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PICTORIAL ESSAY

Congenital anatomic variants of the kidney and ureter: a pictorial essay

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Abstract Congenital renal parenchymal and pelvicalyceal abnormalities have a wide spectrum. Most of them are asymptomatic, like that of ectopia, cross fused kidney, horseshoe kidney, etc., while a few of them become complicated, leading to renal failure and death. It is very important for the radiologist to identify these anatomic variants and guide the clinicians for surgical and therapeutic procedures. Cross-sectional imaging with a volume rendered technique/maximum intensity projection has overcome ultrasonography and IVU for identification and interpretation of some of these variants.

Keywords Horseshoe kidney · Renal agenesis · Zinner's syndrome · Pancake kidney · Intrathoracic ectopia

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Introduction

Congenital anomalies of the kidney and urinary tract occur in 3.3–11.1 % of populations and they account for about 50 % of all congenital abnormalities [1]. Renal tract malformations are a clinically challenging collection of entities because of their diversity and the fact that these disorders can present both before and after birth [1]. Imaging plays an important role in early diagnosis and proper management. Ultrasonography (USG) is a non-invasive modality and has a major impact on evaluating renal anomalies either prenatally or postnatally. However, it may be difficult to diagnose a small damaged kidney or dysplastic kidney with USG [1]. Multidetector computed tomographic (MDCT) urography has the ability to depict the normal urinary tract anatomy, including both the renal parenchyma, collecting structures and ureters. MDCT urography is helpful to diagnose the presence of stones, hydronephrosis or masses. The post-processing images of the MDCT-like maximum intensity projection (MIP), volume rendered technique (VRT) and multiplanar reconstruction (MPR) techniques provide a sensation of three-dimensionality and allow for a more accurate diagnosis [2]. Although MDCT has taken the largest leap, use of iodinated contrast material and radiation exposure is a concern with MDCT. Magnetic resonance imaging (MRI) can be used in case of compromised renal function, severe contrast allergy or in case radiation exposure is a problem, such as in children and pregnant women [3]. MRI and MDCT show comparable accuracy in detection and characterization of most renal lesions [3]. Sometimes typical presentations of common entities and certain uncommon entities may simulate renal neoplasms and may lead to an unnecessary resection because of the concern for renal malignancy; hence, there is a need to know the congenital causes of renal pseudo-tumors [4].

The present article is aimed at documenting various imaging findings in the congenital anatomic variants of kidney and ureters and their complications. The article stresses the need for a multimodality approach to arrive at a correct diagnosis.

Abnormalities of number

Renal agenesis

True renal agenesis results from a failed embryonic process in which the ureteric bud contacts the metanephric blastema to stimulate the development of the normal kidney. Unilateral or bilateral renal agenesis is reported to be 1 in 1,000 and 1 in 10,000 births, respectively [5]. Renal agenesis is commonly associated with other Müllerian (mesonephric) and Wolffian (paramesonephric) ductal anomalies (Fig. 1). Ipsilateral adrenals can be absent or low-lying [2, 4]. Syndromes associated include Zinner's syndrome, Herlyn–Werner–Wunderlich syndrome, Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome and Kallaman syndrome [5, 6].

Bilateral renal agenesis is a rare and fatal event, usually associated with severe oligohydramnios, which produces a characteristic clinical pattern with facial compression and pulmonary hypoplasia (Potter sequence) [6].

Herlyn–Werner-Wunderlich syndrome Herlyn–Werner–Wunderlich syndrome (Fig. 2), also known as obstructed hemivagina and ipsilateral renal anomaly, is a very rare congenital anomaly of the urogenital tract involving Müllerian ducts and Wolffian structures and it is characterized by the triad of didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis [7]. Clinically, this usually presents in post pubertal adolescents or adult women where hematometrocolpos produces a more pronounced mass effect and pain on the side of the obstructed hemivagina [7]. Moreover, the diagnosis is complicated by the infrequency of this syndrome, because Müllerian duct anomalies (MDA) are infrequently encountered in a routine clinical setting. When a high suspicion of MDA exists, USG should be performed initially to look for any abnormalities in the genital tract; however, in order to obtain an accurate MDA classification, MRI must be performed [7].

Zinner's syndrome Zinner's syndrome is a triad of Müllerian duct abnormality comprising of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction. The association was first described by Zinner in 1914 and up till 2000, about 100 cases had been reported. Zinner's syndrome is also considered to be the male counterpart of MRKH syndrome (uterovaginal aplasia) seen in females. Patients are usually diagnosed at adulthood and often present with infertility [8] (Fig. 3). Precise delineation of the renal anomalies and the altered pelvic anatomy is feasible with MDCT. The ideal imaging modality to evaluate the Müllerian duct abnormalities is MRI [8].

Supernumerary kidney

A supernumerary kidney is a third kidney in addition to the two independent kidneys [9]. A supernumerary kidney is a rare urogenital anomaly with less than 100 cases reported [9]. A fused supernumerary kidney is still rarer [9]. They occur due to abnormal cleavage of the metanephric blastema [8]. They are either separate or fused with the parent kidney. Most of the supernumerary kidneys have a separate collecting system. Associated urogenital anomalies are fusion, ectopic ureteric opening, vaginal and uretral atresia, urethral or penile duplication [9].

