DEVELOPMENTAL ANATOMY OF THE KIDNEYS

The full complement of nephrons is present at birth, although newborn nephrons are heterogeneous in glomerular size and proximal tubule length. Anatomy and function mature postnatally. Although fetal urine is excreted into the bladder by 10 to 11 weeks of gestation, the ability to conserve and excrete sodium, concentrate urine, and reabsorb substrates such as glucose evolves to maturity over the first 2 years of life. In utero, the glomerular filtration rate (GFR) is minimal secondary to placental function; at birth, the GFR is 10% of adult values and matures by 12 to 24 months of age. Therefore, an increase in an infant’s creatinine to “normal” adult ranges can indicate pathology.

• Children often cannot differentiate between abdominal pain and groin pain—a complete physical examination is therefore necessary.
• The pathophysiology, clinical findings, and treatment of parapenimosis, testicular torsion, and priapism are similar in both the pediatric and adult population. They are emergencies that require immediate intervention.
• The most common cause of acute renal failure in children is hemolytic-uremic syndrome.
• Poststreptococcal glomerulonephritis is the most common cause of acute glomerulonephritis. IgA nephropathy is the most commonly diagnosed cause of glomerulonephritis in adolescents.
• One third of patients with Henoch-Schönlein purpura have renal involvement. This disorder is the most common form of vasculitis in childhood and is usually characterized by the triad of abdominal pain, arthritis, and purpura.

CRYPTORCHIDISM (UNDESCENDED TESTIS)

By birth, the testes have usually descended from the abdominal cavity into the scrotum; only 3% to 5% of full-term newborns have an undescended testicle. Although spontaneous descent does occur in the first year of life, 0.8% of males are still affected at 12 months of age, and spontaneous descent becomes increasingly unlikely after 6 months. With a careful physical examination, 80% of undescended testes are palpable, most commonly in the inguinal canal. Children with undescended testes are at higher risk for torsion, trauma, and malignancy.

CLINICAL PRESENTATION AND DIAGNOSIS

Children with undescended testes are ideally identified by careful physical examination before any complications occur. The differential diagnosis of testes that are not palpable includes undescended, retractile, and absent testes. Children younger than 1 year should be examined while they are relaxed in a warm bath because the testes can be retracted during
A varicocele is a collection of spermatic venous varicosities in the scrotum caused by incomplete drainage of the pampiniform plexus (Fig. 20.2). They are rare in children younger than 10 years. Varicoceles most commonly develop between 10 and 15 years of age and have an incidence of approximately 15% in males. The majority (85% to 95%) of varicoceles are left sided, the result of spermatic venous incompetence secondary to drainage of the left spermatic vein into the renal vein at a right angle.
**Table 20.1 Age-Based Differential Diagnosis of Pediatric Scrotal Masses and Pain**

<table>
<thead>
<tr>
<th>DIAGNOSIS</th>
<th>AGE AT ONSET</th>
<th>PAIN</th>
<th>POSITION OF TESTES, TENDERNESS</th>
<th>SYSTEMIC SYMPTOMS</th>
<th>COMMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Testicular torsion*</td>
<td>All ages; peak onset at 12-18 yr</td>
<td>60% sudden onset; diffuse tenderness</td>
<td>High-riding horizontal lie</td>
<td>Vomiting common; dysuria and fever uncommon</td>
<td></td>
</tr>
<tr>
<td>Testicular appendix torsion</td>
<td>Prepuberty; average age 10 yr</td>
<td>Acute or gradual; focal tenderness</td>
<td>Normal lie; blue dot</td>
<td>Vomiting, dysuria, and fever uncommon</td>
<td></td>
</tr>
<tr>
<td>Epididymoorchitis</td>
<td>&gt;16 yr</td>
<td>Gradual; posterior tenderness</td>
<td>Normal lie</td>
<td>Vomiting uncommon; dysuria and fever common</td>
<td>Adolescent etiology: STD</td>
</tr>
<tr>
<td>Hydrocele</td>
<td>Most common during first year of life</td>
<td>Painless</td>
<td>Normal lie</td>
<td>None</td>
<td>Usually resolves by age 12 mo</td>
</tr>
<tr>
<td>Inguinal hernia (incarcerated hernia, strangulated hernia*)</td>
<td>Most common during first year of life</td>
<td>Pain with incarceration or strangulation</td>
<td>Normal lie</td>
<td>Vomiting, abdominal pain with incarceration or strangulation</td>
<td>Occurs 10 times more frequently in males than in females; right-sided more common</td>
</tr>
<tr>
<td>Varicocele</td>
<td>10-15 yr</td>
<td>Painless, mild discomfort, “pressure or fullness”</td>
<td>Normal lie</td>
<td>None</td>
<td>Associated with infertility; right-sided varicocele should prompt evaluation for tumor causing IVC compression</td>
</tr>
<tr>
<td>Testicular tumor</td>
<td>Rare; most occur in patients &lt;3 yr old</td>
<td>Painless</td>
<td>Normal lie, nontender mass</td>
<td>None unless advanced tumor</td>
<td></td>
</tr>
</tbody>
</table>

