113 Kidney, urinary bladder, and ureter

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Bilateral anephrogenesis has an incidence of 0.0017%. Heterotopic renal tissue has been reported in the heart and in the suprarenal gland. The kidneys sometimes vary from their normal form, being either longer and narrower or shorter and more rounded. The occurrence of an additional kidney is rare. The characteristic fetal lobulation may persist in the adult (7%). Occasionally, one kidney is very small while the other is proportionally enlarged.

Ectopia or anomalies of ascent-pelvic ectopia

Renal ectopia or ectopic kidney is a congenital anomaly in which at least one kidney is located in an unusual position. It results from the kidney failing to ascend properly from its origin in the true pelvis. Normally, both kidneys are situated posteriorly behind the peritoneum on each side of the vertebral column, at the level of the upper border of the twelfth thoracic vertebra superiorly and the third lumbar vertebra inferiorly (Bergman et al. 2006). Ectopic kidneys, however, can be located anywhere along their path of ascent in the pelvic, iliac, or abdominal areas. The pelvic kidney is opposite the sacrum and below the aortic bifurcation; the lumbar kidney rests near the sacral promontory in the iliac fossa and anterior to the iliac vessels; and the abdominal kidney lies above the iliac crest and adjacent to the second lumbar vertebra (Bauer 2002). There is a slight predominance on the left side and in males. Bilateral pelvic ectopic kidneys can occur with or without fusion. In some cases, one kidney crosses over (crossed renal ectopia) so that both kidneys are on the same side of the body. When a crossover occurs, the two kidneys can grow together and become fused (crossed fused renal ectopia). Since under-ascent is more common than over-ascent, ectopic kidneys are more commonly found in the pelvis or lower abdomen. Over-ascent leads to the kidneys being identified as subdiaphragmatic or intrathoracic (cf. Sfaxi et al. 2002). Factors that can prevent orderly movement of the kidneys include ureteral bud maldevelopment, defective metanephric tissue, genetic abnormalities, maternal illness, and teratogenic causes (Ascher and Rosch 2005). Initially, the kidneys lie close together in the pelvis. They gradually ascend into the abdomen and become separate from each other and inferior to the adrenals by the ninth gestational week. As they ascend they gradually rotate, with the hila starting to be seen anteromedially. Renal ectopia should be differentiated from renal ptosis, in which the kidney is initially located in its proper place with normal vascularity but then moves downward (Bauer 2002). The ureter usually enters the bladder on the ipsilateral side with its orifice situated normally (Bauer 2002).

Incidence of renal ectopia

Ectopic kidneys are thought to occur in approximately 1 in 1000 births, but only about 1 in 10 of these cases is ever diagnosed: it is asymptomatic in most patients, which explains why the incidence in autopsy series (1:1000) is much higher than in clinical presentation (1:10,000) (Moore and Persaud 2008). A radiographic survey of symptom-free potential transplant donors found ectopic kidneys in 2 of 151 individuals (Frick and Goldberg 1980); in earlier reports the incidence was given as 1:2100 and 1:3000 births (Stevens 1937) and 1 in 5000 children (Gleason et al. 1994). A frequency of 1:500 has also been reported by some authors (Bergman et al. 2006). Pelvic ectopic kidney, usually opposite the sacrum and below the aortic bifurcation, has been estimated to occur in 1 of 2100–3000 autopsies without any sex difference (Tsao et al. 2008).

Embryology of renal ectopia

The kidney generally ascends in the developing metanephric blastema in the five- to seven-week embryo. It does not rotate until it reaches its normal lumbar position by the ninth gestational week. The ascent of the kidneys precedes the descent of the gonads into the pelvis. During ascent, each kidney acquires its blood supply from neighboring vessels, initially from the external and internal iliac vessels, but by the eighth week directly from the aorta. An ectopic kidney results from failed ascent of the fetal kidney. If the aortic blood supply is not acquired normally, or if there is an associated abnormality of the spine, cephalad migration of the fetal kidney will not occur (Arnim et al. 2005). During the mesonephros stage, lateral intersegmental arteries (urogenital rete arteriosum), located

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on both sides of the aorta between the sixth cervical and third lumbar vertebrae, provide vascularization to the adrenal glands, kidneys, and gonads. The kidneys receive branches from vessels close to them during their ascent, first from the common iliac artery, then from the distal end of the aorta; when the highest level is attained they receive new branches from the aorta and the former vessels degenerate. A permanent renal artery develops from one of the persisting branches (Sadler 1990). Malascent can occur because of ureteral or metanephric factors. The urinary tract originates from the mesoblast and has three developmental phases: pronephros, mesonephros, and metanephros. The third phase gives rise to the permanent kidneys. Initially, they localize close to each other at the ventral portion of the sacrum and pelvis. As the abdomen and pelvis grow and the body inclination decreases, they ascend and come into contact with the adrenal gland during the ninth week. The renal hila initially face the ventral aspect during ascent, then after a 90 degree medial turn the hilus of each kidney attains its final permanent position facing the anterolateral aspect. During ascent, the kidneys proceed through a bifurcation formed by the umbilical arteries. If one of the kidneys fails to pass this point, it remains within the pelvis next to the common iliac artery (Sadler 1990; Cocheteux et al. 2001; Cinman et al. 2007).

Signs of pelvic kidney

Most patients with an ectopic kidney are clinically asymptomatic. Signs and symptoms, when they occur, usually develop in the third or fourth decade of life and include vague lower abdominal pain, hematuria, and fever (Kato et al. 2000; Bhatnagar et al. 2004; Boyan et al. 2007). Vague abdominal complaints and pain due to an obstructing stone remain the most frequent symptoms of pelvic ectopic kidney. Acute pyelonephritis can present with the characteristic clinical and laboratory features of a ruptured appendix (Tsao et al. 2008). The prevalence of vesicoureteral reflux (VUR) and pelvic dilatation in renal ectopia is estimated at around 20%, although Calisti et al. (2008) found only a 2% prevalence of VUR. Dilatation of the renal pelvis could result from primary ureteropelvic or ureterovesical obstruction and extrarenal collecting systems with malrotation obstructing the ureteropelvic junction (Gleason et al. 1994). VUR occurs in 30% of children with ectopic kidneys (Guarino et al. 2004). The diagnosis of ectopic kidney in the pelvis can be made by ultrasonography (Lu et al. 2011). Malposition of the colon can suggest the ectopic position of a lumbar or pelvic kidney. The diagnosis is easily made when the excretory urogram or renal ultrasound fails to reveal a kidney in its proper location (Bauer 2002). Ectopic kidneys are more susceptible to the development of hydronephrosis, urinary tract infection, and urinary calculus. The abnormal position of ectopic kidneys results in atypical patterns of direct or referred pain and can be misdiagnosed as acute appendicitis. Pelvic organ inflammatory disease and dystocia could be signs of a pelvic kidney in women. A palpable abdominal mass and renovascular hypertension secondary to an anomalous blood supply could also be signs of pelvic and abdominal kidneys (Kumar et al. 2011). An ectopic kidney is often associated with other abnormalities such as agenesis of the opposite kidney, vascular malformation, and genital anomalies (Kato et al. 2000; Bhatnagar et al. 2004; Boyan et al. 2007).

