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Renal calculi in children: imaging features that lead to diagnoses: a pictorial essay

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Renal stones in children are often associated with radiologic findings that lead one to deduce the cause of stone formation. The images presented here show pediatric patients with renal stone disease who have associated imaging findings that should enable one to form an association or "trigger a synapse" between the characteristic associated finding and the cause of the renal stone. It is the elucidation of these associations or imaging features that is the focus of this pictorial essay.

Approximately two-thirds of urinary calculi in North American children are located in the kidney at the time of diagnosis. The other third are in the ureter and the bladder. They are rarely in the urethra. Renal stones occur equally in males and females. Previously, there were no disproportionately affected age groups [1]. However, with recent advances in the care of premature infants, an increasing number of patients with hyaline membrane disease treated with furosemide has resulted in an increased incidence of stones in infants [2].

Symptoms of renal stone disease vary with age. The traumatic incapacitating flank, abdominal, or pelvic pain associated with the passage of stones is unusual in children. However, pain does occur in about half of patients with renal stones [1, 3]. Pain is more commonly the presenting problem in adolescence, whereas preschool children more commonly present with infection. Microscopic or macroscopic hematuria has been reported in 33% to 90% of children with urinary calculi [1]. The typical causes of renal stone disease in children include urologic conditions causing urinary stasis, urinary manifestations of myelodysplasia, recurrent urinary tract infection with urea-splitting bacteria, metabolic abnormalities, immobilization, enteric diseases, and iatrogenic causes [4, 5].

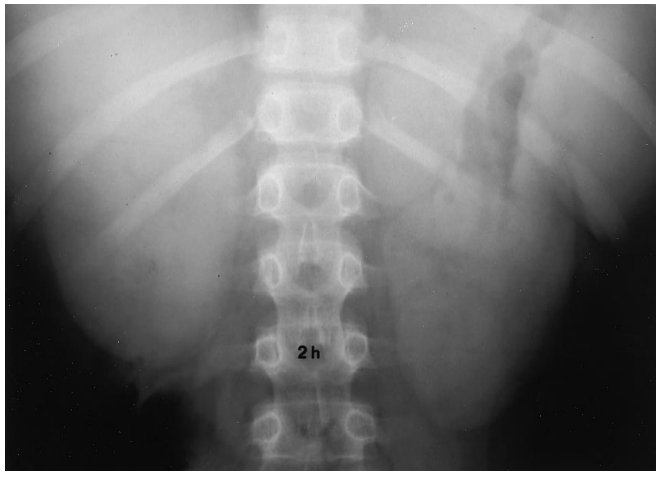


Fig. 1 Acute lymphocytic leukemia on treatment. Persistent nephrograms on 2-h radiograph from excretory urogram represent diffuse tubular block. An abundance of uric acid crystals was found in the urine (the right kidney is smaller than the left because of an unrelated right upper pole nephrectomy in infancy)

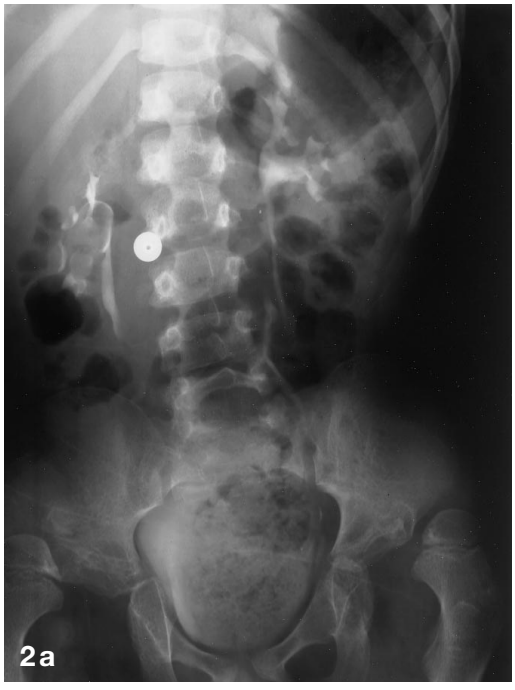
Radiolucent stones

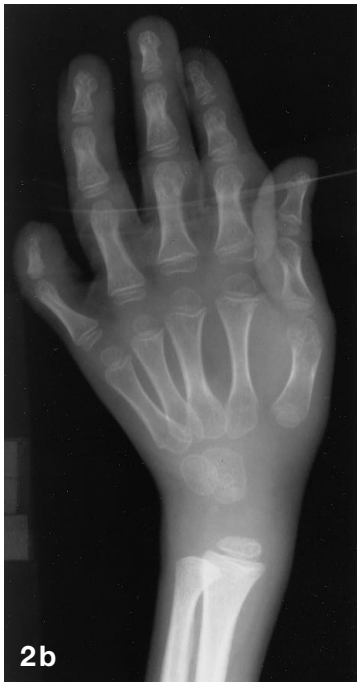
When the stone is seen on ultrasound or CT but is nonopaque on plain film, the differential diagnosis in children includes uric acid, cystine, and xanthine stones.

