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Acquired renal cystic disease

Acquired renal cystic disease

Most patients on haemodialysis develop bilateral renal cysts - after 10 years. On follow-up one-fifth of these patients with - acquired renal cystic disease (ARCD) develop renal cancers. -

Antenatal hydronephrosis

Antenatal hydronephrosis

The prevalence of antenatal hydronephrosis (ANH) ranges from 0.6% to 5.4%. The majority of cases of ANH are transient and resolve after birth. The optimal timing of postnatal US in patients with ANH is at least 48 hours after birth. Diuretic renography can be performed after 4–6 weeks of life. ANH is classified into low , intermediate and high risk. A voiding cystourethrogram (VCUG), antibiotic prophylaxis and functional scan are recommended for high-risk infants along with monthly follow-up, whereas 1- to 3-monthly follow-up with US may su ffi ce in low-risk infants.

Autosomal dominant polycystic kidney disease

Autosomal dominant polycystic kidney disease

Autosomal dominant polycystic kidney disease (ADPKD) is the most common autosomal dominant genetic cystic renal disease causing chronic renal failure requiring dialysis and renal trans plantation. It occurs as a result of mutation in one of two genes (PKD1 on chromosome 16 and PKD2 on chromosome 4). ADPKD gene loci can be identified in individuals with a family history before the development of cysts begins; this is helpful in screening a potential sibling for kidney donation. ADPKD has variable penetration and approximately 50% of a ff ected individuals eventually develop end-stage renal disease (ESRD). Risk factors for the development of ESRD are: /uni25CF early age of presentation; /uni25CF hypertension; /uni25CF male sex; /uni25CF ADPKD gene 1; /uni25CF African ethnic group. ADPKD is associated with cysts in other organs, such as the liver, pancreas, arachnoid membranes and seminal vesi cles. It does not usually manifest before the age of 30 years and in some patients it is never diagnosed. Renal symptoms include abdominal pain, haematuria or a palpable mass. Most patients older than 20 years are hypertensiv e and good control of blood pressure can delay progression to renal fail ure. Novel agents such as vasopressin antagonists (tolvaptan), somatostatin analogues and mammalian target of rapamycin (mTOR) inhibitors have shown potential to prevent cystogen esis, cyst expansion and declining r enal function. Intracranial aneurysms occur in approximately 10–30% of patients with ADPKD and subarachnoid haemorrhage may cause sudden death in young adults. Summary box 82.2 Renal cystic disease /uni25CF - /uni25CF /uni25CF /uni25CF /uni25CF

Double ‘J’ stent Lateral spatulation of the ureteral end Bosniak renal cyst classi /f_i cation is used to grade cysts and probability of malignancy ADPKD – autosomal dominant, systemic disease: Rarely manifests before the fourth decade Hypertension, abdominal pain, haematuria or a palpable /f_l ank mass are common presentations Control of hypertension can delay progression

Benign renal tumours

Benign renal tumours

Incidental detection of renal lesions has increased owing to - the widespread use of abdominal imaging. The lesions may be cystic or solid. Solid renal tumours should be considered - malignant unless proven otherwise. Renal oncocytoma - This derives its name from its cellular appearance on histo - pathology , where uniformly highly granular eosinophilic cytoplasm owing to abundant mitochondria (oncocyte) is seen. It accounts for around 5% of renal tumours. It appears as an enhancing mass on cross-sectional imaging and is di ffi cult to di ff erentiate from RCC. Both RCC and oncocytoma pr esent at around the seventh decade and have a male preponderance. It coexists with RCC in approximately 10% of cases. The characteristic radiological features on axial imaging are the presence of central stellate scarring and a spoke wheel appearance in the angiographic phase. Nephron-sparing sur - - gery , such as partial nephrectomy , should be the preferred option, whenever feasible. The diagnosis is usually confirmed after removal. Histologically , it can be confused with chromo - phobe RCC, particularly the eosinophilic v ariant. They may be di ff erentiated by the use of immunohistochemistry staining, where chromophobe RCC stains positive for cytokeratin-7. Renal angiomyolipoma - Angiomyolipoma comprises a composite mix of fat tissue - with dysmorphic blood vessels and smooth muscle. It is most often detected incidentally and has a female preponderance. Angiomyolipoma may be associated with syndromes such as the tuberous sclerosis complex or it may be sporadic in nature. Spontaneous acute haemorrhage into the mass can present with loin pain. Pregnancy is a potential risk factor for bleeding. US shows a bright echogenic mass lesion on account of the high fat content. CT scan shows an intralesional fat density of –15 to –20 Hounsfield units (HU) within the mass, which sis. Management depends upon the size of the tumour, the risk of haemorrhage and the symptoms. Tumours <4 /uni00A0 cm can be followed up. Nephron-sparing sur gery such as partial nephrectomy is the preferred option. Angioembolisation is the preferred modality of choice in the setting of acute haemor rhage. Drugs that inhibit this pathway (mTOR pathway), such as everolimus and sirolimus, have recently been shown to have excellent response rates in this subg roup of patients with the tuberous sclerosis complex who have activation of the tumor igenic mTOR pathway . Juxtaglomerular cell tumour These are extremely rare tumours that occur at a young age, often presenting with hypertension and hypokalaemia with high renin levels. These tumours are unique in that hyperten sion resolves with surgery .

CONGENITAL DISEASES

Renal agenesis

CONGENITAL DISEASES Renal agenesis

Complete absence of one kidney occurs in 1 in 3000 live births. The other formed kidney is usually hypertrophic. Reproductive tract anomalies are common in females with unilateral renal agenesis. Bilateral renal agenesis is incompatible with life.

Children

Children

See also Chapter 20 . Stones are rare in children. Childhood urolithiasis is more common in males in the first decade and in young adolescent females. Calcium oxalate stones are the most common vari - ety . Genetic disorders are seen in 17% of children with stones. The y may be asymptomatic or may present with non-specific symptoms such as crying, irritability and vomiting. Diagnosis and treatment should be planned such that ionising radiation is ke pt to a minimum. Indications for various modes of treatment are similar to those for adults. - Summary box 82.4 - Urolithiasis /uni25CF /uni25CF /uni25CF - /uni25CF

Causes of stone formation are multifactorial, including age, gender, ethnic origin, family history, environmental factors, geography and diet Ureteric colic is the most common acute presentation. NCCT is the investigation of choice Decreased animal protein intake, decreased salt intake and adequate /f_l uid intake are necessary to prevent recurrence Complete removal and long-term antibiotics are important to prevent recurrence of infection stones

Classification

Classification

- UTI is classified as uncomplicated when it occurs in an immunocompetent host with an anatomically normal and - functional urinary tract. UTIs may also be classified on their site of origin as pyelonephritis (kidney), cystitis (bladder), urethritis or prostatitis. While acute pyelonephritis indicates an acute infection of the kidney , chronic pyelonephritis is only a morphological description of previous infection-related or nuclear imaging. Acute pyelonephritis This commonly occurs as a result of ascending infection from organisms in the lower tract, usually caused by Gram-negative bacteria. Haematogenous spread may be seen in patients with diabetes and in immunocompromised hosts, people who inject drugs and patients with bacterial endocarditis. It is more common in females, especially during childhood, at puberty , after intercourse and during pregnancy . Acute pyelonephritis usually presents with fever, chills, flank pain, nausea and vomit ing. Loin tenderness may be present. Symptoms may vary from mild to severe illness with septic shock and renal failure. Pyuria is almost always present and its absence in a patient with pyelonephritis may point towar ds an obstructed urinary tract. Urine and blood should be collected for culture. Escherichia coli and other Gram-negative organisms are commonly responsi ble. Imaging is necessary when the patient is not responding to antibiotics to rule out pyonephrosis, renal abscess and obstruction. Renal US is often the first imaging modality used. Contrast-enhanced CT (CECT) typically sho ws decreased patchy opacification of the a ff ected parenchyma. Pyelonephritis complicating pregnancy The relaxing e ff ect of progesterone during pregnancy causes ureteral smooth muscle relaxation and dilatation, presumably predisposing pregnant women to ascending upper tract infec tions. It is associated with fetal growth retardation and preterm delivery . Therefore, all pregnant women must be screened in the first trimester for ABU because, untreated, a third of these patients will develop UTI. Lower tract UTI typically occurs in the first trimester whereas pyelonephritis most often presents in the second or third trimester with acute abdominal pain or premature labour. Pyelonephritis is more common in pregnant women with an underlying urological abnormality or diabetes. A renal US is indicated if response to treatment is poor. Anti biotic use during pregnancy is tailored to avoid fetal harm and typically includes fosfomycin, penicillins or cephalosporins. Renal and perirenal abscess A renal abscess results from an ascending UTI in associa tion with an underlying urinary tract abnormality such as obstructive uropathy or VUR. It is usually caused by common uropathogens such as E. coli and other Gram-negative bacilli. Renal abscesses may extend and perforate the renal capsule to form a perirenal abscess. Multiple renal abscesses may conglomerate into a solitary suppurative lesion called a renal carbuncle. This is usually caused by Staphylococcus aureus reaches the kidney by haematogenous spread. The clinical presentation may be insidious and non-specific but patients usually present with persistent fever, back pain, abdominal pain and costovertebral tenderness. Urine exam ination may be normal if the abscess does not