IVC, Inferior vena cava; STD, sexually transmitted disease.

*Emergency condition.

**CLINICAL PRESENTATION AND DIAGNOSIS**

Right-sided varicoceles should prompt an evaluation for intraabdominal pathology such as thrombosis or a tumor causing compression of the inferior vena cava. Similarly, sudden onset of a left-sided varicocele should raise suspicion for renal cell carcinoma with obstruction of the left renal vein. The differential diagnosis of varicoceles and other scrotal masses is outlined in Table 20.1.

Varicoceles can cause mild discomfort and may lead to infertility in adult males. They are usually diagnosed on routine physical examination and are characterized by a full hemiscrotum without skin changes and a classic “bag of worms” finding on palpation.

**TREATMENT**

A varicocele is generally an incidental finding in adolescence and is not an emergency. Referral to outpatient urology is appropriate.

**TESTICULAR TORSION**

The pathophysiology, clinical findings, and treatment of testicular torsion in the pediatric population is similar to that in adults. Detailed discussion of torsion can be found in Chapter 111.

**INGUINAL HERNIA**

An inguinal hernia occurs when an intraabdominal organ, usually intestine, herniates into a patent processus vaginalis (see Fig. 20.1). An incarcerated hernia refers to an intestinal loop that is not reducible. A strangulated hernia results when the blood supply to the intestinal loop is obstructed and bowel ischemia ensues.

**EPIDEMIOLOGY AND PATHOPHYSIOLOGY**

Inguinal hernias occur in 15 of every 1000 live births. The incidence in premature and low-birth-weight infants is much higher (approximately 30%), whereas the incidence in males is three to four times higher than that in females. They are most commonly identified during the first year of life, with a peak in diagnosis during the first month. Indirect hernias are more common on the right side (60%) because of later testicular descent into the scrotum. If a left-sided hernia exists, there is a strong possibility that an occult right-sided hernia is present. A family history of hernia, prematurity, or undescended testicle is associated with inguinal hernias.
CLINICAL PRESENTATION

Incarceration is more common with small hernias and occurs frequently during the first 6 months of life, less commonly after 2 years of age, and rarely after the age of 5. Strangulation and perforation can occur within 2 hours of decreased blood flow. Incarceration occurs more frequently in females; however, it is the ovaries, not the intestines, that herniate and become incarcerated.

TREATMENT

Given the high frequency of incarceration in the first year of life, hernias found on routine examination without symptoms should be referred for surgical repair. Approximately 90% of complications can be avoided if surgery is performed within the first month after diagnosis.

IDIOPATHIC SCROTAL EDEMA

Idiopathic scrotal edema is manifested as painless erythema and induration of the scrotum. More than 75% of cases occur in boys younger than 10 years. Two thirds of cases are unilateral.