Uric acid stones are found in disorders of purine metabolism such as Lesch-Nyhan syndrome. Additionally, uric acid crystals can be associated with treatment of myeloproliferative disorders when there is hyperuricemia secondary to increased purine meta-

Fig. 4 Cystinuria. Renal sonogram shows shadowing foci in the right kidney. These were cystine stones. Plain radiograph showed no opaque calculi ▶

Fig. 5 a, b Cystinuria before and after treatment. **a** Faintly opaque calculus fills the lower calyces. **b** After treatment with penicillamine, the cystine stone has completely disappeared





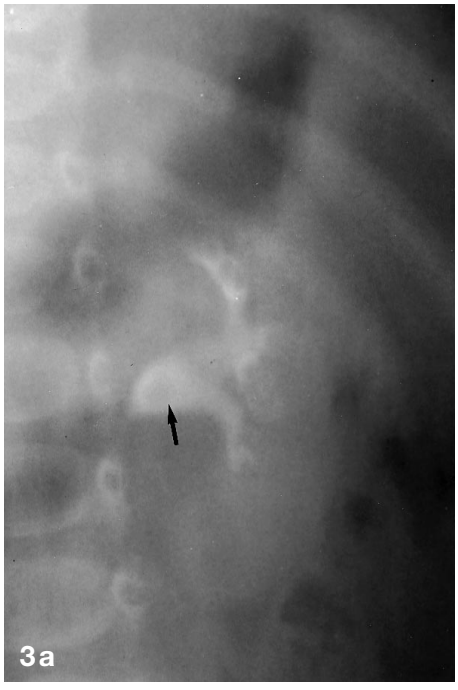
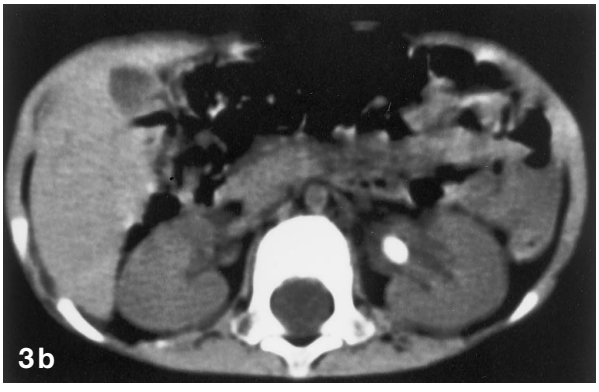
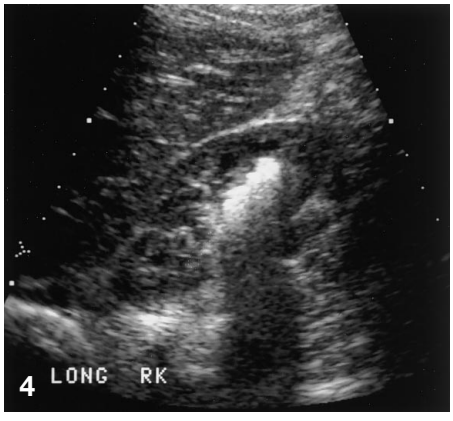
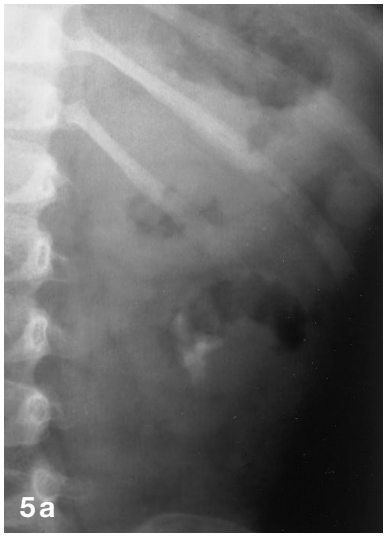