communicate with the collecting system. CECT scan is the investigation of choice to establish the diagnosis. Treatment with antibiotics without drainage may be e ff ec tive in carefully selected patients when the abscess is small (<3 /uni00A0 cm) or in a stab le patient (up to 5 /uni00A0 cm). Empiric antibiotic and other uropathogens causing complicated UTI. Culture- directed antibiotics may be needed for 2 weeks or longer depending on response. Percutaneous aspiration or drainage of pus is indicated in abscesses >5 /uni00A0 cm and in patients not responding to antibiotics. Open surgical drainage is indicated when percutaneous drain - age is inadequate. Emphysematous pyelonephritis This is an acute-onset, rapidly progressive, possibly lethal form - of pyelonephritis characterised by parenchymal necrosis and gas formation, caused by organisms including E. coli , Klebsiella pneumoniae , Pseudomonas aeruginosa and Proteus mirabilis . Most patients have diabetes (up to 90%) and they may have obstruc - tion secondary to calculi or papillary necrosis. Increased glucose levels in those with diabetes may provide a substrate - for carbon dioxide production from fermentation. Symptoms are suggestive of p yelonephritis and an abdominal mass may be palpable. CECT of the abdomen is diagnostic and shows gas in the renal parenchyma, collecting system or both, along with other features of infection such as abscess, obstruction and perinephric stranding. Early diagnosis, intravenous broad-spectrum antibiotics and percutaneous drainage of the abscess and obstructed kidneys have improved outcomes in these patients. Emergency nephrectomy is rarely required and is reserved for patients who do not respond to the described - measures. Xanthogranulomatous pyelonephritis Xanthogranulomatous pyelonephritis (XGP) occurs with severe renal infection in an obstructed kidney and is usually associated with calculi, causing loss of function and paren - chymal destruction. Pathological examination typically shows accumulation of lipid-laden foamy macrophages. Patients may present with flank pain, fever with chills, persistent bacteriuria - and a flank mass. A history of stone disease may be present. It is usually unilateral. CECT of the abdomen is diagnostic and shows a non-functioning enlarged hydronephrotic kidney around a shrunken pelvis with a calculus, also known as the bear’s paw sign (Figure 82.7). Nephrectomy is the defin - - itive treatment. , which - -

Figure 82.7 Xanthogranulomatous pyelonephritis with the ‘bear’s paw sign’.

the pelviureteric junction to be hiked up Putty kidney: extensive dystrophic calci /f_i cation involving all or most of the kidney, seen as calci /f_i ed non-functioning renal tissue Pipe-stem ureter: straightening of the ureter as a result of /f_i brosis of the wall of the ureter Corkscrew ureter: multiple annular strictures along the length of the ureter Golf hole ureteric ori /f_i ce: ureteric ori /f_i ces may become patulous and may be pulled up; tubercles are infrequent in the bladder Figure 82.8 Schematic illustration showing the sequelae of urinary tuberculosis (courtesy of Nivedita Kekre and Dr Madhuri Sadanala).

Clinical presentation

Clinical presentation

Incidentally detected asymptomatic stones are increasingly diagnosed because of the widespread use of imaging. The presenting symptoms depend on the location of the stone, the size and type of stone, underlying infections and complications related to stone disease. Haematuria may be gross or micro scopic, especially during episodes of renal colic. Calculuria is described as sand or gravel accompanying urine. Ureteric colic is acute abdominal pain caused by hyperperistalsis of the ureteric musculature against the obstructing stone. It manifests as sudden-onset ex cruciating pain in the flank that can radiate to the groin, scrotum or labia. Lower ureteric stones close to or lodged at the UVJ can cause symptoms of urgency and frequency . Malaise and weight loss can occur in longstanding infection stones or as a manifestation of renal failure. High- grade fever with chills suggests an underlying UTI and should be considered an emergency . During history taking, informa tion about risk factors such as diet, physical activity , fluid intake, history of urinary tract infections, gastrointestinal symptoms, previous surgical history , family history and previous treatment f or stone disease should be enquired about.

Complications

Complications

Renal and ureteric stones can lead to significant morbidity owing to urinary tract obstruction, infectious complications and loss of renal function. Bilateral obstructing ureteric stones or ureteric calculi in a solitary kidney can present with anuria (calculous anuria). Infectious complications include pyelo nephritis, pyonephrosis, renal abscess or septicaemia. Uncom mon but serious complications include XGP and pyeloenteric or cutaneous fistulae in neglected cases. Nephron loss can occur as a result of recurrent episodes of infection and obstruc tion, causing chronic renal failure.

Congenital megaureter

Congenital megaureter

The normal ureteric diameter in children up to 16 years is 0.50–0.65 /uni00A0 mm. If the ureter is dilated by more than 7 /uni00A0 mm, it - is classed as a dilated or megaureter. This may occur with or without obstruction or reflux. Most cases of megaureter with obstruction present in childhood with severe infections. Renal stones can f orm easily in the dilated systems. Surgical correc - tion is indicated in symptomatic patients who have recurrent urinary tract infections (UTIs), progressive dilation on US and di ff erential renal function of less than 40%. Surgery involves excision of the stenosed distal ur eter and non-refluxing ureteric reimplantation.

Congenital pelviureteric junction obstruction

Congenital pelviureteric junction obstruction

Congenital PUJO is the most common cause of unilateral hydronephrosis with an incidence of 1 in 500 live births. It may result from intrinsic obstruction secondary to an aperistaltic segment at the PUJ due to muscular hypoplasia. Other causes include a high insertion of the ureter into the pelvis and the presence of crossing aberrant vessels at the PUJ. It is more

Ureter draining lower moiety inserts laterally and superiorly into the bladder Ureter draining the upper moiety inserts medially and inferiorly into the bladder – ureterocele and upstream hydroureteronephrosis Sphincter Figure 82.4 Retrocaval ureter with a classic ‘reverse J’ sign seen on intravenous urogram (courtesy of Department of Urology, Christian Medical College, Vellore, India).

common in males and on the left side. Bilateral obstruction occurs in 10% of cases. Historically , PUJO presented as a palpable flank mass in an infant or a child, but most are now detected before birth with antenatal US. Older children may present with intermittent flank pain, UTI or a flank mass. Adults present with back or flank pain or recurrent pyelonephritis. Rarely , a patient may present with a history of severe flank pain following ingestion of large amounts of fluid, which is relieved after passing a large amount of urine (Dietl’s crisis). US may show symmetrical hydronephrosis and a dilated renal pelvis and can provide information on the severity of obstruction by measuring the degree of dilatation, parenchy mal thickness and cortical echogenicity . Isotope diuretic renog raphy is the current investigation of choice. Isotope uptake and washout of the isotope can be followed with time to produce a renogram curve. Usually , half of the peak isotope activity is cleared within 10–15 minutes (T). A rising curve following 1/2max administration of furosemide, a T of greater than 20 min 1/2max utes and a di ff erential function of less than 40% on the a ff ected side is suggestive of significant obstruction and is an indication for surgical intervention (Figure 82.5). CTU or MRU may also be used in the evaluation of PUJO. T he Anderson–Hynes dismembered pyeloplasty is the pro cedure of choice with a wide funnelled, dependent anastomo sis, maintaining good vascularity of the upper ureter and pelvis and excision of the redundant pelvis (Figure 82.6). The indica tions for pyeloplasty are persistent pain, hypertension, haema turia, secondary renal calculi and recurrent UTIs. Endoscopic management in the form of endopyelotomy is reserved for post-py eloplasty strictures. Dietl’s crisis, first reported by Josef Dietl , 1804–1878, in 1864, an Austrian doctor and pathologist known for his work on floating kidneys. James Christie Anderson , 1899–1984, urologist, Royal Hallamshire Hospital, She ffi eld, UK. Wilfred Hynes , 1903–1991, plastic surgeon, The Plastic and Jaw Department, The Royal Hospital, She ffi eld, UK. Anderson and Hynes devised the operation in 1949. Carl Weigert , 1845–1904, German pathologist and anatomist known for work on cellular staining. Robert Meyer ,

1864–1947, German pathologist and gynaecologist in Berlin, removed from his position for being Jewish, emigrated in 1939 to Minneapolis, MN, /uni00A0 USA. Morton A Bosniak , 1929–2016, Professor of Radiology , New Y ork University (NYU) Langone School of Medicine, New Y ork, NY , USA. Congenital anomalies /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF

Figure 82.5 Isotope renal scan using diethylenetriaminepenta-acetate showing an obstructive pattern on the time–activity graph with hold-up of contrast of up to 2 hours. This /f_i nding is consistent with pelviureteric junction obstruction (courtesy of Department of Urology, Christian Medical College, Vellore, India). Congenital anomalies are usually detected incidentally and often only manifest when effected by pathology such as stone disease or malignancy Ectopic ureter should be suspected in a female child who presents with continuous incontinence of urine and also voids normally Weigert–Meyer rule The ureter that drains the upper moiety is at a more inferior and medial position and is prone to obstruction and dysplasia The ureter that drains the lower moiety is at a more superior and lateral position and is prone to VUR Most cases of ANH are transient and resolve after birth

Diagnosis

Diagnosis

The diagnostic approach can be classified into investigations done in the emergency setting and those done in the non emergency setting. The most common acute presentation of stone disease is - ‘ureteric colic’. Small 3- to 5-mm calculi are usually responsible for ureteric colic and commonly lodge at the UVJ. Non- - steroidal anti-inflammatory drugs and paracetamol are - e ff ective. Antispasmodic medications are not necessary to alleviate pain. Abdominal examination may reveal renal angle tenderness. Pelvic examination is especially important in women to exclude tubo-ovarian pathology such as an ectopic pregnancy or twisted ovarian cyst. Table 82.1 lists the - di ff erential diagnoses. /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF - /uni25CF Investigations include urinary examination, blood exam - ination and diagnostic imaging. The majority have micro - scopic haematuria and pyuria. Pyuria may be sterile pyuria or due to infection. An elevated leukocyte count suggests infection and may be an indication for starting antibiotics. Pregnancy should be ruled out. A radiograph of the kidneys, ureters and bladder and US - are good fir st-line tests. Non-contrast CT (NCCT) is the inves - tigation of choice for the diagnosis of stones. It allows for diag - nosis of both radio-opaque and radiolucent stones with the exception of indinavir stones. Most patients respond to medi - cation to alleviate pain. However, if the pain does not reduce with analgesics, or if the patient shows features of sepsis or urinary obstruction, emergency urinary decompression should be planned. Blood and urine should be cultured in patients suspected of sepsis , and empirical broad-spectrum antibiotics should be initiated. If the patient is clinically unstable, initial stabilisation in critical care may be warranted. Emergency uri - nary decompression may be done either with ureteric stenting - or with PCN. However, in the absence of infection, in a certain - select group of symptomatic but surgically fit patients, remov al of stones may be possible by ureteroscopy . Metabolic evaluation - The extent of metabolic evaluation depends on the risk asso - ciated with the recurrence of stone formation. Urinary examination is done to look at crystals and pH in the non-emergency setting. Urine culture is performed if definitive management is planned. Blood chemistry for serum - le vels of calcium, phosphorus and uric acid are done to rule out hypercalcaemia, hypophosphataemia and hyperuricaemia.

TABLE 82.1 Differential diagnoses for ureteric colic. Urinary tract Clot colic Anticoagulation therapy, haemophilia, vascular tumours Papillary necrosis Diabetes, NSAIDs, sickle cell disease Other organs Acute appendicitis Ectopic pregnancy Ovarian torsion Acute intestinal obstruction Abdominal aortic aneurysm Malingering NSAID, non-steroidal anti-in /f_l ammatory drug.

risk patients.

Ectopic kidney

Ectopic kidney

This occurs when the mature kidney fails to reach its normal location in the lumbar region. The incidence is 1 in 500–1200. An ectopic kidney (Figure 82.1) may be found anywhere along the path of ascent: pelvic, iliac, abdominal and rarely thoracic. When the ectopic kidney is located on the contralateral side to its ureteric insertion, it is called crossed ectopia. Renal ectopia may be associated with reflux in the ectopic or orthotopic kidney and with pelviureteric junction (PUJ) and ureterovesical junction (UVJ) obstruction.

Ectopic ureters

Ectopic ureters

An ectopic ureter is one that drains to regions other than the bladder. Ectopic ureters are almost always associated with ureteric duplication and are bilateral in 10%. The female - to-male ratio is 7:1. In females, the ectopic ureter opens either into the urethra below the sphincter or into the vagina (Figure 82.3). Such a child would complain of incontinence of urine despite normal voiding. In contrast, the male child is always continent as the ur eter opens above the external urethral sphincter. Computed tomo-urography (CTU) or magnetic resonance urography (MRU) is diagnostic.

Endourology

Endourology

Endourological procedures are the current preferred mode of - treatment owing to their minimal invasive nature, technological - advancements in instrumentation and more e ffi cient energy sources for stone fragmentation. Current energy sources are pneumatic, US or laser lithotripsy . The type of energy source depends on the type of surgery and stone characteristics. Laser energy can be delivered via flexible instruments. Ureterorenoscopy Ureterorenoscopes (URSs) are long thin scopes that are used to remove ureteric and renal stones. They have working channels that allow for the introduction of energy sources, graspers and baskets. Current models are either semirigid or flexible scopes. A semirigid URS is usually used with a pneumatic lithotripter or laser energy device. Complications include ureteric perfo - ration, avulsion and retropulsion. Ureteric avulsion can be avoided by careful use of baskets under vision. URSs can also be used in patients with bleeding disorders, with a moderate increase in complications. A slimmer and more flexible URS with active deflection of the tip and laser technology with thinner fibres allows for retrograde access to the kidney via the ureteric orifice. This procedure avoids the morbidity associated with percutaneous nephrolithotomy (PCNL). Laser is used as an energy source for stone fragmentation. Indications for retrograde intrarenal surgery (RIRS) /uni25CF Renal stones <2 /uni00A0 cm. /uni25CF Lower pole calculi. /uni25CF Obesity . /uni25CF Musculoskeletal deformities (e.g. kyphoscoliosis) and renal anomalies (HSK or pelvic kidney). /uni25CF Bleeding diathesis. Percutaneous nephrolithotomy PCNL involves removal of renal stones by creating a track between the skin and the pelvicalyceal system. Typically , this procedure is done in the prone position. Fluoroscopy or US is used for localisation. The posterolateral calyx is commonly chosen for entry . US in conjunction with pneumatic and laser lithotripsy is the most common energy source used. Complica tions include bleeding, infection and pleural violation in cases of supracostal puncture. Severe bleeding may require selective angioembolisation. Indications for percutaneous nephrolithotomy /uni25CF Renal stones >2 /uni00A0 cm. /uni25CF Lower pole renal stones with anatomy that is unfavourable for SWL. /uni25CF Failed SWL or RIRS for renal calculi. /uni25CF Staghorn calculi. Contraindications to percutaneous nephrolithotomy /uni25CF Pregnancy . /uni25CF Untreated UTI. /uni25CF Bleeding diathesis. /uni25CF Current anticoagulation. Miniaturised percutaneous nephrolithotomy Miniaturised PCNL (e.g. mini-perc) involves the use of smaller access tracks. The standard PCNL access track is >28Fr compared with miniaturised versions using <22Fr tracks. Miniaturised PCNL is most useful in patients with a smaller stone burden and in children. Lateral and supine PCNL are associated with fewer anaes thetic complications. Moreover, concomitant flexible ureteros copy for endoscopic combined intrarenal surgery can be done to address complex renal stones, multiple stones or stones in challenging locations.

Epidemiology

Epidemiology

The lifetime prevalence varies from 1% to 20% and the causes are multifactorial. Recurrence of stone disease is high, with 50% having recurrence within the first decade of diagnosis. Non-modifiable factors associated with stone formation /uni25CF Age . The adult peak incidence in men is the fourth to sixth decade; women have a bimodal peak in incidence in the third decade and the postmenopausal period. /uni25CF Gender . Men are twice as likely to form stones. /uni25CF Ethnic origin . White people have a higher risk of stone disease than other ethnic groups. Recent evidence suggests that environmental and dietary factors may be more im portant than ethnic origin. /uni25CF Family history . Patients with a family history of stone disease are 2.5 times more likely to develop stone disease themselves. Examples of hereditary forms of stone disease include cystinuria, type I renal tubular acidosis (RTA) and primary hyperoxaluria. Modifiable factors associated with stone formation /uni25CF Environmental factors . People living in hot and arid regions such as the desert or tropical areas have a higher incidence of stone disease owing to increased perspiratory fluid loss. /uni25CF Drugs . Drugs can predispose to stone formation through metabolic e ff ects (e.g. corticosteroids, chemotherapeutic agents).

Genetic renal cysts

Genetic renal cysts

- These cystic renal lesions have a known genetic inheritance. They are usually accompanied by involvement of other organ systems and present earlier in life than sporadic renal cysts.

1.

