop cells, helmet cells, microspherocytes, and burr cells. WBC counts may be elevated, and the platelet count may be less than 50,000/µL. The hemoglobin can be as low as 5 g/dL as a result of the rapid hemolysis that occurs.

Differential Considerations

Considerations in the differential diagnosis of HUS include thrombocytopenic thrombotic purpura, ulcerative colitis, intussusception, and other causes of acquired hemolytic anemia.

Management

Supportive therapy and early peritoneal dialysis account for the reduction in current mortality rates to less than 5%. Patients should be rehydrated; however, it is important not to overload these children with fluids. Hyperkalemia is common and should be treated with sodium bicarbonate, calcium gluconate or chloride, dextrose and insulin, and sodium polystyrene sulfonate (Kayexalate). Patients with severe hyperkalemia, hyperphosphatemia, or severe metabolic acidosis require dialysis.

Packed RBCs (5 mL/kg during 4 hours) typically are administered if the hemoglobin falls below 6 g/dL. Platelet transfusions are required only for life-threatening bleeding or before an invasive procedure. Hypertension is responsive to calcium channel blockers, labetalol, captopril, or nitroprusside in refractory cases. Seizures typically respond to benzodiazepines and phenytoin; however, if they are secondary to hyponatremia, treatment with 3% saline (4 mL/kg) may be indicated. Treatment of the colitis itself also is supportive only because antimotility agents may lead to toxic megacolon. To date, no randomized trials have been conducted to show the effectiveness of antibiotics for the prevention of the development of HUS. Accordingly, because antibiotics may enhance the release of verotoxin from the bacteria, antibiotics should be avoided. Hyperglycemia, ketonemia, and acidosis secondary to pancreatic islet cell necrosis are managed with insulin therapy.

Idiopathic HUS may be treated with plasmapheresis, especially if neurologic involvement is present. If this therapeutic measure is unsuccessful, renal transplantation may be required. Unfortunately, idiopathic HUS also can recur in the transplanted kidney.
Disposition

Patients with HUS require hospital admission, and consultation with a pediatric nephrologist and a urologist should be arranged. Early dialysis and supportive therapy result in return to baseline renal function in up to 90% of patients with acute renal failure.

Principles of Management

Early dialysis and supportive therapy result in return to baseline renal function in up to 90% of patients with acute renal failure.

Priapism

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

Phimosis and Paraphimosis

Steroid cream is first-line therapy for phimosis. In paraphimosis involving vascular compromise of the glans penis, a dorsal slit procedure may be necessary.

Testicular Torsion

Delay in diagnosis and treatment can result in loss of spermatogenesis and, in severe cases, a necrotic, gangrenous testis. Testicular salvage rates are time dependent, with a 96% success rate if detorsion is performed less than 4 hours after symptom onset; with more than a 24-hour delay in time to treatment, the success rate decreases to less than 10%.

Varicoceles

Left-sided varicoceles account for 85 to 95% of the cases. Intra-abdominal disease should be suspected in cases of right-sided varicoceles because these usually are caused by inferior vena cava thrombosis or compression of this vessel by tumors.

Urinary Tract Infections

In children younger than 2 years, a urinalysis alone is inadequate to rule out urinary tract infections because this study yields false-negative results in as many as 10 to 50% of patients.

Hematuria

Hematuria is common in children with causes as simple as a urinary tract infection to more complicated conditions, such as glomerulonephritis or Henoch-Schönlein purpura.

Renal Failure

Renal failure thought to be secondary to dehydration should be corrected immediately with a bolus of normal saline, with repeated laboratory studies showing an improvement in renal function.

Hypertension

Other significant clinical conditions, such as meningitis, brain tumor, intracerebral hemorrhage, stroke, and uremia, may be manifested similar to a hypertensive emergency; these conditions, however, in general have only mildly elevated blood pressure.

The references for this chapter can be found online by accessing the accompanying Expert Consult website.
References