CLINICAL PRESENTATION

Patients may complain of pruritus. Edema and erythema may extend to the phallus, groin, and abdomen. The testes and epididymis will have no palpable masses, and systemic symptoms are rare.

TREATMENT AND DISPOSITION

Idiopathic scrotal edema is a diagnosis of exclusion. If acute pathology has been excluded, patients can be discharged home with outpatient follow-up. Most cases resolve spontaneously within a few days and do not require specific treatment. The recurrence rate is 21%.

CARCINOMA

Testicular and scrotal cancer accounts for 1% of solid tumors in children. The incidence is increased in patients with bilateral cryptorchidism.

CLINICAL PRESENTATION AND DIAGNOSIS

On examination, a painless unilateral mass can be palpated separately from the testis; there may also be a sensation of fullness, tugging, or increased weight in the scrotum associated with testicular enlargement. A reactive hydrocele is present in 7% to 25% of patients and may lead to a delay in diagnosis. Further examination reveals a firm mass, either smooth or nodular, that does not transilluminate. Lymphadenopathy, petechiae, abdominal masses, hepatosplenomegaly, or gynecomastia may be present. Lymphoma and leukemia can metastasize to the testicles and be manifested similarly.

A complete blood count, urinalysis, urine test for human chorionic gonadotropin (produced by germ cell tumors), and testicular ultrasound images should be obtained.

TREATMENT

Management consists of prompt urologic and oncologic consultation for biopsy and staging.

PHIMOSIS

Phimosis is a constriction of the penile foreskin that results in an inability to retract the prepuce over the glans. A fully retractable foreskin is present in 4% of newborns, 50% of 1-year-olds, 80% of 2-year-olds, and 90% of 4-year-olds. In the remaining cases, the foreskin may not be retractable until puberty.

CLINICAL PRESENTATION

Although most cases of phimosis in children are physiologic, symptoms may include pain, hematuria, and in severe cases, urinary obstruction. Adolescents may complain of pain on erection secondary to tension on the foreskin from glandular adhesions. Phimosis can also result from trauma, infection, chemical irritation, and poor hygiene or as a complication of circumcision. Severe stenosis or obstructive uropathy can result from chronic symptomatic phimosis.

TREATMENT

Because the ability to retract the foreskin is age related, parents should not forcefully retract the prepuce. Good hygiene should be taught. If the child has signs of urinary outlet obstruction, dilation of the meatus by gentle use of forceps is warranted.

Steroid preparations (0.05% to 5% betamethasone cream two to four times daily or 1% hydrocortisone cream two to three times daily) have been used with varying success. Severe phimosis may mandate incision of the dorsal inner foreskin by a urologist.

PARAPHIMOSIS

Paraphimosis is a urologic emergency in which the foreskin of an uncircumcised male is retracted behind the glans penis and acts as a constricting band. The resulting venous and lymphatic congestion precludes returning the foreskin to its normal position, threatens arterial blood flow to the glans penis, and can result in penile necrosis, gangrene, or infarction of the glans over a period of hours to days. In infants and young boys, paraphimosis most commonly results after cleaning by a caretaker.
CLINICAL PRESENTATION AND DIAGNOSIS

Balanitis and balanoposthitis can be caused by trauma; by irritation such as contact dermatitis from urine, soaps, powders, and ointments; or by infection. Causative agents of infectious balanoposthitis include group A streptococci (GAS), anaerobic bacteria, *Candida*, and sexually transmitted infections such as gonorrhea and chlamydial infection. Most children with GAS balanoposthitis have a characteristic moist balanoposthitis caused by a nonretractable prepuce, often in conjunction with a current or recent GAS infection at another site. Signs of group A β-hemolytic streptococcal infection include pain, intense fiery redness, and a moist, glistening transudate or exudate under the prepuce. Streptococcal infection can be diagnosed by rapid antigen detection and culture. Gonorrhea and chlamydial infection without frank urethral discharge are unusual in preschool children; however, after puberty, gonorrhea may be detected in the absence of urethral discharge. In severe cases, cellulitis can extend down the penile shaft. Palpable inguinal adenopathy is often present.