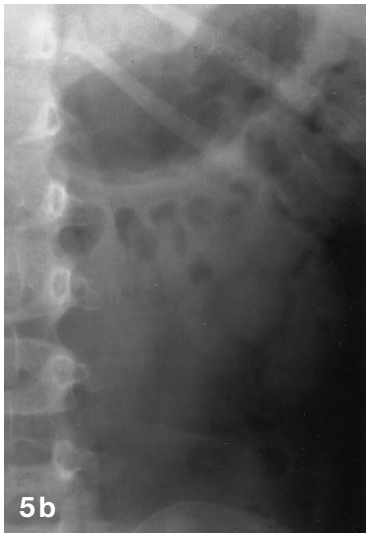
Fig. 2a,b Lesch-Nyhan syndrome. **a** IVP. Scout radiograph revealed no opaque calculi (not shown). Pyelogram shows a large filling defect, a nonopaque uric acid stone, in the right renal pelvis. Both hips are dislocated. **b** Hand radiograph shows the absence of the distal phalanx of the small finger. It had been bitten off by the patient (self-mutilation)

Fig. 3a,b Hereditary xanthinuria from deficiency of the enzyme xanthine oxidase. **a** IVP. Plain radiograph revealed no calculi (not shown). Excretory urogram shows a filling defect in the left renal pelvis *(arrow)*, a xanthine stone. **b** Nonenhanced CT scan. The xanthine stone is well seen in the left renal pelvis









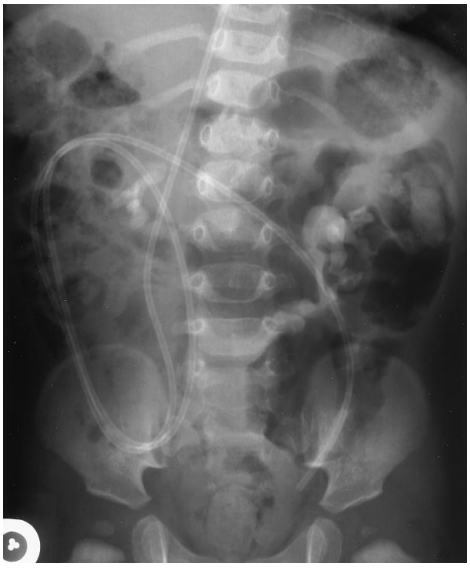
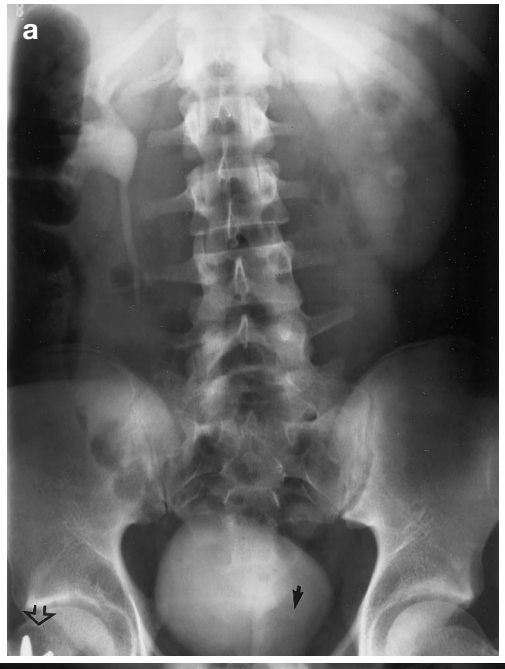


Fig. 6 Infection stone. Patient with ventriculo-peritoneal shunt, neurogenic dysfunction of the bladder, and infection of the urine with *Proteus.* Radiograph shows bilateral staghorn calculi



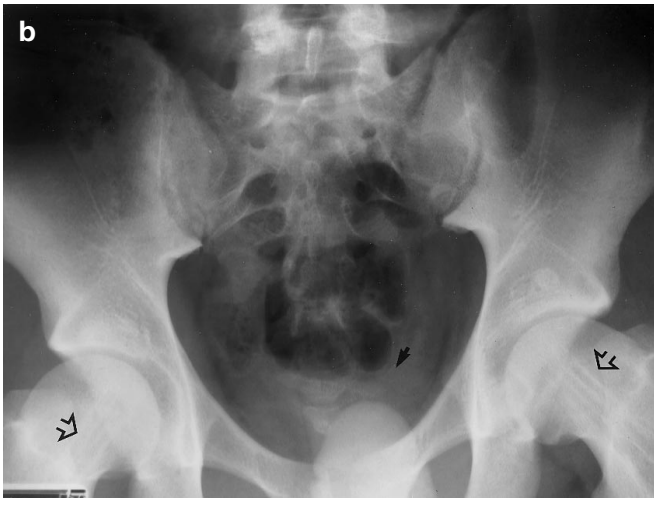


Fig. 7 a, b Immobilization calculi. **a** Excretory urogram shows left ureteral obstruction by a stone *(arrow)* near the end of the edematous distal left ureter. There is elevation of the left hemitrigone. The patient had been immobilized in the past for pinning of right slipped capital femoral epiphysis. The ends of the threaded pins can be seen *(open arrow).* **b** Left flank pain and hematuria led to this radiograph of the pelvis which showed a calculus in the region of the distal end of the left ureter *(arrow).* The patient had been immobilized in the past for pinning of slipped capital femoral epiphyses. The pins have been removed, but the pin tracks *(open arrows)* are still visible