Dependent 2. Wide 3. Funnel shaped 4. Maintain good vascularity 5. Without tension 6. Excise redundant pelvis Dilated renal pelvis Diamond-shaped incision on the pelvis Excised diseased segment PUJ stenosis Figure 82.6 Steps of open dismembered pyeloplasty (courtesy of Nivedita Kekre and Dr Madhuri Sadanala). PUJ, pelviureteric junction.

Horseshoe kidney

Horseshoe kidney

This is the most common renal fusion anomaly , occurring in about1 in 400 live births with a male predominance. The isth mus lies at the level of the fourth to fifth lumbar vertebrae (fused lower poles). This causes failure to ascend and rotate so that the renal pelvis faces anteriorly and vertically with the malrotated calyces pointing posteromedially (Figure 82.2). T he vascular supply is variable and the ureter may insert high on the renal pelvis. Most horseshoe kidneys (HSKs) are asymptomatic but they are associated with an increased incidence of genital anomalies, PUJ obstruction (PUJO) and stone formation. The incidence of Wilms' tumour is higher in HSK. Carl Max Wilhelm Wilms , 1867–1918, German surgeon, described Wilms' tumour in 1899.

The pathophysiology and management of renal and • ureteric stone disease Trauma to the kidney and ureter • Presentation and management of renal neoplasms •

INFECTIONS

INFECTIONS

UTI is very common and a ff ects all ages and both sexes. It can cause significant morbidity and is a rare cause of mortality in patients with serious comorbidities or in patients with urinary tract obstruction. Recurrent UTI is more common in women, a ff ecting 30–40% in the sexually active age group. It can be defined as an inflammatory response of the urothelium (host) - to invading bacteria. Asymptomatic colonisation or bacteriuria (ABU) is also common and can be di ff erentiated from a UTI by the absence of symptoms and pyuria (leukocytes in urine).

Introduction

Introduction

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Learning objectives

Learning objectives

To know: Congenital anomalies of the kidney and ureter • Classi /f_i cation of renal cysts • Classi /f_i cation, de /f_i nitions, pathogenesis and management • of urinary tract infections

Multicystic dysplastic kidney

Multicystic dysplastic kidney

Multicystic dysplastic kidney (MCDK) is the second most common cause of an abdominal mass in newborns after hydronephrosis due to PUJO. The unilateral incidence is 1 in 1000–4000 live births. It has a ‘bunch of grapes’ appearance -

Figure 82.1 Computed tomography scan showing a pelvic kidney with calculus.

with multiple non-communicating cysts of varying sizes with out identifiable renal parenchyma. MCDKs can be diagnosed on antenatal ultrasound (US), with multiple cysts being evident as early as 15 weeks’ gestation. Isotope renal scan will show a photopenic area in the renal fossa with surrounding back ground activity . Newbor ns may present with a palpable renal mass but nephrectomy is not necessary as the majority undergo involution within 5 year s. Bilateral MCKD is incompatible with life.

(b) Figure 82.2 (a) Horseshoe kidney (courtesy of Nivedita Kekre and Dr Madhuri Sadanala). (b) Intravenous urogram image at 5 minutes, showing horseshoe kidney and posterior orientation of the calyces (courtesy of Department of Urology, Christian Medical College, Vellore, India).

Non-endourological surgical management

Non-endourological surgical management

Open surgery such as pyelolithotomy and anatrophic nephro lithotomy is reserved for complex and infected stones with anatomical abnormalities. High-risk stone formers should be advised to follow preventive measures to reduce recurrence. General measures advised to all patients include: /uni25CF fluid intake of more than 2.5 litres per day; /uni25CF dietary calcium should not be restricted; supplemental cal - cium, if necessary , should be taken at meal times; /uni25CF reduce intake of animal protein and salt.

Non-surgical management of stone disease

Non-surgical management of stone disease

This involves watchful waiting, medical expulsive therapy , SWL and stone dissolution therapy . Watchful waiting Patients with small (<5 /uni00A0 mm), non-obstructive, asymptomatic, lower pole renal calculi with preserved renal function may be kept on follow-up. Up to 90% of 4-mm stones and 50% of 6- to 10-mm stones pass spontaneously . Medical expulsive therapy Tamsulosin is an α -adrenergic adrenoreceptor blocker 1 (α -blocker) that causes smooth muscle relaxation of the distal ureteric muscle. It can be used for distal ureteric stones larger than 5 /uni00A0 mm and to assist passage of fragments following SWL. Extracorporeal shockwave lithotripsy SWL is a non-invasive method introduced in 1980 by Christian Chaussy that allowed stones to be treated on an outpatient basis. Mechanism of action The stone is localised using either fluoroscopy or US or both. Then acoustic pulse waves are generated and focused on the stone. Stone fragmentation occurs as a result of mechanical stress caused directly by the energy transmitted by the incident shockwave and indirectly by the collapse of bubbles. The e ffi cacy of SWL reduces with an increasing number of stones and volume of stone burden. Steinstrasse is a German word meaning ‘street of stones’. It describes a row of closely gathered stone fragments that line the distal end of the ureter (Figure 82.10). This occurs when the stone burden is high or when the stones are hard. These stones are usually asymptomatic and pass spontaneously; how ever, they may cause obstruction, requiring surgical interven tion. ‘Clinically insignificant residual fragments’ are residual stone fragments of 4 /uni00A0 mm in size or less after treatment that are expected to pass spontaneously . However, 20–40% of these fragments may not clear and for m a nidus for stone regrowth.

Pathogenesis

Pathogenesis

Stone formation results from a cascade of events that occur during and after urine formation. When the concentration of culprit salts such as calcium and oxalate overwhelm inhibitory factors (e.g. citrate, potassium, magnesium, Tamm–Horsfall mucoproteins, pH changes), they Sushruta , 600 /uni00A0 /b.sc/c.sc/e.sc , authored Sus ´ruta-sam hita ¯, considered the father of plastic surgery . . Igor Tamm , 1922–1995, an outstanding cytologist, virologist and biochemist, pioneer in the study of viral replication, professor at the Rockefeller Institute for Medical Research, New Y ork, NY , USA. Frank Lappin Horsfall Jr , 1906–1971, American microbiologist specialising in pathology , worked at the Rockefeller Institute, New Y ork, NY , USA. The Tamm– Horsfall protein was first purified in 1952 during his work with Igor Tamm. Alexander Randall , 1883–1951, American urologist, first described the plaques in 1937 as part of a postmortem case series using a hand lens. o ff with the flow of urine or they may anchor onto sites like renal papillae to form Randall’s plaques. Variations in the pH of urine may also facilitate or inhibit stone gr owth; acidic pH precipitates the formation of uric acid stones and alkaline pH precipitates the formation of calcium phosphate stones. Hence, the manipulation of pH through medication can help in preventing new stone formation. Stasis of urine also promotes stone formation. Stasis stones - are usually multiple, round and have a smooth surface. These are called ‘milk of calcium stones’.

Pregnancy

Pregnancy

Renal colic is the leading cause of non-obstetric hospital admission in pregnancy . The physiological changes that take place during preg - nancy include an increase in glomerular filtration rate by 50%; increased excretion of calcium, uric acid and sodium; and increased excretion of inhibitors of crystallisation such as citrate and magnesium. Urine pH is alkaline and so the pre - dominant stone type seen in pregnancy is calcium phosphate stones. US is the primary mode of investigation for renal colic. MRI can be used as a second-line investigation to define the - level of obstruction. Most stones pass spontaneously . Ho wever, stones can cause loss of pregnancy and prematur e labour. Hence, emergency ureteroscopy is a reasonable first-line option in well-selected distal ureteric stones. Inter nal stenting or PCN can be used in the interim and a definitive procedure can be planned fol - lowing childbirth. Pregnancy is an absolute contraindication to SWL.

RENAL CYSTS

RENAL CYSTS

Renal cysts can be broadly classified into sporadic, acquired and genetic causes.