Abnormalities associated with a pelvic kidney

Many urological abnormalities are associated with the orthotopic kidney, suggesting that renal ectopia is part of a urinary tract malformation complex. This underscores the need for a thorough examination including voiding cystourethrogram (VCUG) and radioisotope studies (Gonzalez et al. 2007). The associated abnormalities most commonly involve the genitourinary, musculoskeletal, and cardiovascular systems. Abnormalities of the uterus are found in 20–66% of women with renal ectopia: unicornuate uterus with or without rudimentary horn, bicornuate or absent uterus, and vaginal atresia or vaginal duplication have been reported (Bauer 2002) and cryptorchidism, hypospadias, or urethral duplications can occur. In most patients with an ectopic kidney, the contralateral kidney is normal. The most common congenital cardiovascular anomalies are septal or valvular defects; pulmonary agenesis and dextrocardia have rarely been reported with an ectopic kidney (Eroglu et al. 2005). Ureteral ectopia can be associated with imperforate anus and tracheoesophageal fistula (Cinman et al. 2007). Prenatal diagnosis of pelvic kidney is available after 24 weeks of gestation (Meizner et al. 1995). In men, associated genital abnormalities are seen in 10–20% of patients, including undescended testes, urethral duplication, and hypospadias (Kocak et al. 2001). The most common arterial variation of an orthotropic kidney is the presence of accessory arteries (28%). Accessory renal arteries vary in size and are usually derived from the aorta (26–30% of all reported kidneys); they can enter the kidneys at any point. The renal veins are less variable than the renal arteries; variations are more common on the right side (28%) (Bergman et al. 2006). Up to 50% of renal units contralateral to pelvic kidneys demonstrate some form of anatomical abnormality, and up to 10% are absent. The ectopic kidney is usually smaller than normal and does not necessarily conform to the usual reniform shape because fetal lobulations are retained. The axis of the kidney is slightly medial or vertical but can be tilted as much as 90° laterally, so that it lies in a true horizontal plane (Someren 1989; Gleason et al. 1994; Benchekroun et al. 2002). The association of pelvic kidney with aorto-iliac aneurysm is rare and has been reported in only 17 cases (Hanif et al. 2005; Marone et al. 2008). An ectopic (pelvic) kidney can be wrongly interpreted as a bulky lymph node, potentially subjecting the patient to operation (Bader et al. 2005). A severe atheromatous involvement of the distal aorta can impair the blood supply to the ectopic kidney (Dretler et al. 1971). Most ectopic kidneys have hydronephrosis of the collecting system. Half of these cases result from obstruction at the ureteropelvic or the ureterovesical junction

(Bauer 2002). An anteriorly placed collecting system of a pelvic kidney and an anomalous vasculature can partially block the major calyces and predispose the ectopic kidney to hydronephrosis. In addition, there is an increased risk of injury from blunt abdominal trauma because the low-lying kidney is not protected by the rib cage (Glodny et al. 2001; Benchekroun et al. 2002; Arnim et al. 2005; Ho et al. 2013). Association of a fused ectopic multicystic dysplastic kidney with ureterocele has been reported (Kiddoo et al. 2005).

Complications of pelvic kidney

Available data suggest no adverse effects on blood pressure or kidney function in children with renal ectopia. Vascular variations associated with an ectopic kidney can predispose to iatrogenic trauma during interventional procedures and emergency operations (Kara et al. 2011). Giant hydronephrosis in an ectopic kidney is an extremely rare disorder that can present as an asymptomatic abdominal mass (Hsieh et al. 2013). The vessels supplying an ectopic kidney usually arise from the nearest large vessels such as the iliac vessels. Trauma to these vessels can cause considerable bleeding and can lead to partial renal infarction or nephrectomy (Aslan et al. 1999). Ectopic kidneys can be associated with a variety of vascular anomalies, including anomalies of the contralateral renal vessels and branches and tributaries of the abdominal aorta and inferior vena cava (Belsare et al. 2002). A pelvic kidney interpreted as a lymph node conglomerate can create some diagnostic confusion (Chao 2012). Ectopic pelvic kidneys have been utilized successfully for kidney transplantation. However, all these donor nephrectomies were performed by open surgery (He and Mitchell 2012). In a woman with breast carcinoma presenting for a follow-up bone scan, initial inspection suggested sacral metastasis; on closer review an absence of renal activity in the left flank was however noted (Pryma and Akhurst 2005). Correlation with a CT scan confirmed a hydronephrotic ectopic kidney overlying the sacrum. Dystocia from a pelvic kidney is a very rare finding, but when it does occur early recognition is mandatory and cesarean section is indicated (Bauer 2002). In acute and elective obstetrical and gynecological operations, the pelvic kidney is susceptible to iatrogenic trauma (Dabiri and Cheung 2006). A pelvic kidney creates potential difficulties in the management of patients requiring aortic surgery (Krahn and Taylor 1993). The renal pelvis is usually anterior to the parenchyma, and surgical operation for acute appendicitis could create complications (Kara et al. 2011). Ectopic kidneys can be associated with autosomal dominant polycystic kidney disease (ADPKD) and a pelvic kidney has been associated with a reverse sigmoid colon (Chen et al. 2010). Renal cell carcinoma (RCC) is a very rare phenomenon in an ectopic kidney; there are only seven reports of RCC in pelvic kidneys (Mahmoudnejad et al. 2009). Surgical approach to ectopic kidneys merits caution because of the uncertain vascular anatomy (Hernández Toriz et al. 2006). In patients with pelvic kidneys, anterior views must be obtained during radionuclide scanning if unnecessary nephrectomy is to be avoided (Allen et al. 2005).

Horseshoe kidney

Horseshoe kidney (HSK) consists of two distinct kidneys lying vertically on either side of the midline and connected at their respective lower poles by a parenchymatous or fibrous isthmus that crosses the midplane of the body. It was first recognized during an autopsy by DeCarpi in 1521 (Weizer et al. 2003). It represents one of the most commonly renal anomalies with a reported incidence of 1/400 to 1/1600 based on autopsy series performed half a century ago. It combines three anatomical abnormalities: sectopia, malrotation, and vascular changes. In rare cases the isthmus connects the upper poles and an inverted HSK is created (Yoshinaga et al. 2002). The kidneys are somewhat low lying and close to the vertebral column, so that a line drawn through the midplane of each kidney bisects the midline inferiorly; this is the characteristic orientation of the collecting system, which is directly posterior to each renal pelvis (Strauss et al. 2000). The failure of normal rotation in HSK cases is responsible for the atypical arrangement of the pelvicalyceal system. The renal hilum lies anteriorly to the calyces, which can point inward toward the spinal column, downward, or both (Gupta et al. 2007).

The presence of HSK can be confirmed by an ultrasound scan or an IVP. When abdominal and retroperitoneal images from 15,320 radiographs, especially from computerized tomography (CT), were evaluated, 23 patients were identified with HSK giving an overall incidence of 1/666. This closely matches past autopsy series in which the reported incidences were 1/400 to 1/1600. The abnormality has been discovered clinically in all age groups ranging from fetal life to 80 years; it is more common in males by a margin slightly greater than 2:1 (Basar et al. 1999; Hobbs et al. 2002; Weizer et al. 2003). No genetic and racial predominance is known, although HSK has been reported in identical twins and in siblings within the same family (Gleason and Kraemer 1995; Yoshinaga et al. 2002). In a study of 1957 potential living kidney donors, solitary or horseshoe or pelvic kidney was found in 0.2% (Lorenz et al. 2010).