bolism. Cell breakdown following chemotherapy increases the likelihood of uric acid crystal formation. This is seen urographically as diffuse tubular block [5] (Fig. 1). Uric acid stones are soluble in alkaline urine, of clinical importance in their treatment.

The Lesch-Nyhan syndrome, also called congenital hyperuricosuria, is a hereditary basal ganglia disorder in children that is characterized by choreoathetosis, mental retardation, and self-mutilation. It is caused by a defect in purine metabolism [5]. Plain radiographs frequently show signs of self-mutilation (such as truncated, chewed-off fingertips), and neuropathic arthropathy of the hips, but not the lucent uric acid urinary stones (Fig. 2).

Xanthine stones (xanthose [Greek] = yellow) occur in the rare autosomal recessive deficiency of xanthine oxidase that results in increased urinary excretion of xanthine and hypoxanthine, resulting in stone formation. This type of stone is most often seen in patients of Lebanese descent [7] (Fig. 3).

Cystine stones [cyst (Latin) = bladder] were first identified from bladder stones, hence the name. An autosomal recessive disorder of amino acid transport in the renal tubule causes impaired reabsorption of cystine, ornithine, lysine, and arginine [8]. The only recognized clinical consequence is cystine urolithiasis, which typically begins in the second decade of life (Fig. 4). Cystine stones are soluble in alkaline urine and are either nonopaque or faintly opaque on plain radiographs. Imaging before and several months after "competitive binding" penacillamine therapy often demonstrates disappearance of the stones (Fig. 5).

Infection stones

In a patient with urinary calculi and urinary infection, *Proteus* is the most likely organism and *Proteus mirabilis* is the most common organism associated with renal stones [9] (Fig. 6). *Proteus* and some species of *Pseudomonas, Klebsiella, Escherichia coli,* and *Staphylococcus* produce urease, which catalyzes conversion of urea to ammonia. Ammonia hydrolyzes to form OH- and NH_4^+, making urine alkaline and in turn increasing the concentration of PO_4^{3-}. A supersaturated solution of magnesium ammonium phosphate results, and stones form. High urine pH also promotes crystallization of hydroxyapatite, which is commonly incorporated into the stone [5]. Recently, nanobacteria, the smallest known bacteria with a cell wall, a new type of bacteria in both human and cow blood, has been shown to produce a structure that can precipitate

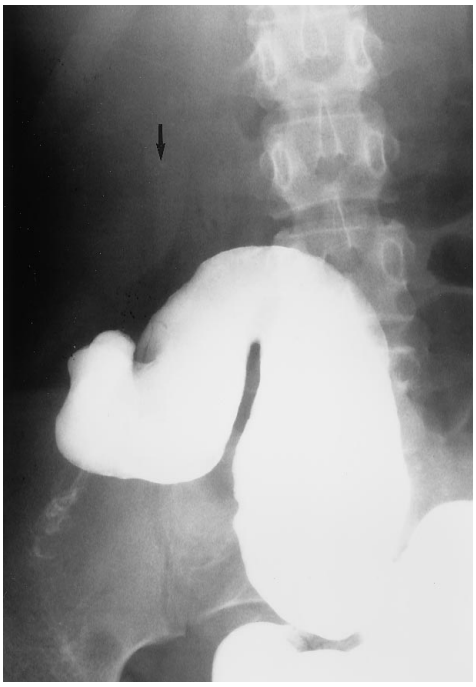


Fig. 8 Crohn disease. This girl with Crohn disease had resection of the diseased portion of the right colon and terminal ileum. On this postoperative barium enema, a small calculus was present in the right renal pelvis *(arrow)*

carbonate apatite crystals, found in the core of many kidney stones [10].

Stones associated with immobilization

Stones that form in the kidneys of immobilized patients are caused by the hypercalcemic/hypercalciuric state due to the mobilization of calcium from bone [11]. Immobilization can be secondary to a variety of causes such as fracture, slipped capital femoral epiphysis, prolonged bed rest for any reason, and also exposure to the microgravity environment of space flight [12]. Radiographs may show stones as early as 8 days after immobilization [11]. Often associated imaging findings enable one to suggest immobilization as the cause of the stone (Fig. 7). Examples include orthopedic hardware or their sequelae, and disuse osteopenia in an immobilized extremity.