Renal cell carcinoma

Renal cell carcinoma

RCC is the most common solid neoplasm of the kidney . It accounts for around 90% of renal tumours and constitutes 2–5% of all cancers in adult men and 1–3% in adult women. There has been a recent steady increase in the incidence of RCC. It may be sporadic or familial. Familial renal cell carcinoma von Hippel–Lindau (VHL) syndrome is the most common familial syndrome associated with RCC. VHL disease is a rare autosomal dominant disorder that is characterised by multiple pathologies, including clear-cell RCC (ccRCC), phaeochromocytoma, retinal angiomas and haemangioblastomas of the brainstem, cerebellum or spinal cord. Aetiology Cigarette smoking, obesity and hypertension are the major risk factors associated with RCC. Others include diuretics, occupational exposure to petrochemicals and dyes and ARCD in patients on long-term haemodialysis. Clinical presentation The classic triad of flank pain, haematuria and a palpable mass is now uncommon as most renal masses are detected incidentally . Symptoms and signs may be non-specific. The most common presenting symptom is haematuria. Patients may have constitutional symptoms such as fever, malaise and weight loss in advanced disease. Advanced disease can present with bilateral lower limb oedema or recent-onset non-reducing right-sided varicocele owing to thrombus in the IVC. Paraneoplastic syndromes (PNSs) are found in up to one-third of patients with RCC. The most common PNS is an elevated erythrocyte sedimentation ra te (ESR) followed by hypertension, anaemia and hypercalcaemia. Up to a quarter of the patients may have evidence of metastatic disease on pre sentation. The most common site of metastasis is the lung and Eugen von Hippel , 1867–1939, Professor of Ophthalmology , Göttingen, Germany , first described angiomas in the eye in 1904. Arvid Vilhelm Lindau , 1892–1958, Swedish pathologist, described angiomas of the cerebellum and spine in 1927. may occasionally present as pathological fractures. Pathology ccRCC is histologically an adenocarcinoma arising from the - proximal renal tubular epithelium. They are slow growing and bulge out of the renal contour (Figure 82.15). Most are soli - tary , but bilateral and multiple tumours are found in familial RCC. The prognosis of ccRCC varies depending on various - histopathological features, suc h as nuclear grading. Other histological variants are papillary RCC, chromophobe RCC and, rarely , collecting duct carcinoma and renal medullary carcinoma. Table 82.2 summarises the salient features of subtypes of RCC. The tumour can spread directly , invading the perinephric - tissue through the capsule or at times directly extending into the renal vein as a tumour thrombus. V ein wall invasion is asso - ciated with poor prognosis. Diagnosis Laboratory findings Evaluation should include blood count, ESR, serum creatinine, liver function tests, lactate dehydrogenase (LDH), corrected serum calcium, coagulation markers and urine analysis. Increased alkaline phosphatase should prompt further investi - gation to rule out liver and skeletal metastases. LDH is useful in risk stratification of metastatic disease. -

Figure 82.15 Cut surface of a kidney showing a large, well-demarcated clear-cell renal cell carcinoma in the upper pole with foci of yellowish areas signifying the lipid content of the tumour (courtesy of Dr Vikram Raj Gopinathan, Department of Pathology; photo credit: Sekhar, Christian

Medical College, Vellore, India).

/uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Radiological investigations Although US can diagnose the tumour, triphasic CECT is the investigation of choice for diagnosis and staging. RCC typically shows contrast enhancement after contrast injection (a change of >15 /uni00A0 HU is considered significant). The CT also provides additional information on the function of the opposite kidney , primary tumour extension, venous involvement, enlargement Dimitrie D Gerota , 1867–1939, Romanian anatomist, physician and radiologist. and intra-abdominal metastatic disease (Figure 82.16). MRI provides similar information to CT , but can be superior at detecting tumour infiltration into the vein wall and the level of thrombus. A chest radiograph should be obtained in all cases. A bone scan is necessary in a patient with elevated alkaline phospha - tase, bone pain or hypercalcaemia. Tumour staging The treatment and prognosis of RCC depends on its patho - logical staging. The most important factors are the size of the tumour and whether it is confined within the renal capsule and Gerota’s fascia. Involvement of the lymph nodes, renal sinus and vein wall are associated with a poorer prognosis than the presence of tumour thrombus in the renal vein or IVC. Currently the most commonly used system for staging RCC is the TNM classification (Figure 82.17). Prognostic factors Currently , the grading system proposed by the International Society of Urological Pathology (WHO/ISUP) is used for grading renal cancer. Anatomical factors such as tumour size, venous invasion, renal capsular invasion and adrenal involve - ment herald a poorer prognosis. Certain histological types, e.g. sarcomatoid, have a worse prognosis. Management Nephron-sparing surgery Radical nephrectomy remains the gold standard treatment for localised disease. However, with the recent increase in incidental detection of small renal masses (tumours <4 /uni00A0 cm), more nephron-sparing surgery is being performed. Partial nephrectomy should be the treatment of choice in tumours less than 4 /uni00A0 cm, in well-selected tumours between 4 and 7 /uni00A0 cm, in bilateral tumours, in tumours in solitary kidneys and in patients with pre-existing renal dysfunction. Minimally invasive tech - niques by laparoscopy or robots have reduced postoperative morbidity . Because of the limitation of ischaemia time, mini - mally invasive techniques should be reserved for tumours with non-complex anatomy , as predicted by nephrometry scores. Alternative techniques such as surveillance, cryoablation or radiofrequency ablation of small renal tumours may be o ff ered in patients with high surgical risk (e.g. elderly patients, patients with multiple comorbidities). Activ e surveillance is based on the fact that most incidental tumours detected in the elderly grow slowly and have a low chance of local invasion or metas - tasis. Radical nephrectomy Classically , radical nephrectomy involved removal of the entire kidney enclosed in Gerota’s fascia with the ipsilateral adrenal gland and regional lymphadenectomy . Most are now performed laparoscopically and the adrenal is spared if there is no involvement on CT/MRI. Lymphadenectomy is indicated only in high-risk patients with large primary tumours and enlarged lymph nodes.

subtypes of renal cell carcinoma (RCC). RCC subtype Salient features Clear-cell RCC Most common subtype Usually sporadic May be associated with loss of chromosome 3p and a mutated von Hippel–Lindau gene Papillary (type I and Second most common II) RCC Usually sporadic but may be familial Type 1 tumour has a better prognosis than type 2 Chromophobe RCC Usually sporadic but may be familial Good prognosis Collecting duct Uncommon (1–2%) carcinoma Aggressive tumour Arises from the renal medulla, hence centrally located tumour Renal medullary Rare (<0.5%) carcinoma Very aggressive Associated with a younger age and sickle cell trait Centrally located

tumour Figure 82.16 Contrast-enhanced computed tomogram showing a left renal mass with left renal vein thrombus extending into the inferior vena cava (IVC) (hypoattenuated linear area within the IVC) (courtesy of Department of Urology, Christian Medical College, Vellore, India).

Surgical management of inferior vena cava thrombus Renal tumours are associated with IVC tumour thrombus in 5–10% of cases. Tumour thrombus may extend as far as the right atrium. Thrombus extending to the retrohepatic or suprahepatic segment of the IVC requires full mobilisation of the liver, and thrombus extending to the right atrium may require cardiopulmonary bypass and circulatory arrest. Management of metastatic renal cell carcinoma Up to one-third of patients with RCC will present with disseminated disease. The International Metastatic Renal Cell Carcinoma Database Consortium risk stratification classifies a patient with metastases into risk groups based on performance status, time from diagnosis to systemic therapy , haemoglobin levels, calcium levels and platelet and neutrophil counts. The median survival of the good risk group is little more than 3.5 years, compared with just under 2 years in the intermediate-risk group. The expected median survival of the poor risk group is just over 7 months. Tyrosine kinase inhibitors inhibit vascular endothelial growth factor (e.g. sunitinib, pazopanib) and these drugs have improved survival in metastatic ccRCC. RCC is an immu nogenic tumour and responds to immunotherapy . The first generation of agents were interleukins and interferons. More recently , targeted therapy in the form of immune checkpoint inhibitor s and anti-programmed death 1/programmed death ligand-1 inhibitors have been used. Cytoreductive nephrectomy may be beneficial in good and intermediate-risk patients. Palliative nephrectomy may be consider ed for intractable haematuria, pain and symptomatic PNS. Angioembolisation of renal tumour can be performed in medically unfit patients with intractable haematuria.

vena cava Aorta Figure 82.17 Staging of renal cell carcinoma is based on size, position and lymph node involvement: Stage I: tumour <7 cm in the largest dimension, limited to the kidney. Stage II: tumour >7 cm in the largest dimension, limited to the kidney. Stage III: tumour in the major veins or adrenal gland with intact Gerota’s fascia, or regional lymph nodes involved. Stage IV: tumour beyond Gerota’s fascia. Adrenal Lymph gland node Stage I Stage III Stage II Gerota’s fascia Stage IV

Retrocaval ureter

Retrocaval ureter

- This is due to anomalous development of the inferior vena cava (IVC) with persistence of the posterior subcardinal vein. The right ureter passes behind the IVC rather than lying on its right side and may lead to ureteric obstruction, hydronephrosis and calculi. Most cases remain asymptomatic. Contrast - imaging with IVU, CTU, MRU and diuretic renogram aid the diagnosis. The classic sign of a dilated upper ureter at L3/L4 with proximal hydronephrosis and the ureter passing medially behind the IVC is described as the reverse ‘J’ sign (Figure 82.4). Surgical correction is indicated in symptomatic patients with ureteroureterostomy or pyeloplasty depending on the level of obstruction.

Lower moiety draining ureter – pelviureteric junction obstruction Lower moiety draining ureter – vesicoureteric re /f_l ux Upper moiety draining ureter – ectopic ureter drains below the sphincter in females and above the sphincter in males Figure 82.3 Complete duplication of the ureter and associated anomalies (courtesy of Nivedita Kekre and Dr Madhuri Sadanala).