HSK is usually asymptomatic and it is revealed incidentally during imaging, but gastrointestinal symptoms such as pain, nausea, and vomiting can occur. In 5% of cases HSK is diagnosed when patients are evaluated for an abdominal mass, a vague abdominal pain possibly radiating to the lower lumbar region. Symptoms, if any, are related to hydronephrosis, infection, or calculus formation. A possible sign is abdominal pain, nausea, and vomiting on hyperextension of the spine. Horseshoe kidneys have been detected after angiography for evaluation of an abdominal aortic aneurysm (Huber et al. 1990; de Brito et al. 1991). An obstruction causing significant hydronephrosis occurs in one-third of patients. Multiple retinal arteries are found in most HSKs and can be an intraoperative hazard for patients due to the risk of unexpected hemorrhage. Precise imaging using modern cross-sectional imaging techniques is therefore very important for preoperative planning (Natsis et al. 2014).

The ureteral bud arises from the Wolffian duct at the end of the fourth week and grows craniad toward the urogenital ridge, acquiring a cap of blastema by the fifth week. The developing metanephric tissue and ureteral bud migrate cephalad, rotating medially on its long axis. The entire process is completed by the eighth week of gestation. The abnormality arises between the fourth and sixth weeks, after the ureteral bud has entered the renal metanephric blastema. Posterior nephrogenic cells migrate abnormally to form an isthmus or connection between the two developing kidneys, creating the horseshoe shape. Because the kidney fails to rotate, the calyces point posteriorly and the axis of each pelvis remains in the vertical or obliquely lateral plane (Weizer et al. 2003).

There are two hypotheses regarding the embryogenesis of HSK. The classical proposal is that during the metanephric stage (fourth week of gestation) while the kidneys are still in the pelvis, their lower poles come into contact and fuse at the midline, forming a HSK with a fibrous isthmus. This fusion can be attributed to abnormal flexion or growth of the developing spine and pelvic organs, causing fusion of the nephrogenic blastemas of the immature kidneys, which lack renal capsules. Normally, the kidneys migrate out of the pelvis during the seventh and eighth weeks and at the same time rotate so that the anteriorly facing renal pelvis turns medially. At the level of the lower lumbar vertebrae, the inferior mesenteric artery (IMA) blocks their upward migration (Cascio et al. 2002; Tijerina et al. 2009). The alternative proposal is that HSK results from a teratogenic event involving the abnormal migration of posterior nephrogenic cells to form a parenchymal isthmus. This could explain the increased incidence of malignancies associated with HSK, such as Wilms' tumor and HSK carcinoid (O'Brien et al. 2008). Prenatal diagnosis is also possible as early as the first trimester of pregnancy, using high-frequency transvaginal sonography (Bronstein et al. 1990).

HSK patients are at risk of reflux, stasis, stone, and infection. Infections complicate up to one-third of HSK patients and are a major cause of death among them (Fernbach and Davis 1986). A variety of benign and malignant tumors are also associated with HSK. The increase in malignancy is attributed to teratogenic factors. Most cases are associated with renal cell carcinoma. Transitional cell carcinoma is related to chronic infections and stone. The incidence of Wilms' tumor is higher than in the general population. Benign tumors that usually accompany HSK include renal angiolipoma and oncocytoma (O'Brien et al. 2008). Horseshoe kidney should not adversely affect pregnancy or delivery. The lower ureter usually enters the bladder normally and is rarely ectopic. Any ureteropelvic junction (UPJ) obstruction probably results from high insertion of the ureters into the renal pelvis. In rare cases, crossing of the ureter over the isthmus can also contribute to a higher prevalence of UPJ. The main complication of HSK is stone formation (16–60% of cases). The orientation of the calyces impairs proper drainage, resulting in stasis and hence stones which are often multiple; there is a significantly increased incidence of staghorn stones (Natsis et al. 2005). HSK predisposes the patient to serious injuries following abdominal trauma (Natsis et al. 2013).

The isthmus is bulky and consists of parenchymatous tissue with its own blood supply. Occasionally it is just a flimsy midline structure composed of fibrous tissue. It usually lies anterior to the aorta and vena cava. Teratogenic factors could be responsible for the abnormal migration of nephrogenic cells to form an isthmus and for the increased potential for carcinoma development in this portion of the kidney (Doménech-Mateu and Gonzalez-Compta 1988; Hohenfellner et al. 1992). The incidence of Wilms' tumor in horseshoe kidneys is more than twice that in the general population. Renal carcinoma has been reported within a horseshoe kidney (Mesrobian et al. 1985; Neville et al. 2002). The isthmus is situated at the level of the third or fifth lumbar vertebra, under the origin of the inferior mesenteric artery (IMA) and anterior to the abdominal aorta (AA) and inferior vena cava (IVC). Rarely, the isthmus is reported to lie posteriorly to the vessels or to run between them. Sometimes it is located at the pelvis or at the sacral promontory (O'Brien et al. 2008).

There is wide variation in the arterial supply to HSKs, considering that the renal arteries (RA) can originate from the AA, the common iliac arteries (CIA), and the IMA. The isthmus can be supplied by a single vessel derived from the AA, or alternatively its blood supply can originate from CIA or IMA (Ferko et al. 1997). The HSK can be supplied by both normal and accessory RAs. With the exception of some of the arteries to the isthmus, no vessel on the dorsal aspect of the kidney has been described. Multiple RAs have been reported in 45 out of 71 reported HSK cases (63%); one or two RAs have been reported in the other 26 (37%). Because of these variations, *en bloc* removal of an HSK with large segments of the AA and IVC is proposed when the HSK is considered as an allograft organ (Stroosma et al. 2000; Goyal et al. 2003). The HSK is frequently accompanied by a retrocaval ureter (Shen et al. 2012) and there are IVC abnormalities such as double, left, and pre-isthmic IVCs in 5.7% of cases (Radermecker et al. 2008; Ichikawa et al. 2012). It is also argued that HSK is associated with cardiovascular malformations (Greenwood et al. 1976) and it is frequently associated with vascular and ureteral abnormalities: ureteral duplication, ureteroceles, UPJ obstruction, vesicoureteral reflux, hydronephrosis, and nephrolithiasis. In 30% of cases there is one RA per kidney. The blood supplies to the isthmus and lower poles are also variable; the isthmus and adjacent parenchymal masses can receive a branch from each main renal artery or have their own arterial supplies originating directly from the aorta or even from branches originating from the inferior mesenteric, common or external iliac, or sacral arteries.

There are many variations in the number and origin of renal-collecting systems in HSK. As a rule, calyces are located in the upper two-thirds of each kidney and an extrarenal calyx or an independent ureter drains the isthmus. The ureters normally end in the bladder, but they can also be ectopic (Cascio et al. 2002). Retrocaval ureter (RCU) is a rare congenital abnormality

associated with HSK, and a pre-isthmic IVC is the direct cause of hydronephrosis and UPJ obstruction. Because the kidney fails to rotate, the calyces point posteriorly and the axis of each pelvis remains in the vertical or obliquely lateral plane.

There are central nervous system problems in 3% of children with HSK. Anorectal abnormalities are frequently encountered in these patients (Salerno et al. 2000). Duplication of the ureter occurs in 10% of patients; vesicoureteral reflux has been noted in more than half of affected individuals (Cascio et al. 2002). DMSA scanning (radionuclide scanning using dimercaptosuccinic acid) of 22 patients with HSK revealed asymmetrical function in 63% (Kao et al. 2003). Metabolic evaluation of 37 patients with HSK revealed altered calcium, oxalate, uric acid, and citrate excretion in 50% of stone formers, suggesting an additional underlying metabolic etiology (Raj et al. 2004). Patau and Gardner syndromes (trisomy 13 and deletion q15q22) have been reported to coexist with HSK. It has also been mentioned that 20% of Down and Edwards syndromes (trisomies 21 and 18, respectively) and 60% of Turner syndromes possess a HSK. The HSK has rarely been related to oral–cranial–digital syndrome or other skeletal abnormalities such as kyphosis, scoliosis, and macrognathia, or to neurological abnormalities. Other associated abnormalities include supernumerary kidneys with horseshoe elements, undescended testis, septate vagina and hypospadias (Mandell et al. 1996; O'Brien et al. 2008).