Inflammatory bowel disease (IBD)

In children with inflammatory bowel disease such as Crohn disease, stone formation is due to multiple factors including treatment with steroids as well as bowel dysfunction from inflammation and ileostomy [13]. Bowel dysfunction leads to altered oxalate metabolism. Excessive intestinal fat binds intraluminal calcium ions that would normally combine with oxalate to form insoluble calcium oxalate and be eliminated in the feces. Instead, oxalate remains soluble and is absorbed in excessive amounts [13]. Hyperoxaluria and urinary tract calcium oxalate stone formation follow. A plain radiograph of the abdomen showing an opaque renal stone plus osteopenia and its sequelae, sacroiliitis, and/or an ostomy overlying the abdomen should lead one to suspect IBD as the possible etiology for the stone (Fig. 8).

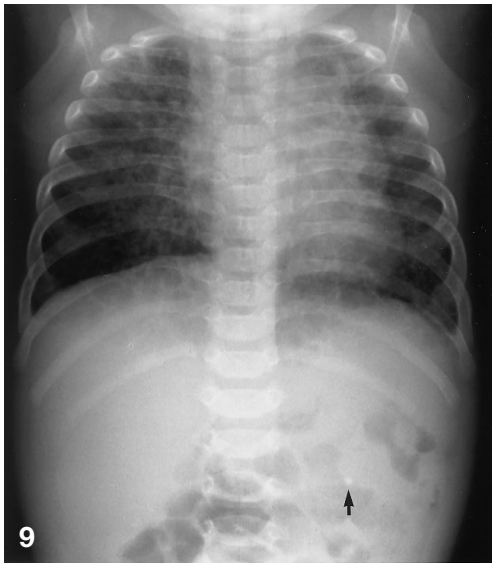


Fig. 9 Furosemide (Lasix) stone. Radiograph of chest and upper abdomen of premature infant shows chronic lung disease of prematurity. The baby had hyaline membrane disease and was treated with furosemide (Lasix). There is a stone in the left kidney *(arrow)*

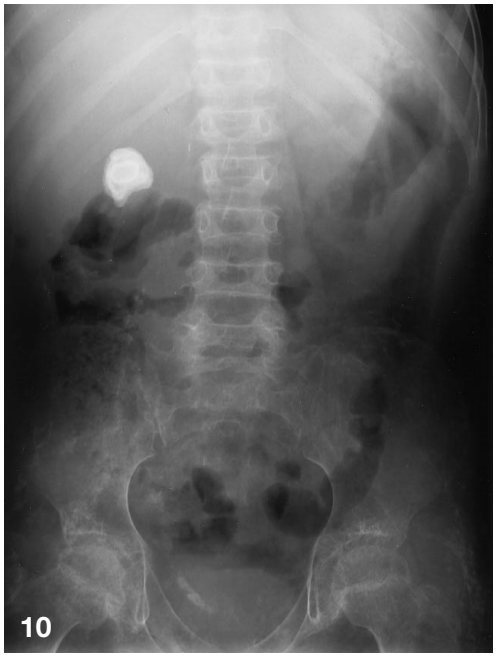


Fig. 10 Dermatomyositis on steroids. On this abdominal radiograph, there is a laminated renal calculus that fills the right renal pelvis. Additionally, there are soft-tissue calcifications. The bones are osteopenic and the vertebral bodies are partially collapsed as a result of the steroid therapy

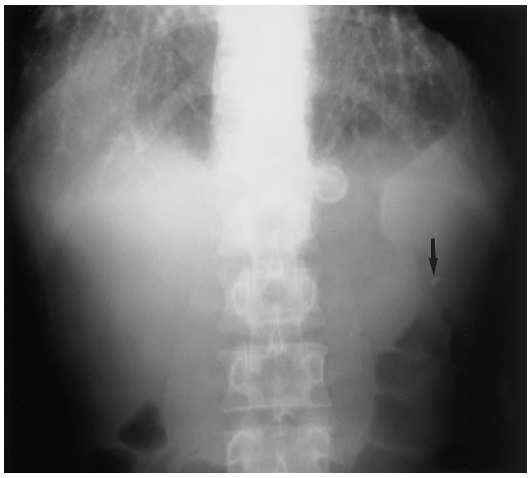


Fig. 11 Cystic fibrosis. Abdominal radiograph including the lower lungs shows a gastrostomy tube in the stomach. Chronic changes of cystic fibrosis are seen in both lung bases. There is a left renal calculus *(arrow)*