TREATMENT AND DISPOSITION

Noninfectious balanoposthitis should be managed with careful attention to hygiene, use of warm water sitz baths, and avoidance of causative agents. Treatment of infectious balanoposthitis should be tailored to the particular infection: minor, polymicrobial infection can be treated with topical mupirocin or bacitracin; GAS infection should be treated with 10 days of ampicillin, amoxicillin, a cephalosporin, or clindamycin. Children with sexually transmitted infections should be admitted to the hospital and evaluated for sexual abuse. Indications for admission include severe infection and urinary retention.

MEATAL STENOSIS

Meatal stenosis is narrowing of the urethral meatus, usually secondary to recurrent episodes of subclinical meatitis. It can be caused by ammonia diaper irritation in circumcised boys and by recurrent balanoposthitis in uncircumcised boys. Acquired meatal stenosis more commonly occurs in circumcised boys because the foreskin in uncircumcised boys acts as a protective cover for the meatus. Congenital meatal stenosis is rare.

CLINICAL PRESENTATION AND DIAGNOSIS

Obstructive symptoms occur occasionally, including hesitancy, straining, urgency, frequency, and postvoid dribbling. An abnormal urinary stream may be seen, but urinary retention is rare. An erythematous swollen meatus is noted, often with purulent discharge. Radiographic studies are seldom necessary.

TREATMENT

Treatment of purulent meatitis includes sitz baths and the administration of oral antibiotics (e.g., cephalaxin) for 7 days.
Urinary retention is an indication for urology consultation and hospital admission; otherwise, prompt outpatient follow-up is sufficient.

**URETHRAL STRICTURE**

In the United States, most cases of urethral stricture are acquired by infection and trauma. Gonococcal urethritis or infection resulting from an indwelling Foley catheter are the most common infection-related causes. In developing countries, sexually transmitted urethritis is the most common cause. Trauma can result in urethral stricture secondary to pelvic fracture and straddle injury.

**CLINICAL PRESENTATION AND TREATMENT**

Clinically, the patient has difficulty passing urine. Slowing, spraying, or dribbling of the urine stream may be noted. When patients with this condition are seen in the ED, outpatient management, including prophylactic antibiotics, is appropriate. Urethrography or voiding cystourethrography should be performed before dilation of the stricture if this procedure is necessary.

**URETHRAL FOREIGN BODY**

A urethral foreign body in older boys may have been inserted for sexual purposes. Most objects are palpable if they are in the anterior urethra. Retained foreign body should be included in the differential diagnosis of a male with signs and symptoms of recurrent urinary tract infection and no urogenital abnormalities. Most foreign bodies can be removed endoscopically.

**URETHRAL PROLAPSE**

Prolapse of the urethra is most common in young African American girls (Fig. 20.4). The prolapsed mucosa is visible and may be irritated, congested, and hemorrhagic. Though quite alarming on physical examination, it is not associated with sexual abuse. Treatment consists of sitz baths three times a day.

**Fig. 20.4** Two examples of urethral prolapse. A, Urethral prolapse in a 4-year-old girl who had bloody spotting on her underwear. B, Urethral prolapse. (A, From Behrman RE, Kliegman RM, Jenson HB. Nelson’s textbook of pediatrics. 18th ed. Philadelphia: Saunders; 2007. Fig. 544-10; B, from Zitelli BJ, Davis HW. Atlas of pediatric physical diagnosis. 5th ed. Philadelphia: Mosby; 2007. Fig. 14-50.)