Sigmoid-shaped kidney

The sigmoid or S-shaped kidney is the second most common anomaly of fusion. The crossed kidney is inferior with the two kidneys fused at their adjacent poles. Each renal pelvis is oriented correctly and they face in opposite directions from one another. An S-shaped kidney is the source of trouble and urogram appearance. The estimated occurrence of a sigmoid-shaped kidney is 1:1000. The lower convex border of one kidney is directly opposite the outer border of its counterpart. The adrenal glands are normally situated. The ureter from the normal kidney courses downward anterior to the outer border of the inferior kidney, and the ectopic kidney's ureter crosses the midline before entering the bladder; at times, the ectopic ureter may pass behind the rectum as it crosses the pelvis.

Intrathoracic kidney

Intrathoracic renal ectopia is a rare congenital malformation. It is often asymptomatic and diagnosed incidentally during the work-up for a possible lung mass near the midline and on the posterior mediastinum along the posterior aspect of the diaphragmatic leaflet on a lateral view (Vázquez et al. 2008). It does not require treatment. Its incidence ranges over 0.5–5% of renal ectopia, with a prevalence of one in 10,000 cases. The condition is rarely bilateral; it occurs mostly on the left side (61%) and in males (1.7:1) (Subramanian and Goldfarb 2008). Four forms of intrathoracic kidney have been documented: a true thoracic ectopia with normally developed diaphragm, congenital diaphragmatic eventration, diaphragmatic hernia, and renal ectopia associated with traumatic rupture of the diaphragm (Pfister-Goedeke and Brunier 1979; Donat and Donat 1988; Hidaka et al. 2012). Wolfromm (1940) reported the first case of intrathoracic kidney, clinically diagnosed by retrograde pyelography in a 43-year-old woman. Intrathoracic ectopia denotes either a partial or a complete protrusion of the kidney above the level of the diaphragm into the posterior mediastinum. A high-positioned kidney can lead to focal eventration of the overlying diaphragm, mimicking a supradiaphragmatic renal position.

Congenital intrathoracic kidney is a rare developmental anomaly; in 13,000 autopsies 27 ectopic kidneys were encountered, and only one of them was intrathoracic (Campbell 1930). This condition is to be differentiated from a congenital or traumatic diaphragmatic hernia, in which other abdominal organs as well as the kidney have advanced into the chest cavity (Drop et al. 2003).

A superior ectopic kidney can appear above, below, or partly through the diaphragm. In the supradiaphragmatic position the kidney lies opposite T7–T9, while in the transdiaphragmatic position it lies opposite T7–11. True thoracic ectopia with a normally developed diaphragm is less common than other types (Kubricht et al. 1999). The kidney usually has a rotation anomaly with the hilus facing anteriorly, a high origin of the renal vessels, a long normal ureter, and medial deviation of the lower pole. Nevertheless, the function of the affected kidney is usually well preserved (Sözübir et al. 2005; Obatake et al. 2006; Dingeldein et al. 2008; Fadaii et al. 2008). A combination of intrathoracic liver and kidney has been reported (Lee et al. 2006). The ureter is elongated to accommodate the excessive distance to the bladder, but it never enters ectopically into the bladder or other pelvic sites.

The adrenal gland is below the kidney, in its normal location, in most of these patients (N'Guessan and Stephens 1983). An abnormally high ascent of the metanephros generates a diaphragmatic defect and subsequently an ectopic kidney in the thorax. An intrathoracic ectopic kidney can be either congenital or acquired. The mechanism by which the kidney migrates to a superior position is not known. It reaches its adult location by the end of the eighth week of gestation when the diaphragmatic leaflets are formed as the pleuroperitoneal membrane, which separates the pleural from the peritoneal cavity. Mesenchymal tissues associated with this membrane eventually form the muscular component of the diaphragm. Delayed closure of the diaphragmatic leaflets allows the kidneys to ascend above the level of the future diaphragm (N'Guessan and Stephens 1983). Delayed involution of mesonephric tissue has also been proposed as a causal factor (Angulo et al. 1992), and intrathoracic kidneys occur in only 0.25% of patients with a diaphragmatic hernia (Donat and Donat 1988). Renal angiography demonstrates either a normal site or a more cranial origin for the renal artery takeoff from the aorta supplying the thoracic kidney.

Plain chest X-ray usually reveals a paravertebral mass in the posterior mediastinum. It should be differentiated from other posterior mediastinal masses. Intravenous urography has been used as a routine method of diagnosis for differentiating other lesions such as mediastinal tumors and hernia (Obatake et al. 2006; Rouanne et al. 2010). The affected hemidiaphragm is usually slightly elevated. The diagnosis should be considered in a patient with a mass at the base of the lung on a chest radiograph (Sidhu et al. 2001). The diagnosis is confirmed by CT or contrast-enhanced CT scan and intravenous urography or magnetic resonance imaging of the kidney (William and Jeans 1996; Clarkson and Potter 2009). Renal cortical scintigraphy could be used to confirm the functional status of the kidney tissue (Louzir et al. 1999; Sözübir et al. 2005). In a reported case of atypical chest pain after myocardial scintigraphy that revealed a large thallium-avid mass in the right hemithorax, subsequent photon emission computed tomography/computed tomography demonstrated the mass to be an ectopic kidney (Du et al. 2010).

Most affected individuals remain asymptomatic. Pulmonary symptoms are exceedingly rare and urinary symptoms are even more infrequent. Most previously reported cases were asymptomatic and were discovered incidentally during investigations for other medical problems. The usual complications such as infection, stone formation, and ureteropelvic junction obstruction that accompany inferior ectopic kidneys such as pelvic kidney are rarely seen in thoracic cases. There are occasional reports of association with nonspecific chest, abdominal, and flank pain (Oon et al. 2005; Fadaii et al. 2008; Subramanian and Goldfarb 2008; Ahmed et al. 2011), urinary stones, or renal carcinoma (Kubricht et al. 1999; Lenz et al. 2003). Spontaneous intracranial hypotension presenting as aggravated headache after sitting and standing was the presenting sign in a young adult with intrathoracic kidney; a focal paraspinal area of increased uptake first supposed to be a site of CSF leakage was then identified by radionuclide cisternography as an intrathoracic renal ectopia (Chiu et al. 2012; Pandey et al. 2012). Left thoracic kidney associated with left Bochdalek hernia has been reported in late adulthood (Karaoglanoglu et al. 2006; Fiaschetti et al. 2010). History of trauma should be considered in patients with intrathoracic kidney (Suárez and de Jesús 1998). Intrathoracic kidney can also be discovered incidentally in the elderly (Sfaxi et al. 2002).