Sporadic renal cysts

Sporadic renal cysts

Sporadic renal cysts are usually benign. Cysts with thin, sharply defined walls and clear fluid content are known as simple renal cysts. This category of cysts may be diagnosed with certainty by US. Apart from a few thin septa, any variation in the nature of the fluid, thickness of the cyst wall or septa or the presence of either calcification or a solid nodule would require further imaging with either computed tomography (CT) scan or - magnetic resonance imaging (MRI) to rule out cystic renal cell - carcinoma (RCC). Bosniak proposed a four-tiered classification of the malignant potential of cystic renal lesions. Category I cysts represent benign lesions that require no further follow-up, whereas categories III and IV have a higher probability of malignancy and require surgical excision. Category II cysts - can be safely followed up.

Surgical management

Surgical management

Indications for surgical intervention /uni25CF Failure of medical management. /uni25CF Impaired renal function. /uni25CF Chronic infection – staghorn calculi, matrix calculi. /uni25CF High-risk occupation or geographical location – pilots, long-distance locomotive drivers, sailors. /uni25CF Patient’s preference. The choice of therapy depends on multiple factors such as surgical fi tness, body habitus and stone characteristics. The Christian G Chaussy , b. 1945, German urologist. general principle is to choose the least invasive method possible for that particular stone.

Figure 82.10 Steinstrasse formation after extracorporeal shock wave lithotripsy at the right distal ureter (courtesy of Department of Urology, Christian Medical College, Vellore, India).

TUMOURS OF THE KIDNEYS AND URETERS Upper tract uro

TUMOURS OF THE KIDNEYS AND URETERS Upper tract urothelial cancer

Primary urothelial neoplasms of the renal pelvis and ureter are rare. They account for less than 10% of all urothelial tumours. They are more common in adult men. Important risk factors are tobacco consumption, occupations in the dye, petrochem - icals and rubber industries, analgesic abuse, high arsenic content in drinking water, exposure to cyclophosphamide and the presence of chronic infl ammation. Chronic infl ammator y conditions are also associated with squamous cell carcinoma.

ureteral mobilisation, taking spatulate freshened ends approximate with interrupted care to preserve periureteral on opposite sides; place /f_i ne absorbable sutures without adventitial tissue double ‘J’ stent Step 2: excise unhealthy devascularised ends Figure 82.13 Technique for upper ureteric injury repair. Steps 1 and 2: freshen the devascularised edges. Step 3: spatulate both ends and place an internal double J stent. Step 4: approximate both ends with interrupted absorbable sutures (courtesy of Nivedita Kekre and Dr Madhuri Sadanala). (a) U-shaped bladder /f_l ap (c) Double ‘J’ stent Suture the bladder incision with continuous or interrupted absorbable sutures Figure 82.14 Steps of Boari /f_l ap creation. (a) U-shaped bladder /f_l ap. internal DJ stent is placed in the ureter with a distal loop in the bladder. Nivedita Kekre and Dr Madhuri Sadanala). tension (b) Apex of the bladder /f_l ap is sutured to the end of the ureter; place double ‘J’ stent (d) (b, c) The apex of the bladder /f_l ap is sutured to the ureteric end and an (d) Suture the bladder incision with absorbable sutures (courtesy of

Patients commonly present with gross haematuria, with or without flank pain and occasionally clot colic. Passage of long, slender, worm-like clots is suggestive of upper tract involvement. Patients with known bladder tumours should always be screened for upper tract tumours. V ery few present with advanced constitutional symptoms and a palpable mass. Microscopic haematuria should be evaluated to exclude urothelial malignancy in the high-risk adult (chronic smokers, occupational exposure, older age) population. Pathology Both the PCS and the ureter have a thinner muscular layer than the bladder. Therefore, aggressive tumours of the upper tract can easily invade the muscle layers; hence, the prognosis is poor. Most of these tumours are caused by a field change; hence, tumours tend to be multifocal and may be associated with carcinoma in situ (CIS) in normal-looking urothelium. This also explains the higher incidence of recurrence of tumours in the bladder after successful treatment of upper tract disease. Therefore, long-term bladder follow-up with cytology and cystoscopy is necessary . Histological grading is of great prognostic significance. Low-

grade tumours follow a relatively benign course with mul tiple recurrences. High-grade tumour s are potentially invasive with a poor prognosis. Histological variants, such as micropap illary , neuroendocrine, sarcomatoid and squamous tumours, hav e a worse prognosis. Urothelial tumours can invade surrounding tissues, metas tasise to regional lymph nodes and spread haematogenously to lungs, liver and bones. Squamous cell and adenocarcinoma are rare non-urothelial malignancies involving the upper tract. They usually present at an advanced state and hav e a very poor prognosis. Squamous cell carcinoma occurs pr edominantly in the renal pelvis. Diagnosis Urinalysis may reveal numerous RBCs and white blood cells. Urine cytology should be obtained. The presence of atypical or malignant cells in a freshly voided sample has a high speci ficity for urothelial malignancies. CTU, cystoscopy , retrograde pyelogram and flexible ureterorenoscopy are required for diagnosis. CTU is the investig ation of choice. Findings suggestive of urothelial carcinoma are radiolucent filling defects, incomplete visualisation of calyces and the presence of hydronephrosis. CT also provides important staging information about local spread and lymph node involvement. Flexible ureterorenos copy may be used to visualise the ureter, renal pelvis and col lecting system and to biopsy suspicious lesions. URS biopsies can determine tumour grade and help in planning treatment. Incorporation of narrow-band imaging and blue light have improved the diagnostic capability of ureterorenoscopy . Staging Staging of upper tract urothelial cancer is similar to that for urothelial bladder cancer by the TNM system. Unifocal, small (<1 /uni00A0 cm), low-grade disease with no evidence of invasion on CTU is characterised as a low-risk tumour. Upper tract urothelial cancers that invade the muscle wall usually have poor prognosis. The 5-year survival is <50% and <10% for pathologically proven T2/T3 and T4 tumours, respectively . Management The management depends upon the stage, grade and risk stratification. Low-risk localised tumours may be managed with endo - scopic ablation or segmental excision. Kidney-sparing surgery is important in patients with solitary kidney , renal insu ffi ciency and synchronous bilateral tumours. High-risk tumours warrant radical nephroureterectom y with bladder cu ff resection with or without lymphadenectomy . Locally advanced disease is usually treated with cisplatinum-based neoadjuvant chemotherapy to downstage the disease prior to surgical ablation. Adjuv ant chemotherapy has been shown to improve surviv al.

Tuberculosis of the urinary tract

Tuberculosis of the urinary tract

Genitourinary tuberculosis (GUTB) accounts for 15–20% of extrapulmonary cases of TB. It is secondary and caused by haematogenous spread of tubercle bacilli from the thoracic lymph nodes or the lungs. GUTB occurs as a result of either reinfection or reactivation of old TB granulomas. Blood-borne organisms are deposited close to the glomeruli, causing an inflammatory reaction. Macrophages react and granulomas are formed. If bacterial multiplication goes unchecked, caseous necrosis results in the formation of tubercles. Multiple tubercles coalesce and rupture into the collecting system, caus ing intermittent tuberculous bacilluria and pyuria. The disease spreads through the collecting system with ulceration initially . When bacterial multiplication is halted by the immune system, sequelae due to fibrosis appear (Figure 82.8). Tubercular obstructing or destructive lesions in the kidneys and ureter responsible for renal function loss. Involvement of the bladder is secondary to renal disease. The disease gradually involves the bladder musculature, which is replaced by fibrous tissue, causing a decrease in the size and capacity of the bladder (‘thimble’ bladder). Urinary bladder involvement is responsible for urinary frequency , which is the most common symptom of GUTB. Epididymal tuberculosis presents as a painless epididy mal nodule, usually involving the tail of the epididymis, or a chronic discharging sinus in the posterior scrotal wall. Patients may present with urinary frequency , colicky flank pain, hae maturia and, rarely , fe ver and constitutional symptoms. They may also present with symptoms suggestive of recurrent UTIs William ‘Bill’ K Kerr , a Canadian urologist, described his eponymous sign in 1967. and, rarely , calcified tubercular lesions may be misdiagnosed as urinary tract calculi. For microbiological confirmation, a t least three consecutive early-morning specimens of urine are examined for acid-fast obiological diagnosis is urine bacilli. The gold standard f or micr culture. Nucleic acid amplification tests (NAATs) pro vide rapid diagnosis (within hours). When the diagnosis remains uncer - tain, bladder biopsy , tissue culture and tissue NAATs may be required. Imaging with CTU may also help and can show early signs such as calyceal distortion and papillary necrosis, hydronephrosis, poor function of r enal segments secondary - to parenchymal destruction, fibrosis and chronic obstruction. Ureteric strictures and proximal dilatation may also be seen. IVU can pick up the earliest signs of disease activity , such as calyceal distortion. Treatment involves short-course antituberculous ther - s are apy (ATT). Rifampicin, isoniazid and pyrazinamide are used sometimes with ethambutol as first-line drugs. The primary aim of therapy is preservation of renal func - tion and avoidance of fibrotic sequelae . Ureteric strictures may require double J (DJ) stenting to preserve function until defin - itive reconstruction is attempted. Percutaneous nephrostomy (PCN) is recommended in obliterativ e strictures to achieve prompt decompression. Definitive surgery is usually done 3–6 - weeks after starting A TT . The choice of reconstructive proce - dure depends on the type and location of sequelae. Open surgical repair is generally superior to balloon dila - - tation for tubercular ureteric strictures. Augmentation entero - cystoplasty (usually using