**Renal Disorders**

**NEPHROTIC SYNDROME**

Nephrotic syndrome is the clinical manifestation of a variety of primary and secondary glomerular disorders characterized by the following findings:

- Hypoproteinemia (serum albumin <3 g/dL)
- Marked proteinuria (>40 g/m²/hr in a 24-hour period)
- Edema
- Hyperlipidemia (predominantly triglycerides and cholesterol)

**PATHOPHYSIOLOGY**

Nephrotic syndrome is classified as primary, defined as nephrotic syndrome without systemic disease, or secondary, defined as nephrotic syndrome in the presence of systemic disease. Ninety percent of children with nephrotic syndrome have the primary syndrome, the most common of which is minimal change disease (idiopathic nephrotic syndrome).
Idiopathic nephrotic syndrome is classified by its response to corticosteroids, with the majority being steroid responsive and approximately 20% being unresponsive to steroids. Lack of response to steroids portends a poor prognosis and a 50% risk for progression to end-stage renal failure.

**EPIDEMIOLOGY**

The incidence of nephrotic syndrome in children younger than 16 years is 2 to 7 per 100,000 per year, and the prevalence is 15 cases per 100,000. Boys are more likely to be affected than girls. The typical age at onset of primary nephrotic syndrome is 18 months to 6 years. Children with nephrotic syndrome at an age older than 5 years are more likely to have the secondary form. Neonates are most likely to have the congenital (Finnish) type, which is steroid resistant and generally fatal. In adolescents, nephrotic syndrome is most often associated with a primary or secondary form of an underlying nephritis.

**CLINICAL PRESENTATION**

The usual initial sign in a child with nephrotic syndrome is edema. The edema starts with early-morning periorbital swelling, often misattributed to a cold or allergies. As the edema spreads to the abdomen, trunk, and extremities, children have increasing difficulty fitting into their pants and shoes. Parents can mistake this for weight gain. The child otherwise appears well, although ascites, pleural effusion, or pulmonary edema may be present. Ascites and an edematous intestinal wall can be manifested as abdominal pain, nausea, vomiting, or diarrhea.

**COMPLICATIONS**

Complications include infection, hypercoagulability, hypovolemia, respiratory distress, and acute renal failure.

**INFECTION**

Increased susceptibility to infection results from disease-mediated impairment of the immune system, as well as treatment with immunosuppressive therapy, both of which may mask the typical signs of infection. The most common bacterial infection is peritonitis, although cellulitis, pneumonia, sepsis, and meningitis are also seen. Infection with encapsulated bacteria, such as *Escherichia coli*, *Haemophilus influenzae*, and *Streptococcus pneumoniae*, is the main cause of death in children with nephrotic syndrome.

**HYPERCOAGULABILITY**

Hypercoagulability occurs in 3% of patients with nephrotic syndrome. Thromboembolic events can involve the arteries and veins, most commonly the pulmonary arteries, renal veins, and deep leg veins. A sudden onset of gross hematuria or renal failure should prompt investigation for renal vein thrombosis.

Because prednisone exerts an antithrombin effect, practitioners should not attempt deep venous punctures on children treated with steroids unless no alternative exists.

Do not attempt deep venous punctures in children who have nephrotic syndrome and are receiving chronic prednisone therapy because they are at increased risk for thromboembolic events.

**RESPIRATORY DISTRESS**

Pleural effusions, pulmonary edema, and massive ascites can cause respiratory distress.

**TREATMENT**

The goals of ED management are restoration of volume, treatment of symptomatic edema, and evaluation for and treatment of infectious complications. Patients in shock should be treated by isotonic hydration consisting of a 20-mL/kg bolus per hour until they are normotensive. If the patient is clinically dehydrated with hemoconcentration but not in shock, a trial of orally administered sodium-deficient fluids at twice the maintenance dose is preferable to intravenous solutions. Small amounts of oral hydration fluid should be administered frequently to avoid vomiting caused by an edematous gut.

If the patient is well hydrated and exhibits symptomatic edema or anasarca, diuretics may be warranted but should be used judiciously because these children have decreased circulating volume and are prone to thromboembolic events. Intra-venous or oral administration of furosemide, 1 to 2 mg/kg/24 hr divided into two doses, can be used. Loop diuretics are most effective, but additional diuretics such as hydrochlorothiazide or spironolactone may be administered if the response is inadequate. Diuretics are not effective with albumin concentrations of less than 1.5 g. Albumin infusions may be necessary before administration of diuretics (0.5 to 1 g/kg given as 25% salt-deficient albumin, followed by 0.5 to 1 mg/kg of furosemide).