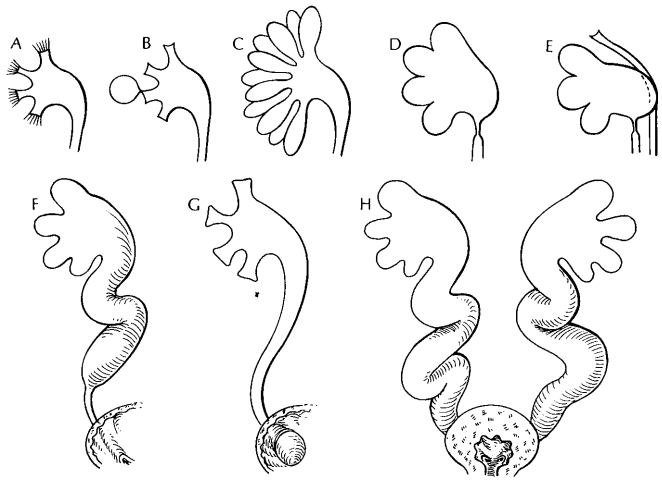


Fig. 12 Diagrammatic representation of the congenital anomalies that lead to stasis of urine and stones. *A* Tubular ectasia (medullary sponge kidney); *B* calyceal diverticulum; *C* congenital megacalyces; *D* ureteropelvic junction obstruction; *E* ureteropelvic junction obstruction of lower pole of duplex kidney; *F* primary megaureter; *G* ureterocele; *H* posterior urethral valves

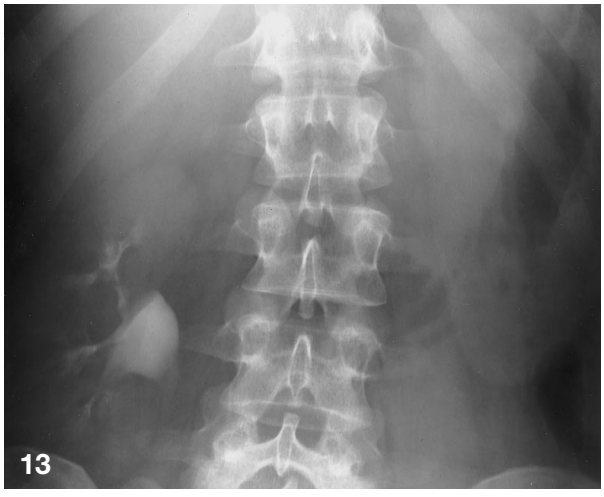
Fig. 13 Stasis stone. Tubular ectasia. IVP in patient with tubular ectasia (medullary sponge kidney) who presented with left flank pain and hematuria. The right kidney shows tubular blush typical for medullary sponge kidney. On the left, there is an obstructed nephrogram. Subsequently, a small left calculus was passed

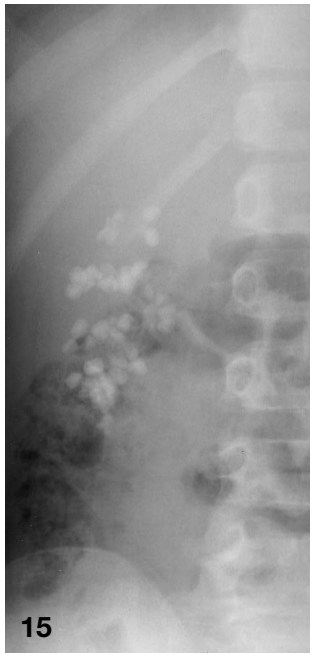
Fig. 14 Stasis stones. Calyceal diverticulum. Tomogram of right side of abdomen shows a cluster of calculi in a calyceal diverticulum

Fig. 15 Stasis stones. Congenital megacalyces. Plain radiograph of the right kidney shows stones in many of the dilated calyces

Medication-induced stones

Patients being treated for many illnesses may be taking drugs that cause renal stone formation. At the initial stage of the disease, radiographs may be normal. After ongoing therapy, renal stones may develop. Stones are now frequently seen in low-birth-weight premature infants treated for prolonged periods (usually at least 2–3 weeks) with furosemide for chronic lung disease of prematurity or heart failure [2] (Fig. 9). Furosemide leads to decreased tubular reabsorption of calcium, causing hypercalciuria. Calcium phosphate and calcium oxalate stones result. Most recently, stones