ileum) for small-capacity bladders,

radiological sign seen in an intravenous urogram, caused by microulceration of the calyces Caseous necrosis Ureteric stricture Thimble bladder: extremely small capacity bladder owing to /f_i brosis

ureteric reimplantation with or without a Boari flap (bladder tube) for lower ureteric stricture and ileal replacement of the ureter for multiple long ureteric strictures may be required. Nephrectomy is done for major renal lesions with a poorly functioning kidney . Urinary infection in childhood and vesicoureteric reflux All children with UTI must be evaluated for underlying predis posing conditions as recurrent pyelonephritis can cause renal scarring and loss of renal function. UTIs account for 7% of childhood febrile illness. In the age group <3 months, it is more common in males and in the age group >1 year , it is more common in females. Structural and functional abnormalities of the urinary tract such as VUR and posterior urethral valves predispose to UTI. Reflux is considered primary when it is due to an incompetent UVJ and secondary when it is due to increased bladder pressure or outlet obstruction. Presenting symptoms in neonates and infants include febrile illness or sepsis and may not be localised to the urinary tract. The method of urine sampling, especially before toilet training, is crucial and may involve suprapubic aspiration or per urethral catheter collection. A bacterial count of 50 /uni00A0 000 colony-forming units per millilitre is generally considered a positive culture result in children, although a lower count from a suprapubic aspirate in a symptomatic child is significant. The most important complication of UTI in a child is renal scarring secondary to renal parenchymal inflammation. US should be performed in all children, and children with recurrent UTIs or a first time UTI with pyelonephritis should be evaluated further. VCUG is the investigation of choice to diagnose reflux and should 99m be performed in high-risk children. Tc dimercaptosuccinic acid (DMSA) radionuclide cortical scan is the best modality to detect parenchymal lesions. VUR is present in approximately 30% of children with UTI and in up to 90% of children with Achille Boari , nineteenth century urological surgeon from Ferrara, Italy , described the technique of a bladder flap in dogs in 1894; it was first performed in a patient in 1936. renal scarring. Renal scarring may cause hypertension in up to 20% and is an important cause of renal failure. The grades of VUR are summarised in Figure 82.9 , with grades I–III generally resolving spontaneously . Low-dose nocturnal antibiotic prophylaxis to prevent scar-inducing pyelonephritis is the mainstay of treatment as the majority of reflux cases resolve with time. Ho wever, sur - gery (ureteric reimplantation, periureteric injections of Teflon or collagen) should be considered if episodes of acute pyelo - - nephritis recur despite antibiotic therapy or if severe reflux is accompanied by a surgically cor rectable malformation such as a paraureteric bladder diverticulum. Summary box 82.3 Infections /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF

Grade 1: Grade 2: Grade 3: re /f_l ux into undilated re /f_l ux into undilated re /f_l ux causing mild to ureter ureter and pelvis moderate dilatation of the ureter, renal pelvis and mild forniceal blunting of the calyces Figure 82.9 Grades of vesicoureteric re /f_l ux (courtesy of Nivedita Kekre and Dr Madhuri Sadanala). Grade 4: Grade 5: re /f_l ux causing moderate re /f_l ux causing gross ureteral ureteral tortuosity and tortuosity and gross pelvicalyceal dilatation pelvicalyceal dilatation with loss of papillary impressions of the calyces UTI: in /f_l ammatory response of the urothelium (host) to invading bacteria ABU: colonisation of urine with bacteria with no evidence of in /f_l ammation All pregnant women must be screened in the /f_i rst trimester for ABU because, untreated, one-third of

these patients will develop UTI GUTB: Always due to either reinfection or reactivation of old tuberculosis Urine examination: sterile pyuria in acidic urine Close monitoring of upper tract during the initial phase of ATT and timely urological intervention may prevent renal loss UTI in children: More common in girls after 1 year of age Sample collection method is important to avoid contamination VUR occurs in 30% of children with a UTI Renal scarring is a possible long-term consequence

Urolithiasis is as old as mankind. The first documented cystolithotomy was described by Sushruta, an ancient Indian surgeon in almost 600 /uni00A0 /b.sc/c.sc/e.sc . The development of shockwave lithotripsy (SWL) and endourological procedures with multiple e ffi cient energy-generating devices (such as US, pneumatic, electrohydraulic) for stone fragmentation have revolutionised the management of stone disease. Although the incidence of bladder stones has declined progressively owing to the allevi ation of poverty and the improvement in basic nutrition, the modern world is witnessing a steady increase in the incidence of renal calculi.

Types of stones

Types of stones

Calcium oxalate stones This is the most common type of stone, constituting 60–85% of all stones. Hypercalciuria, hypercalcaemia, hyperoxaluria, hyperuricosuria and hypocitraturia are known metabolic abnormalities that can predispose to its formation. Hypercalci - uria is the most common metabolic abnormality and occurs as a result of dysregulation of transport at various sites, including the intestine, bone or kidney . Primary hyperparath yroidism is the most common disease associated with hypercalcaemia and stone disease. Increased parathyroid hormone causes increased bone resorption and increased synthesis of 1,25-dihydr oxyvitamin D3. This causes increased intestinal absorption of calcium, leading to hypercal - - caemia and hypercalciuria. Hyperuricosuria causes uric acid crystal formation, espe - cially in association with acidic urine, over which calcium oxa - late crystals aggregate. Calcium phosphate stones Pure calcium phosphate stones are rare. Common forms seen are apatite and brushite stones. Apatite is seen with infection and brushite stones are usually seen with distal RTA. Uric acid stones Hyperuricosuria promotes the formation of both calcium oxalate and uric acid stones. Uric acid precipitates into crystals in acidic urine and remains soluble in alkaline urine. Conditions that can cause hyperuricosuria are gout and myeloproliferative disorders after cytotoxic treatment. Infection stones These are struvite and apatite stones. They form as a result of urease-producing bacterial infections, such as those caused by Proteus , Klebsiella , Serratia or Enterobacter . Alkalinisation of urine takes place as urease hydrolyses urea to carbon dioxide and ammonium. Staghorn calculi are infection stones that grow in a branch - ing pattern, taking the form of the pelvicalyceal system. significant morbidity , which includes loss of renal function owing to chronic infection and obstructive uropathy . Com plete clearance of a staghorn calculus is necessary , as residual fragments after treatment can cause rapid recurrence and per sistence of bacteriuria. Long-term chemoprophylaxis is man datory for a few months after successful r emoval of infection calculus. Cystine stones Cystine stones constitute approximately 1% of stones. Cystin uria is an autosomal recessive inherited disease that causes decreased reabsorption of cystine from the intestine and the proximal tubule of the kidney . Cystine is insoluble even at physiological pH and wor sens with increasing acidity . Cystine stones are very hard stones as a result of disulphide bonds and do not fragment with SWL.