Although patients may arrive in shock from intravascular volume depletion alone, sepsis should be excluded. Any child with nephrotic syndrome and an unexplained fever must be considered to have a bacterial infection until proved otherwise. Because persons receiving steroid therapy may not demonstrate abdominal pain or typical signs of infection, diagnostic paracentesis is necessary to exclude bacterial peritonitis in children with abdominal pain or fever in the setting of ascites. Treatment with cephalosporins or ampicillin (with or without gentamicin) is recommended. Prophylactic antibiotics are not necessary unless infection is suspected, and material should be collected for culture.

The mainstay of treatment of nephrotic syndrome is steroid therapy. Generally, patients initially receive prednisone, 2 mg/kg/24 hr (maximum, 60 mg/24 hr) divided into two or three doses. Approximately 90% of patients with idiopathic nephrotic syndrome respond to steroid therapy by the end of a 4-week course, with response defined as trace or negative amounts of urine protein for 3 days. Failure to respond to steroid treatment increases the likelihood of a cause other than minimal change disease.
Hemolytic-uremic syndrome is the most common cause of acute renal failure in children, with an incidence of 1 to 10 cases per 100,000 children younger than 5 years. The mean age at diagnosis is 3 years, and the diagnosis becomes increasingly unlikely after 5 years of age. Caucasian children are more often affected than others, and there is no gender preference.

PATHOPHYSIOLOGY

Hemolytic-uremic syndrome is defined by the presence of the classic triad of microangiopathic hemolytic anemia (MAHA), thrombocytopenia, and acute renal failure (Box 20.1).

In contrast to the adult form of the disease (thrombotic thrombocytopenic purpura), the microthrombi of hemolytic-uremic syndrome are usually confined to the kidneys. Thrombotic thrombocytopenic purpura has a predominantly neurologic manifestation, a higher mortality rate, and a better response to plasmapheresis and fresh frozen plasma. Renal involvement is the defining feature of hemolytic-uremic syndrome.

The usual cause of epidemic hemolytic-uremic syndrome is the verotoxin-producing strain of *Escherichia coli* serotype O157:H7, although it can be caused by *Shigella* organisms that produce a similar toxin. Transmission is through person-to-person contact or ingestion of contaminated food, such as unpasteurized dairy products or undercooked beef. The verotoxin binds to and destroys the colonic mucosa, thereby leading to bloody diarrhea.

**BOX 20.1 Hallmarks of Hemolytic-Uremic Syndrome**

- Acute renal failure
- Microangiopathic hemolytic anemia
- Fever
- Thrombocytopenia

**CLINICAL PRESENTATION**

The epidemic form begins with a prodrome of nausea, vomiting, watery diarrhea, crampy abdominal pain, and occasionally fever. Bloody stools typically develop on the second or third day of symptoms. Usually between 5 and 10 days after the prodromal gastroenteritis, there is a sudden onset of pallor, listlessness, irritability, and oliguria because of development of the classic triad (MAHA, thrombocytopenia, renal failure). Additional signs on clinical examination may include dehydration, edema, hypertension, petechiae, hepatosplenomegaly, jaundice, and neurologic manifestations (obtundation, hemiparesis, seizures, brainstem dysfunction).

Gastrointestinal complications such as toxic megacolon, ischemic colitis, intussusception, and perforation are also possible. Pancreatic insufficiency from microinfarction in the pancreas can lead to permanent insulin-dependent diabetes mellitus.

**DIAGNOSTIC TESTING**

Hemolytic-uremic syndrome is diagnosed clinically by symptoms coupled with consistent laboratory findings. The presence of MAHA is necessary to establish the diagnosis. The peripheral blood smear demonstrates signs of a microangiopathic process: teardrop cells, helmet cells, spherocytes, and burr cells.

A complete blood count may show leukocytosis, profound anemia (with hemoglobin levels of 5 to 9 g/dL), and mild to moderate thrombocytopenia (platelet counts are generally around 40,000/mm³).