In the neonatal period, sudden breathing difficulties and recurrent attacks of pulmonary infections, cough, wheezing, and vomiting have been reported as the first presenting sign (Karaoglanoglu et al. 2006; Maduekwe et al. 2011). Intrathoracic ectopia with a Bochdalek defect has been reported as a right posterior mediastinal mass and intestinal gas in the right lung field (Obatake et al. 2006). Color Doppler can demonstrate an abnormal course of the renal artery, arising from the aorta and feeding the intrathoracic right kidney; this could provide a diagnosis of thoracic kidney in the prenatal period (Masturzo et al. 2001). Only 13 cases of intrathoracic kidney have been reported in the pediatric age group over the past 25 years. Although the early literature recommended an aggressive surgical approach for all intrathoracic kidneys, surgeons have taken a more selective approach during recent decades. Surgical treatment has been reserved for children with associated bowel herniation or respiratory compromise, as in the case of a child with bilateral intrathoracic kidneys. There is concern about the ongoing growth and development of intrathoracic kidneys due to the potential for vascular or ureteric obstruction. Most symptomatic patients present with respiratory distress (Murphy et al. 2012). A persistent cough for five months with no history of pulmonary infection, and respiratory distress immediately after birth, were the first clues to intrathoracic kidney in a newborn infant and a neonate (Nouri-Merchaoui et al. 2011). It is extremely rare for an intrathoracic kidney to be detected prenatally in a fetus with congenital diaphragmatic hernia (CDH) (Pfister-Goedeke and Brunier 1979; Hidaka et al. 2012). Pneumomediastinum has been reported as the presenting sign of intrathoracic kidney in a neonate (Matsubara et al. 2000).

When the kidneys are not in their normal position but the bladder and amniotic fluid volume are normal, thoracic kidneys should be suspected (Muttarak et al. 2001). Color Doppler ultrasound imaging facilitates identification of the kidneys, as the renal arteries can be seen coursing in a cephalad direction (Suresh et al. 2007). Ureteropelvic junction obstruction in a thoracic kidney causing flank pain has been reported (Hamptom and Borden 2002).

A case of intrathoracic kidney with superimposed renal cell carcinoma has been reported; the patient first presented with hyponatremia and inappropriate antidiuretic hormone secretion (Kubricht et al. 1999). It is usually accompanied by a defect in the hemidiaphragm. Right-sided Bochdalek hernia with dilated colon loops and right kidney within a right hemithorax has been reported (Karaoglanoglu et al. 2006). Rarely, an intrathoracic kidney possesses congenital anomalies such as ureteropelvic junction obstruction, malpositioning, and duplication of the renal pelvis and ureters (Hamptom and Borden 2002). The combination of a duplex system with an intrathoracic renal ectopia is very rare (Beltrán Armada et al. 2004). Association of bilateral intrathoracic kidneys with bilateral diaphragmatic hernias has been reported (Dingeldein et al. 2008).

In the absence of other anomalies or abnormal renal function, an intrathoracic kidney does not interfere with the techniques used during coronary bypass surgery in cases with more than one renal artery. In various studies, a possible association between multiple renal arteries and hypertension has been reported (Glodny et al. 2001; Kem et al. 2005).

An intrathoracic kidney associated with bowel in the chest should undergo standard repair and nephropexy. An isolated intrathoracic kidney without evidence of bowel herniation can safely be ignored (Murphy et al. 2012); in the absence of other renal pathology it requires no surgical intervention (Subramanian and Goldfarb 2008; Murphy et al. 2012). Congenital intrathoracic kidney requires neither medical nor surgical treatment; in cases of posttraumatic diaphragmatic hernia it must be promptly treated surgically (Rouanne et al. 2010),

reducing the intrathoracic kidney into the abdomen (Nouri-Merchaoui et al. 2011).

Ectopic kidneys other than HSK

Cross-fused ectopic kidney is a congenital displacement of one kidney to the opposite side of the body, and is associated with an increased incidence of congenital and acquired anomalies (Collura et al. 2004; Chung et al. 2009). The causes of a crossed ectopic kidney are unclear; the autopsy incidence is one in 22,000. Crossed renal ectopia usually occurs from left to right, is found more often (3:2) in males than in females (Stimac et al. 2004; Kwon et al. 2004), and can be of various types such as unilateral fused kidney with inferior ectopia, sigmoid or S-shaped kidney, lump or cake-shaped kidney, L-shaped or tandem kidney, disk, shield, or doughnut-shaped kidneys, and unilateral fused kidneys with superior ectopia (Stuart et al. 1992). The ectopic kidney usually functions well unless malrotation affects its excretory capacity. In crossed renal ectopia the kidney is located contralateral to the side where the ureter enters the bladder, usually below the orthotopic organ, and in 90% of cases is fused to the normal kidney (Sood et al. 2005). In this position the wolffian duct (and with it the ureteral bud) crosses over the midline and fuses with the contralateral nephrogenic cord (Cook and Stephens 1977a, b). Crossed renal ectopia, in which one kidney lies on the contralateral side but its ureter passes to the ipsilateral side, is the second most common renal fusion anomaly following horseshoe kidney (Kwon et al. 2004; Boyan et al. 2007).

L-shaped kidney

L-shaped (tandem) crossed renal ectopia is a rare congenital anomaly. It occurs when the crossed kidney assumes a transverse position at the time of its attachment to the inferior pole of the normal kidney. The crossed kidney lies in the midline or in the contralateral paramedian space anterior to the L4 vertebra. Rotation about the long axis of the kidney can produce either an inverted or a reversed pelvic position. The ureter from each kidney enters the bladder on its respective side. Abdominal pain is a major symptom and staghorn calculi in both renal moieties have been reported (Chung et al. 2009; Mishra et al. 2013). Associations of an inverted L-shaped renal ectopia with inverted-Y ureteral duplication, ectopic ureter, and bicornuate uteruses have also been reported (Liu et al. 2010).

Disk kidney

Disk, shield, doughnut, or pancake kidneys are unusual anatomical anomalies that result when both the upper and lower poles of the embryonic kidney become fused at their medial borders to produce a doughnut- or ring-shaped mass. The renal parenchymal mass is located in the pelvis, generally with two pelves and two ureters and with no intervening fibrous septum. The lateral aspect of each kidney retains its normal contour (Heidempergher et al. 2012). This type of fusion differs from the lump or cake kidney in that the reniform shape is better preserved due to the somewhat less extensive degree of fusion. The pelves are anteriorly placed and the ureters remain uncrossed. Renal rhabdomyosarcoma (RMS) is a rare pediatric tumor that was discovered in a 4-year-old boy with metastases to the lung, pelvis, and bone marrow (Walther et al. 2013). This anomaly creates a great technical challenge during aortic reconstruction (Eze et al. 1998).

Lump kidney

A pelvic cake kidney in which there is complete fusion of both kidneys into a single renal parenchymal mass has been described as the rarest of renal fusion anomalies. It can be diagnosed at any age group from childhood to the eighth decade of life and is not necessarily associated with a poor prognosis (Rosenkranz et al. 2010; Schwartz et al. 2010). However, complications potentially associated with anatomical malformations such as urinary stasis, infection, formation of stones, and vascular involvement can cause serious clinical problems. Cake kidneys probably form because the nephrogenic blastemas are compressed between the umbilical arteries at the beginning of the cranial migration of the ureteral buds, and this could lead to their fusion. There has been extensive joining over a wide margin of the maturing renal anlage. The total kidney mass is irregular and lobulated. Ascent usually progresses only as far as the sacral promontory, but in many instances the kidney remains within the true pelvis. Both renal pelves are anterior and they drain separate areas of the parenchyma (Rosenkranz et al. 2010; Schwartz et al. 2010). The ureters do not cross. Most diagnosed cases have been reported to present malformations in other organs or in their blood supply, including abnormal testicular migration, Fallot's tetralogy, vaginal or sacral agenesia, and anal abnormalities. A pelvic cake kidney most commonly drains via two separate ureters; fewer than 10 reports demonstrate a cake kidney drained by a single ureter. In such instances other concomitant abnormalities such as unicornuate uterus or bilateral absence of the vas deferens has been reported (Goren and Eidelman 1987; Martinez-Lazaro and Cortes-Blanco 2000; Calado et al. 2004; Rosenkranz et al. 2010; Schwartz et al. 2010).