URETERS Renal trauma

URETERS Renal trauma

Kidneys are retroperitoneal structures; they are relatively fixed by their vascular pedicles and are well protected by perinephric fat, strong posterior abdominal wall muscles and the lower rib cage. Renal trauma is usually a part of polytrauma and is present in only 5% of all trauma cases. Mechanism of injury Trauma may be penetrating or blunt, the latter being far more common. Common modes of injuries are head-on collisions in road tra ffi c accidents, falls from height and contact sports. Sudden, rapid deceleration can cause avulsion injury to the ureters at the PUJ or renal pedicle. Penetrating injuries cause direct tissue disruption and are usually associated with adjacent organ injuries. Presentation Patients with polytrauma may present with loss of conscious ness and haemodynamic shock because of associated injuries. Haematuria – gross or microscopic – is pathognomonic of renal trauma; however, its absence does not exclude serious renal trauma. Similarly , the sev erity of renal trauma does not correlate with the degree of haematuria. Trauma causing lower rib fractures with or without vertebral fractures and abdominal pain with flank contusions should raise suspicion about the underlying renal injury and should be evaluated further. Management CECT of the abdomen is necessary to delineate the type and extent of renal injury . It provides information about the pres ence of parenchymal laceration, its depth, extension into the pelvicalyceal system and the extent of urinary extravasation (Figure 82.11). It also provides valuable informa tion about other abdominal injuries and the status of the contralateral kidney . Based on the CECT , renal injuries are classified accord ing to the renal injury scale of the American Association for the Surgery of Trauma (Figure 82.12). Common indications for CECT in abdominal trauma include: /uni25CF abdominal trauma with gross haematuria; /uni25CF microscopic haematuria with hypotension (systolic <90 /uni00A0 mmHg); /uni25CF rapid deceleration injury; /uni25CF children with microscopic haematuria (>5 red blood cells [RBCs]); /uni25CF all penetrating injuries. Conservative management with close surveillance is su ffi cient in the majority of cases of isolated renal trauma. Lower grade injuries, sometimes including grade IV , can be managed conservatively with bed rest, antibiotics in case of penetrating injuries and serial haemoglobin estimation. Reimaging is usu ally done after 2–4 days in cases of falling haemoglobin le fever or expanding flank mass. Persistent urinary leak can be managed with an internal DJ stent or PCN. Indications for emergency surgical exploration are: - /uni25CF expanding or pulsatile retroperitoneal haematoma; /uni25CF PUJ avulsion; /uni25CF renal pedicle injury; /uni25CF haemodynamic instability .

Figure 82.11 Axial image showing pelviureteric injury with contrast in the peripelvic region (courtesy of Department of Urology, Christian Medical College, Vellore, India).

Ureteral duplication

Ureteral duplication

Duplication of the ureter (Figure 82.3) and renal pelvis is a common anomaly , with an incidence of approximately 1 in 150 births. Unilateral duplication is six times more common than bilateral. It is more common in females. The duplication may be incomplete (Y-shaped ureter) or complete. It is associ ated with vesicoureteric reflux (VUR), PUJO and ureterocele. Incomplete duplex ureters with a ‘Y’ ureter arise when the ureteric bud bifurcates after its initial development from the Wol ffi an duct. Complete ureteric duplication occurs when there are two separate ureteric buds that develop into two sepa rate ureters, which drain the upper and lower kidney moieties separately . The lower moiety ureter has a shorter submucosal tunnel and is prone to VUR. PUJO is more common with the often associated with a concomitant ureterocele. The upper moiety of the kidney is often dysplastic.

Ureteral trauma

Ureteral trauma

Most ureteral injuries are iatrogenic and occur during surgery near the ureter. Gunshot or penetrating injuries to the abdomen can cause ureteric injury . Management of ureteral injuries as a result of external trauma is dictated by the severity of trauma and associated visceral injuries. - Iatrogenic ureteral injury The overall incidence of iatrogenic ureteral injury varies between 0.5% and 1.0%. Hysterectomy accounts for most of these cases, followed by ureteroscopy . Common sites of injuries to the pelvic ureter are at the pelvic brim, where it might be - injured while ligating the infundibulopelvic ligament; at the bifurcation of the common iliac artery , while ligating the internal iliac artery; or at the paracervical region, while devel - oping the ureteric tunnel or while clamping and dividing the upper vagina. During open surgery , ureteral injury may be identified intraoperatively . However, most injuries (70–80%) are identi - fied postoperativ ely . The postoperative course of these patients can be di ffi cult and presentation may include abdominal pain, fever or sepsis. It is not uncommon to miss ureteric injuries; if left unrecognised, they can lead to significant morbidity , such as formation of urinoma, abscess, ur eteral stricture and uri - nary fistula. - Management of iatrogenic ureteral injury Triphasic abdomen and pelvic CECT is the imaging modality of choice. The choice of treatment is based on the location, type, - extent and timing of presentation. If an injury is recognised vels, intraoperatively or in the immediate postoperative period, it should be surgically repaired immediately . The viability of the ureter must be assessed. In cases of contusion, DJ stenting must be performed. In cases of partial transection, primary repair over a DJ stent may be performed. A tension-free spatulated ureteric anastomosis using fi ne absorbable sutures can be done for short segment loss. This is usually done in upper ureteric injuries (Figure 82.13). Longer segment loss, especially in the pelvic ureter, is managed by ureteroneocystostomy with or without a Boari fl ap. Longer defects up to 15 /uni00A0 cm can be repaired by mobilising and hitching the bladder to the psoas Summary box 82.5 Urinary tract trauma /uni25CF /uni25CF /uni25CF /uni25CF major muscle (psoas hitch) with a Boari fl ap (Figure 82.14). Transureteroureterostomy (anastomosing the injured ureter to the contralateral ureter) can be an option in selected situations. Delay in diagnosis would result in late presentation and delay the defi nite repair by 2–3 months to allow resolution of urinoma and periureteric infl ammation. In such situations an initial endourological approach either with a retrograde ureteric stent or with PCN would decrease the morbidity associated with urinoma and help to preserve renal function.

Kidney capsule Peritoneum Subcapsular Haematoma haematoma Laceration <1 cm Grade IV Laceration into collecting system Figure 82.12 Classi /f_i cation of renal trauma. Suspect renal injury in abdominal trauma in adults with modes of injury such as sudden deceleration, penetrating injury directed towards the renal bed, hypotension and haematuria and in children Triphasic (arterial, nephrogenic and delayed phase) CT is the investigation of choice to diagnose and grade urinary tract injury in a haemodynamically stable patient Most grade I–IV renal trauma (including penetrating injuries) can be managed conservatively Most ureteric injuries are iatrogenic and

prompt diagnosis will prevent morbidity Laceration

> 1 cm Grade V Renal artery or vein injury Arterial blood clot from endothelial Avulsion injury of hilum Kidney shattered

Ureterocele

Ureterocele

Ureterocele is a cystic enlargement of the intramural ureter, which probably occurs as a result of atresia of the ureteric orifice. It has a female-to-male ratio of 4:1 and occurs bilat - erally in 10%. Similar to ectopic ureters, ectopic ureteroceles frequently drain the upper pole and are often associated with dysplastic or non-functional renal tissue. In childhood, they usually present with infection. When large , they can obstruct the bladder neck or even the contralateral ureteric orifice. The classic feature of a ureterocele on an intravenous urogram (IVU) is the ‘cobra head’ sign. The treatment of simple uret - eroceles is surgical excision with reimplantation of the ureter. Endoscopic incision of a ureterocele is the preferred treatment method for simple ureteroceles in infants and small children, but may result in subsequent ureteric r eflux. A non-functioning kidney may need nephrectomy . -

Wilms’ tumour

Wilms’ tumour

See also Chapter 17 . This is the most common tumour of childhood, account - ing for 5% of all childhood cancers. They are bilateral in 5% of cases and familial in 1%. The tumour has mixed elements derived from the embryonic nephrogenic tissue, namely blas - temal or undi ff erentiated tissue, epithelial tubules and stroma. The typical presentation is a child aged between 1 and 4 years of either gender with a large, palpable abdominal mass that may cross the midline. It may also be associated with haema - turia, hypertension, fever and weight loss. Pain is relatively uncommon. The large tumour can rupture and present as an acute abdomen. Other causes of renal masses include neuro - blastoma, congenital mesoblastic nephroma, RCC, clear-cell sarcoma and rhabdoid tumour. US can confirm the r enal origin and solid nature of the - mass. Further definitive imaging with either CECT or MRI is necessary to stage the disease. Up to 13% of patients hav e bilateral tumours. The tumours usually infiltrate the kidne ys and normal renal parenchyma is compressed at the periphery around the tumour (claw sign). A CT of the chest should be obtained as the lung is the most common site of distant metastasis. Current treatment is nephrectomy with pre- or postoperative chemotherapy . Both regimes have a comparable survival of ~90%. Tumours of the kidney and ureters /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Brierley JD, Gospodarowicz MK, Wittekind C (eds). TNM classification of malignant tumours , 8th edn. Oxford: Wiley , 2016. Available from https://www .uicc.org/8th-edition-uicc-tnm-classification-malig - nant-tumors-published/. Khan F , Ahmed K, Lee N et al . Management of ureteropelvic junction obstruction in adults. Nature Rev Urol 2014; 11 (11): 629–38. Moore EE, Shackford SR, Pachter HL et al . Organ injury scaling: spleen, liver, and kidney . J Trauma 1989; 29 (12): 1664–6.

Rule out urothelial malignancy in high-risk adults (chronic smokers, occupational exposure, older age) with microscopic haematuria Nephron-sparing surgery should be considered in small renal masses to preserve renal function, more so in patients with compromised renal function PNSs are found in up to 30% and IVC tumour thrombus in 5–10% of patients with RCC Targeted therapy and immunotherapy have improved survival in metastatic RCC Wilms’ tumour is the most common renal tumour in children <15 years old and should be treated in a multidisciplinary setting