C-reactive protein levels may be elevated. The coagulation profile is usually normal, and although serum fibrin split products might be elevated, fulminant disseminated intravascular coagulation is rare. Chemistry abnormalities include hypocalcemia, hyperkalemia, azotemia, metabolic acidosis, hyperbilirubinemia, low total protein as a result of proteinuria, and elevated lactate dehydrogenase. Urinalysis often shows hematuria, proteinuria, and pyuria. Granular and hyaline casts are seen in the urine sediment.

Specific serologic testing for antibodies to the lipopolysaccharide of *E. coli* O157:H7 is necessary because routine stool cultures may not always detect the bacteria or verotoxin.

**TREATMENT AND DISPOSITION**

Severe anemia, defined as a hemoglobin concentration lower than 6 g/dL, requires transfusion of packed red blood; platelet transfusion is recommended only with active bleeding or in preparation for a required invasive procedure.

Indications for dialysis in children with hemolytic-uremic syndrome are similar to those with renal failure: signs and symptoms of uremia, azotemia, severe fluid overload, and electrolyte disturbances not responsive to medical therapy. Rehydration should be done slowly to avoid fluid overload.

Hypertension is generally responsive to the administration of calcium channel blockers, labetalol, captopril, hydralazine, or in refractory cases, nitroprusside. Seizures will respond to benzodiazepines and phenytoin; however, hyponatremic seizures do occur and should be evaluated for and treated with...
except for the change in urine color. Rarely, patients with stolic) may be found. Some children may be asymptomatic ties. On examination, hypertension (both systolic and dia-

hematuria and proteinuria. The patient has had a sudden onset of brown,
streptococcal infection or 3 to 6 weeks after the onset of the glomeruli. The latency period for both the pharyngeal and
ence.

The signs and symptoms of acute glomerulonephritis are pre-

The most common form of acute glomerulonephritis in chil-

The family should be advised to monitor the patient’s urine
output and weight and observe for signs of congestive heart
 failure or hypertension. In general, the prognosis is excellent. Approximately 80% to 90% of these children recover without any persistent renal abnormalities except for microscopic hematuria, which may persist for up to 18 months.

IgA NEPHROPATHY

IgA nephropathy, also known as Berger disease, is the type of glomerulonephritis most commonly diagnosed in adolescence. This disease accounts for up to 25% of cases of glomerulonephritis in Asia and Europe and up to 10% in the United States.

CLINICAL PRESENTATION AND DIAGNOSIS

The classic finding is hematuria or proteinuria with a preceding upper respiratory infection. The diagnosis is confirmed by a renal biopsy specimen with deposition of IgA in the mesangium.

TREATMENT AND DISPOSITION

Gross hematuria resolves spontaneously within days without serious sequelae in 85% of patients. Because the prognosis is good in most patients without significant proteinuria or renal dysfunction, treatment is not usually needed. However,
proteinuria, hypertension, or renal insufficiency portends a poor prognosis and should be managed in consultation with nephrology.

**HENOCH-SCHÖNLEIN PURPURA WITH RENAL INVOLVEMENT**

Henoch-Schönlein purpura is the most common form of small-vessel vasculitis in childhood. Most patients have the triad of abdominal pain, arthritis, and purpura. A third of the patients have renal involvement, 80% of whom will have asymptomatic hematuria. The populations most affected are school-age children and young adults, and it occurs more commonly in Caucasians and males. One third to three fourths of patients have a preceding respiratory infection.

**CLINICAL PRESENTATION**

Usually occurring within the first month of illness, renal involvement develops in approximately one third of patients, with progression to end-stage renal disease in less than 1%. Predictors of a poor prognosis include late onset of renal involvement in older children and massive proteinuria; 20% of such cases result in end-stage renal failure. Approximately 50% of patients in whom nephritic syndrome with Henoch-Schönlein purpura develops will progress to end-stage renal disease within 10 years.