Symptoms

Most cases of renal ectopia remain asymptomatic during life and are found incidentally or detected only when complications arise. If manifestations do occur, they usually develop during the third or fourth decade of life and include vague lower abdominal symptoms such as urinary infections, urolithiasis, and abdominal mass (Kwon et al. 2004; Boyan et al. 2007; Ghosh et al. 2008). In different autopsy series it is detected in 1 in 1000–7000 (Hwang et al. 2002), and the number of cases found clinically is estimated to be only 1 in 10,000 patients (Bhatnagar et al. 2004; Guarino et al. 2004). In 0.2% of 12,000 patients investigated for the cause of abdominal pain, renal ectopia was found (Asghar and Wazir 2008), a condition of "pancake kidney"

eventually associated with polycystic disease and abnormal vascular supply. Hypertension and microscopic hematuria were the only clinical signs.

Associated anomalies

There have been reports of uncertain anatomy with multiple renal arteries and veins. Surgeons should be aware of ectopic and fused kidneys to minimize perioperative complications because of this uncertain anatomy. The surgical procedure must be planned according to the anatomical variations of each patient, and the anatomy of the patient's urinary system should be assessed with CT, IVP, and angiography. Crossed ectopia is always combined with abnormally located ureters, which cross the midline at the level of the distal aorta or its bifurcation to enter the bladder at the normal position (Yano et al. 2003; Guarino et al. 2004; Sood et al. 2005). Demonstration of crossed renal ectopia is important because it is a predisposing factor for obstruction, infection, and neoplasia of the urinary system (Boyan et al. 2007). Patients with fusion anomalies have a significantly higher risk of developing certain renal and urothelial tumors possibly related to embryogenic mechanisms, urinary stasis, and infection (Miller and Kropp 1992; Stimac et al. 2004). Rarely, crossed renal ectopia is associated with herniation of the ureter into an inguinal hernia (Rocklin et al. 1989; Hwang et al. 2002). It is usually associated with congenital anomalies of the gut. Development of malignancy in crossed fused ectopic kidney is uncommon. Many other associated syndromes and anomalies have been reported such as Klippel–Feil syndrome, various urogenital anomalies, imperforate anus, TAR syndrome, or cardiovascular septal defects. In cases of familial incidence, autosomal dominant inheritance has been reported (Rinat et al. 2001). There are reported cases of renal cell carcinoma, squamous cell carcinoma of renal pelvis origin, Wilm's tumor, and adenocarcinoma associated with crossed fused renal ectopia (Cook and Stephens 1977a, b; Redman and Beryl 1997; Aquilera Tubet et al. 2005). A lump in the lower abdomen and painless hematuria are signs of malignant involvement of an ectopic kidney (Soni et al. 2012). Associations with the absence of the vas deferens, cryptorchidism, and skeletal abnormalities have been reported. Caudal regression syndrome consists of multiple congenital anomalies, including imperforated anus, rectovesical fistula, sacral agenesis, vertebral anomalies, and club feet. Crossed fused renal ectopia can be associated with fused ureters and urinary obstruction (Duh et al. 2007). Polycystic changes in a pancake kidney have been reported (Heidempergher et al. 2012).

Diagnosis

It has been suggested that crossover and fusion with the opposite kidney could be caused by over-bending and rotation of the caudal end of the embryo. The increased prevalence of crossed renal ectopia in patients with scoliosis supports this hypothesis (Bauer et al. 1992). In most cases the fusion is between the lower pole of the orthotopic kidney and the upper pole of the ectopic kidney. Association with mullerian agenesis has also been reported with a 40% incidence of renal anomalies (Sen and Kapoor 2006). Genitourinary anomalies can present a formidable challenge to the vascular surgeon during abdominal aortic reconstruction. Because of the risk for injury to the kidney during surgery, preoperative evaluation of this anomaly must include computed tomography, angiography, and intravenous pyelography. Preoperative placement of a ureteral catheter can also prevent injury to the anomalous ureter (Yano et al. 2003). Pancake kidney has been associated with bilateral iliac artery aneurysms (Eckes and Lawrence 1997).

Crossed renal ectopia can usually be diagnosed by intravenous urography. Other methods include ultrasonography, intravenous pyelography, computerized tomography, and andrenal scintigraphy (Sharma et al. 2006; Boyan et al. 2007). CT also helps to identify metastatic disease. MRI is another modality for characterizing a focal renal mass. RCC is typically mildly hypointense on T1-weighted and hyperintense on T2-weighted images. Association of squamous cell carcinoma or RCC with crossed fused renal ectopia is relatively rare. CT or MR angiography is a required preoperative study for imaging the supplying vessels; if there are malignant mass lesions a heminephrectomy can be performed (Kraft et al. 2007; Kumar et al. 2008; Liu et al. 2010). Renal cortical scintigraphy is useful for locating the poorly functioning kidney for surgical removal (Gharagozloo and Lebowitz 1995). Diagnosis of renal ectopia can be an accidental finding during scintigraphy performed for other reasons (Cohn and Roach 2003).

Treatment

Treatment of crossed-fused renal ectopia is indicated when complications rather than the anomaly itself require intervention (Ghosh et al. 2008; Liu et al. 2010). Most individuals with crossed renal ectopia have normal longevity and good prognosis. However, some patients are at risk of developing urinary tract infections or renal calculi; Boatman et al. (1972) noted that one-third of their symptomatic patients required a pyelolithotomy. Extracorporeal shock wave lithotripsy therapy and percutaneous nephrolithotomy have rendered most of these unusual patients stone-free (Semerci et al. 1997; Desai and Jasani 2000). If investigations demonstrate an obstruction, surgery to prevent complications can be required (Taslim et al. 2012).

Abnormalities of rotation

Rotational anomalies of kidney are rare (Das and Amar 1984; Nathan and Glezer 1984). During normal development the kidney ascends from its original pelvic position to its final location at the end of the eighth week. It also undergoes rotation so that the hilum, which initially faced its ventral aspect, comes to lie on its medial aspect. This occurs during the ascent of the kidney, between 38 and 49 days of development. When renal rotation takes place before definitive vascularization (Pollack

and McClennan 2000) the hilum is directed anteriorly rather than medially. The true incidence of this condition is underestimated because in many patients it is asympotomatic. Different types of rotation abnormalities have been reported: in unrotated kidneys the renal pelvises face ventrally; in incomplete rotation they face venteromedially; in some cases there is excessive rotation. In the rare reverse and excessive rotation cases, the renal pelvis presents itself in a position depending upon the number of degrees of rotation completed (Das and Amar 1984; Atasever et al. 1992; Pollack and McClennan 2000). Bilateral non-rotation of the kidneys has been reported in an 82-year-old female (Moore et al. 2011).