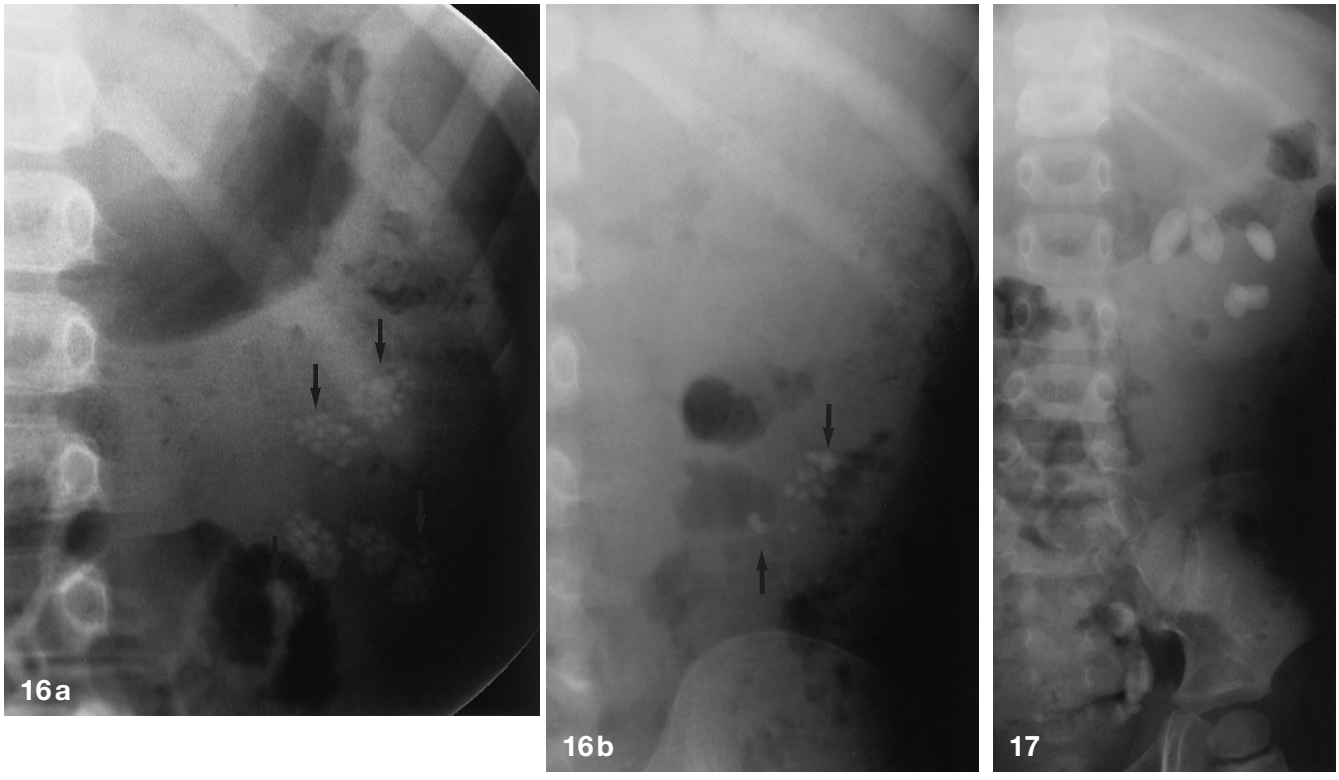


Fig. 16a, b Stasis stones. Ureteropelvic junction obstruction. **a** UPJ obstruction in a kidney with a single collecting system. Numerous stones have formed in many of the dilated calyces *(arrows)*. **b** UPJ obstruction of lower pole of left duplex kidney. Stones have formed only in the dilated lower pole calyces *(arrows)*

Fig. 17 Stasis stones. Left primary megaureter. Radiograph shows large stones in the calyces, renal pelvis, and distal ureter

have been seen as the result of treatment with indinavir sulfate (a protease inhibitor) in patients with AIDS [14]. The patient in Fig. 10 has soft-tissue calcifications from dermatomyositis and was being treated with steroids.

Cystic fibrosis

In addition to the well-recognized lung manifestations of the disease, disease-related complications include pancreatic insufficiency, malabsorption, and consequent malnourishment, which may result in formation of renal stones [15, 16] (Fig. 11).

Stones associated with urinary stasis

Stasis favors stone formation by causing retention of crystal aggregates and other potential niduses. Stones formed are usually calcium phosphate, although any of the crystals may precipitate. Stasis can occur along the urinary axis from the renal tubule to the urethra. Conditions in children associated with stone formation due to urinary stasis include (Fig. 12):

Tubular ectasia (medullary sponge kidney) (Fig. 13)
Calyceal diverticula (Fig. 14)
Congenital megacalyces (Fig. 15)
UPJ obstruction; single system or lower pole of duplex kidney (Fig. 16a, b)
Primary megaureter (Fig. 17)
Ureterocele (Fig. 18a, b)
Posterior urethral valves (Fig. 19)

In conclusion, the images presented will assist in the formation of associations between the characteristic findings of renal stones on imaging studies and the causes of the stones.