The diagnosis is made clinically and not based on laboratory studies.

**TREATMENT**

Prednisone, 1 to 2 mg/kg/day for 2 weeks (max 60 mg/day), with tapering of the dose over a 2-week period, has been shown to improve gastrointestinal and joint symptoms and lessen the severity of nephritis. About one third of affected patients will have a recurrence of at least one symptom. Fortunately, most patients recover quickly in several weeks with supportive treatment.

**RENAL TUBULAR ACIDOSIS**

The normal response to acidemia is to reabsorb all the filtered bicarbonate and to increase excretion of hydrogen, primarily by excreting ammonium ions in urine.

Renal tubular acidosis occurs when the renal tubules are unable to perform these functions. Accumulation of ammonium ions and subsequent metabolic acidosis can cause growth retardation, kidney stones, bone disease, and progressive renal failure.

Four subtypes of renal tubular acidosis are recognized:

- **Type 1**—distal (classic form)
- **Type 2**—proximal (bicarbonate wasting)
- **Type 3**—a combination of types 1 and 2
- **Type 4**—hyperkalemic (rare in children)

Types 1 and 2 are encountered most frequently in children. The diagnosis of all types requires a serum electrolyte panel and urinalysis with urine pH.

**TYPE 1 RENAL TUBULAR ACIDOSIS**

Distal renal tubular acidosis results from a defect in the tubular transport of hydrogen in the distal nephron. The most common form in children is hereditary, but it can be a complication of systemic diseases seen more commonly in adults such as Sjögren syndrome, lupus erythematosus, and hyperparathyroidism. Patients have failure to thrive, anorexia, vomiting, and dehydration. Hyperchloremic metabolic acidosis and hypokalemia may be seen. Hypercalciuria can be manifested as rickets, nephrocalcinosis, nephrolithiasis, and renal failure. Urine pH usually exceeds 6.5.

The classic diagnostic test for distal renal tubular acidosis is an acid load from ammonium chloride; however, this is a tedious test that can generate severe acidosis. Distal renal tubular acidosis can be permanent, but children may outgrow it by the time that they are of school age. Treatment is focused on correction of the acidosis with sodium bicarbonate or sodium citrate, which may also prevent kidney stone formation. Target serum HCO₃ levels should be between 20 and 22 mEq/L in infants and between 22 and 26 mEq/L in children.

**TYPE 2 RENAL TUBULAR ACIDOSIS**

Proximal renal tubular acidosis is the most common form found in children. Because approximately 85% to 90% of bicarbonate reabsorption occurs in the proximal tubules, proximal renal tubular acidosis is characterized by an alkaline urine (usually pH >7), loss of bicarbonate in urine, and mildly reduced serum bicarbonate concentration.

Proximal renal tubular acidosis can result from inherited disorders (hereditary fructose intolerance, Wilson disease, cystinosis, Fanconi syndrome, Lowe syndrome), exposure to medications (chemotherapy agents, acetazolamide, sulfonamides), anatomic abnormalities (obstructive uropathy, reflux), or exposure to heavy metals. Treatment consists of alkaline therapy with citrate solutions (Bicitra, Polycitra) or sodium bicarbonate. Potassium supplements may also be required because the added sodium load of the sodium bicarbonate may increase potassium loss in the distal tubule.

**URINARY RETENTION**

Urinary retention is defined as failure to urinate for more than 12 hours. More than 90% of all newborns void within the first 24 hours of life and 99% within the first 48 hours. In male children, posterior urethral valves are the most common cause of retention. Other causes include urethral polyps, urethral stricture, urethral diverticulum, meatal stenosis, and fecal impaction. In female infants, the differential diagnosis includes prolapsing ureterocele, urethral prolapse, and foreign bodies. Infections, medications, spinal cord abnormalities, and sexual abuse can cause retention in both male and female children. Diagnostic tests in the ED include blood urea nitrogen, creatinine, and urinalysis.

**REFERENCES**

References can be found on Expert Consult @ www.expertconsult.com.
REFERENCES