Renal arteries develop from the lateral splanchnic arteries, which form a network on either side of the aorta. Accessory renal vessels arise as a result of the complicated development and positional anatomy of the kidneys (Bayramoglu et al. 2003). Anomalies associated with rotation of the kidneys, either unilateral or bilateral, are often caused by or related to aberrant vessels and often receive an accessory arterial branch. The accessory arteries usually enter the renal cortex at one of the poles. Accessory renal arteries most commonly pass anterior to the ureter; they are sometimes encountered in patients with hydronephrosis (Malament et al. 1961; Hollinshead 1971; Nathan and Glezer 1984). The accessory vessels can also be located posterior to the ureter, closely related to the ureteropelvic junction (Bayramoglu et al. 2003), creating a clinically important condition.

Although rare, rotational anomalies have important surgical implications because they can be mistaken for a paravertebral mass, and the anterolateral deviation of the urinary system sometimes creates an impression of a lower pole mass. Abnormal rotation creates problems during percutaneous nephrectomy. Anomalies of renal rotation are associated with renal ectopia and fusional abnormalities but can be exhibited with no additional abnormality (Braasch 1931). The possible presence of an accessory renal artery should be considered in abnormally rotated kidney cases (Braasch 1931; Olsson and Wholey 1964). A rare anomaly of the kidneys and its vessels was found in a white adult male cadaver. The anomaly consisted of unrotated kidneys with partially extrarenal calices and pelves. In addition to the normal vessels, each kidney received a branch from a common trunk, originating from the inferior end of the aorta (Nathan and Glezer 1984).

Nephroptosis

Nephroptosis has been defined as renal descent of 5 cm or more (or two vertebral bodies) on changing from the recumbent position to orthostasis. Noted as a common finding, it rarely produces symptoms. It is said to be more common in women and to affect the right kidney more often than the left; it is bilateral in 20% of cases. It has been observed predominantly in slim young women with little supportive perirenal fat. The classical history is of flank pain in the upright position that reduces or is relieved by lying down. It can be associated with intermittent functional excretory obstruction, with forceful traction on the renal artery leading to renal ischemia and with traction on the perirenal nerves. Historically a number of symptoms have at one time or another been linked to this finding. However, it is nephroptotic pain that dominates the picture, although recurrent upper tract urosepsis, hypertension, and renal stone disease have also been linked. Nowadays, a diagnosis based on symptoms can be confirmed by contrast imaging and/or renography, which demonstrate the classical renal descent with significant rotation and decrease in renal blood flow on moving from the supine to the erect state, respectively (Tatevosian et al. 2004). Erect and supine IV urograms or renal scans documenting obstruction are practical diagnostic tests for the condition (see *Campbell's Urology*). New criteria for diagnosing symptomatic nephroptosis and the introduction of imaging and laparoscopic surgical management have replaced the more exotic and unnecessary surgical procedures employed in the past (Moss 1997; Hatzinger et al. 2007).

History

The idea of renal 'ptosis' was described by Matthew Baillie in 1825. Shortly afterwards in 1939, Rayer concluded that this condition was far more common than formerly appreciated; it was frequently overlooked and the patient was often wrongly diagnosed with nervous colic, hypochondria, or lumbosciatic neuralgia. The first nephropexy procedure was performed in 1881 (McWhinnie and Hamilton 1984; O'Reilly and Pollard 1988; Moss 1997; Boccardo 1999). Renal ptosis has a contentious and colorful history. Recently trained physicians are often unfamiliar with it. Over the centuries the condition has received numerous labels including: the moveable kidney, the floating kidney, renmobilis renmigrans, and Wanderniere. Nephropexy, once a routine operation, almost disappeared from American practice more than 40 years ago. Nearly 200 surgical procedures described for this condition were suddenly eclipsed. The label 'an ineffective treatment for an imaginary disease' then stuck to the condition of nephroptosis and its surgical management (Barber and Thompson 2004; Hatzinger et al. 2007).

Floating kidney

Floating kidney and nephroptosis are exactly the same entity; the former term seems to have referred to a self-reducible ptosis. Pierre Rayer discussed the issue extensively in his great monograph on the kidney (published in 1839), noting that the right kidney is more prone to floating than the left. Rayer also recounted the sad tale of a doctor who felt his own kidney floating while taking a bath, and promptly retired from practice to prepare himself for death (Bynum 2001). The first nephropexy was performed in 1881. From that time, surgical treatment of nephroptosis has remained a subject of discussion. By 1936, approximately 170 different surgical methods existed for fixation of the kidney. An accurate diagnosis is imperative before

performing nephropexy. There were times when this operation was carried out much too often, giving it a bad reputation (Moss 1997; Hatzinger et al. 2011). In the past the diagnosis has probably been frequently mistaken, but we should caution against completely rejecting it; it can happen in some special cases. By definition of the 'floating state', the kidneys move at least two vertebral bodies downward in the vertical position, and this movement can be detected by radiography. Also, retrograde pyelography revealed a delay in emptying the renal pelvis in one patient with suspected floating right kidney. Although diagnosis has frequently been wrong in the past and many unnecessary operations have been performed, present imaging techniques make it possible to diagnose the occasional genuine cases of nephroptosis (Newling 2001).

Primary megaureter

Megaureter was first described by Caulk in 1923. Congenital megaureter is a condition usually diagnosed in neonates and children; it is hypothesized to result from failure of the ureterovesical junction (UVJ) to mature or from ureteral wall muscle-fiber abnormalities (Hanna and Wyatt 1975; Gosling and Dixon 1978). Diagnostic criteria include: dilated ureter, absence of vesicoureteral reflux, absence of infravesical obstruction, and absence of distal ureteral obstruction (Domini et al. 1999; Khoury and Bagli 2007). Primary non-refluxing megaureter is usually a functional and benign congenital malformation that resolves spontaneously. Only a few cases require surgical treatment; this should be reserved for patients who develop ureteral dilation, a decrease in renal function, and/or severe symptoms during follow-up (Areses Trapote et al. 2007). A primary megaureter is an anomaly with a pre-vesical or overall dilated ureter with a diameter of more than 6 mm.

It is important to distinguish between cases of primary non-refluxing and primary obstructive-refluxing megaureters as they require completely different treatments. The basic diagnostic work-up includes ultrasonography and voiding cystourethrography. Diuretic renography is used to detect the degree of upper urinary tract obstruction. In contrast to most cases of primary non-refluxing megaureter, obstructive refluxing megaureters commonly need to be corrected. Antibiotic prophylaxis can be indicated in infants with a primary obstructive megaureter during the first six months of life because there is a higher risk of complications due to pyelonephritis (Anheuser et al. 2013). Most cases of primary megaureter (PM) resolve spontaneously, but long-term follow-up is recommended because symptoms can develop years later (Di Renzo et al. 2013). A high rate of spontaneous resolution of or decrease in urinary tract dilatation is expected for most cases. Long-term follow-up of children with prenatally diagnosed PM with mild to moderate hydronephrosis confirms a high incidence of resolution and improvement. Ultrasonography should be continued periodically until the child reaches adulthood (Shukla et al. 2005). In adults and adolescents with primary megaureter, large stones can develop in the dilated portion of the ureter due to urinary stasis (Delakas et al. 2002; Hemal et al. 2003; Rosenblatt et al. 2009). Cases of megaureter because of uretropelvic junction obstruction can present with acute flank pain crisis during childhood (Hanna et al. 1976; Anderson et al. 2012). Congenital megaureter is a diagnosis that urologists and radiologists need to consider in the setting of isolated distal ureteral dilation. Although its presentation in adult life is rare it is sometimes identified by point-of-care ultrasound DC, usually during the third or fourth decade, and it is usually unilateral (most often left-sided) (Dorairajan et al. 1975).