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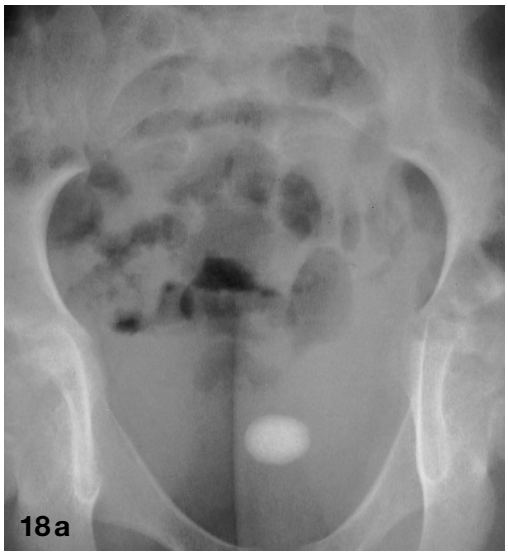

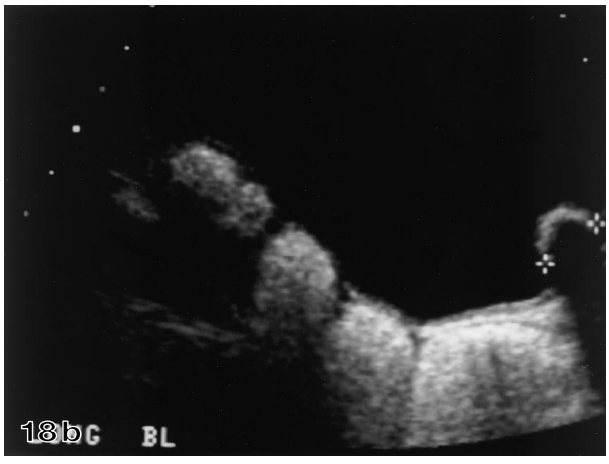

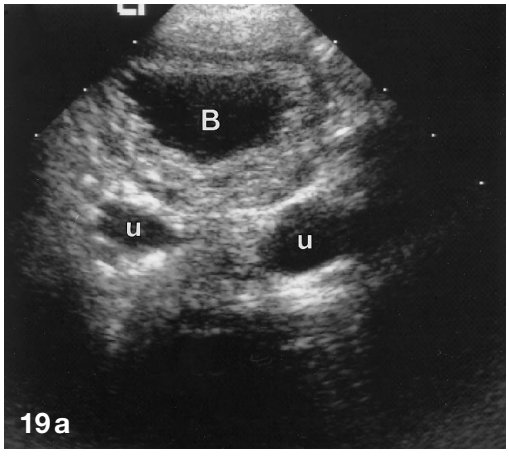

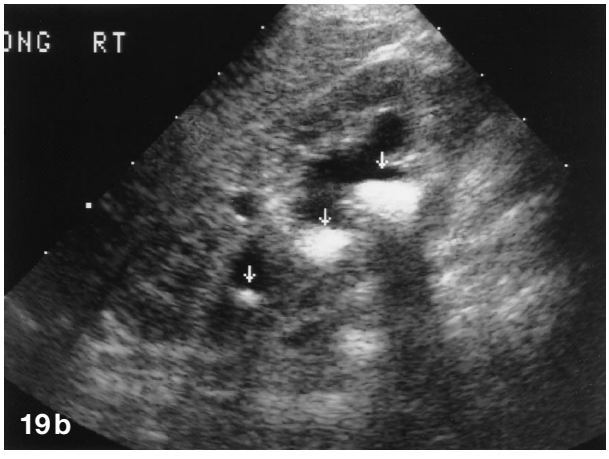

Fig. 18 a, b Stasis stone. Simple ureterocele. **a** Plain radiograph of pelvis shows a laminated calculus on the left side of the pelvis near the midline. **b** Sonogram of the urinary bladder shows the stone that casts an acoustic shadow. The stone did not move with change in position. At cystoscopy, it was found to be in the ureterocele at the end of the single left ureter

Fig. 19 a, b Stasis stones. Posterior urethral valves. **a** Sonogram of the urinary bladder *(B)* shows a thick bladder wall and bilateral dilated ureters *(U)*. **b** Sonogram of right kidney shows shadowing foci (calculi) in many of the dilated calyces *(arrows)*. The left kidney (not shown) had a similar appearance

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