Supernumerary kidney

Supernumerary kidney is a rare congenital urinary tract abnormality. The supernumerary kidney is a definitive accessory organ with its own collecting system, blood supply, and distinct encapsulated parenchyma. It is rarely symptomatic but can become symptomatic in early adulthood. It is among the rarest congenital urological anomalies with fewer than 100 cases reported in the English literature. The embryological anomaly results from premature division of the metanephric bud; the number of kidneys probably reflects the number of abnormal divisions of the progenitor cells. It affects males and females equally and usually affects the left side. Bilateral supernumerary kidney is even rarer and only three cases have been reported in the literature. Computed tomography (CT), excretory urography (IVP), and CT angiography (CTA) can be used to confirm the diagnosis of supernumerary kidney (Oto et al. 2002).

Because of the atypical presenting symptomatology, this entity frequently causes a diagnostic as well as a therapeutic dilemma. Back pain has been reported as the presenting sign (Tada et al. 1981). A noncontrast CT scan revealed a left-sided para-aortic mass. An intravenous contrast CT scan revealed an anatomically and functionally supernumerary kidney and no percutaneous biopsy was required (Bernik et al. 2001). The condition is generally discovered when complications arise, for example hydronephrosis (Jira et al. 2002). An acute episode of abdominopelvic pain, secondary to hydronephrosis of a third kidney situated in the pelvis behind the bladder, has been reported (Eberle et al. 2002). A 33-year-old female patient was investigated for right lower quadrant pain. She was treated surgically for a cyst of the right ovary. Finally, excretory urography and a computed tomography examination revealed a normally functioning extra kidney on that side (Koureas et al. 2000). An ectopic supernumerary pelvic kidney is a rare cause of a pelvic mass that can be found incidentally on a computed axial tomographic (CT) examination (Flyer et al. 1994), but most cases are asymptomatic and only an imaging study reveals separate extra kidneys.

Solitary functioning kidney (SFK)

SFK in childhood can be either of primary (congenital) origin (pSFK) or secondary after unilateral nephrectomy (sSFK). These are different anomalies. Unilateral renal agenesis (URA) is defined as congenital absence or a kidney on one side. In a systematic review of 43 cohorts comprising over 2600 patients with URA, the general incidence of URA was 1 in 2000. Because it is difficult to diagnose, the pre-renal incidence of URA is reported around 1 in 8000 (Westland et al. 2013b, 2014). It is suggested that most cases diagnosed as URA are actually cases of renal aplasia, which is not easily detectable on ultrasound (Westland et al. 2013b, 2014). Both agenesis and aplasia are referred to as URA. It should be discriminated from a unilateral nonfunctioning kidney resulting from multicystic renal dysplasia (MCRD); in the latter condition the renal parenchyma exhibits dysplastic and cystic differentiation, with a normally atretic ureter due to abnormal fetal renal development (Schedl 2007; Weber 2012). Its incidence is 1 in 4300 births (Westland et al. 2013b, 2014). Fetal ultrasound now enables individuals with a solitary functioning kidney to be identified prenatally. Congenital solitary kidney affects 1 in 1000 persons, whereas bilateral renal agenesis affects 1 in 5000 and is incompatible with life (McPherson 2007); however, in a meta-analyses of 43 studies (total number of patients 2684, 63% male) the general incidence of URA was 1 in approximately 2000 (Westland et al. 2013b).

Acquired solitary kidneys are frequently due to nephrectomy of a pathological contralateral kidney or to donation of a kidney for renal transplantation. Having a solitary functioning kidney (SFK) entails having an abnormally low number of nephrons, and this leads to adaptive hypertrophy and hyperfiltration in the remaining nephrons with proteinuria, arterial hypertension (AH), and diminished glomerular filtration rate (GFR) as possible sequelae (Gonzalez et al. 2005).

Although hyperfiltration secondary to a 50% reduction of renal mass in humans is considered not to lead to loss of function of the remaining parenchyma, there is an increased risk for proteinuria and progressive renal failure (Fotino 1989; Novick et al. 1991; Gluhovschi et al. 2013). However, other researchers believe that renal function in children with solitary kidney declines gradually over longer periods of follow-up (Abou Jaoudé et al. 2011; Westland et al. 2013a). A recent study demonstrated that 40–50% of adults with URA required dialysis by the age of 30 (Sanna-Cherchi et al. 2009; Westland et al. 2013b). Early presentation can be occult blood in the urine (Anraku et al. 2012). Hydronephrosis combined with ureteral diverticulum has been reported in the solitary kidney because of the blind-ended bifid ureter (Anraku et al. 2012). Offspring and other relatives of individuals with congenital solitary kidney have a significantly increased incidence of renal disease (agenesis, dysplasia, cystic dysplasia) (McPherson 2007). A retrospective study of renal injury markers in children with SFK demonstrated that nearly one-third of the patients had hypertension or albuminuria or used renoprotective medication at a young age (Kaneyama et al. 2004; Dursun et al. 2005). We therefore emphasize that clinical follow-up of all children with an SFK is needed. URA is also associated with extra-renal involvement of systems such as gastrointestinal, cardiovascular, and musculoskeletal (Westland et al. 2013b).

A dromedary hump is a common variant of the left kidney and represents the splenic impression from pressure of the splenic border along the upper lateral half of the renal contour.

Urinary bladder and ureters

Urinary bladders of variable shape (e.g., hourglass bladder) have been reported. The bladder may be doubled. Agenesis has a reported incidence of 1:600,000 (0.00017%).

Variations of the calices, renal pelvis, and ureter include the following.

1. Calices, including absent, aberrant, or unusual number, size or position.
2. Renal pelvis, including absent, rudimentary, double (bifid), multiple (unilateral or bilateral), hydronephrosis (congenital), intrarenal pelvis (unilateral or bilateral), extrarenal pelvis (unilateral or bilateral), or diverticulum of renal pelvis.
3. Ureter, including (a) variations in number, for example single (one ureter absent), double on one side, triple, quadruple, quintuple, or sextuple; (b) complete, incomplete, or branching; or (c) variations in form, size, and position, for example diverticulum of ureter (simple or multiple), inperforate ureter, stricture of ureter (pyo-ureter, hydroureter, megalo-ureter), dilation of ureter (total or partial), "golf hole" type of ureteral orifice, ureter ectopic, misplaced in bladder (at fundus, in diverticulum, near bladder neck, etc.), with extravesical orifice (opening into another ureter, urethra, genital tract, or bowel), ureter twisted or occluded, ureters crossed, or ureter with valve formation.

The calices majores may pass downward for some distance beyond the hilum, and end by joining to form the ureter without undergoing any obvious expansion. In such cases, the pelvis is absent; if the calices dilate, one or two pelves may be present.

The principal variation in the ureter is a more or less complete division into two. As a rule, the two ureters unite a little above the bladder, so that there is only one vesical orifice. When the division is complete there are two separate openings into the bladder. In one study of 26,480 autopsies, 136 double or bifid ureters were found (0.5%).

In rare cases, three or four ureters may be found. Several instances are recorded in which a supernumerary ureter, proceeding from the upper part of the kidney, opened directly into the urethra. Ureters have also been reported to open into the vagina, the seminal vesicle, or the prostatic urethra. The calices minores may vary in number over the range 6–14, the smaller number indicating that some of the calices minores embrace the

apices of several pyramids. Variations in the calices minores and pelvis are more marked. The right ureter has been found passing behind the inferior vena cava (postcaval or retrocaval ureter) between that vessel and the aorta.

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