The supernumerary kidney needs to be differentiated from the more commonly occurring duplex kidney, which is defined as having two pelvicalyceal systems. The supernumerary kidney, in contrast, is thought to be an accessory

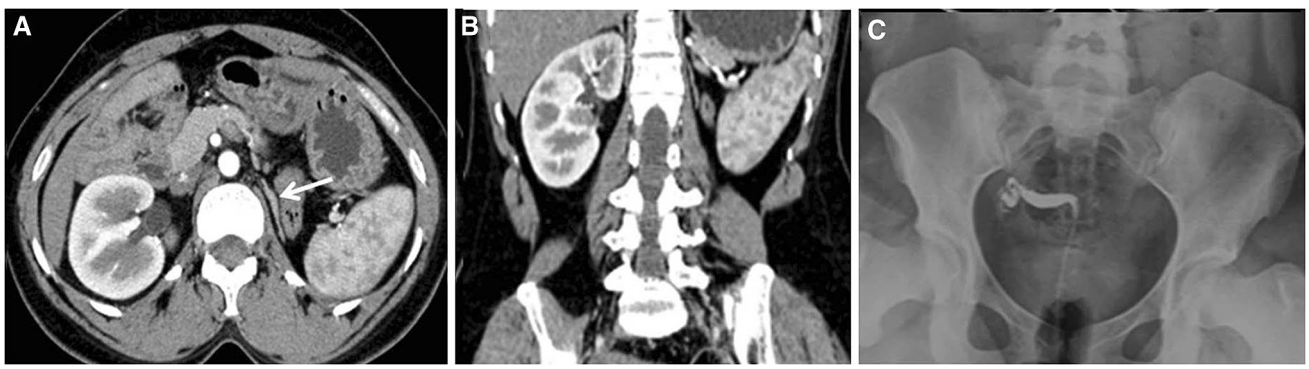

Fig. 1 Left renal agenesis with unicornuate uterus. Axial (**a**) and coronal reformatted CT images (**b**) showing unilateral left renal agenesis with low lying left adrenal gland (*white arrow*). Same patient had unicornuate uterus which is well identified on hystero-salphingography (**c**)

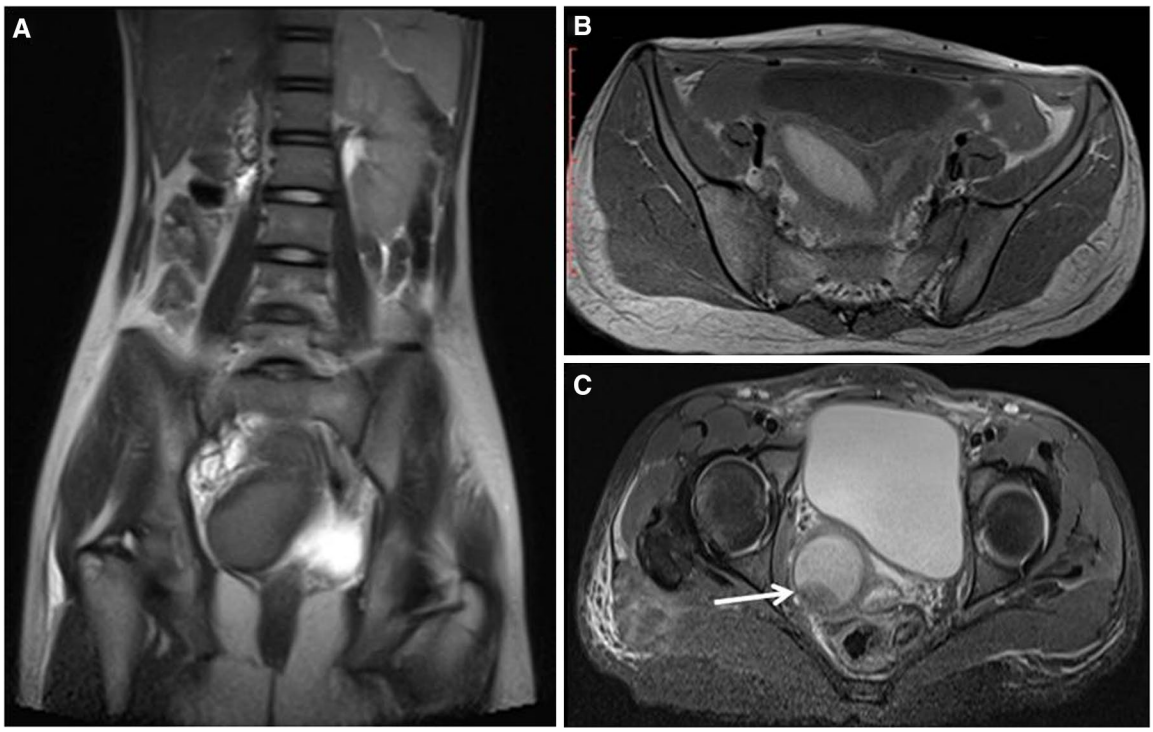

Fig. 2 MR images of Herlyn–Werner–Wunderlich syndrome. T2-weighted coronal image (**a**) shows absence of right renal tissue with uterine didelphys in T1-weighted axial image (**b**). T1-hyperintense contents noted within the right horn of the uterus hematometrocolpos. T2-weighted axial image (**c**) shows right-sided hemivaginal septum (*white arrow*)

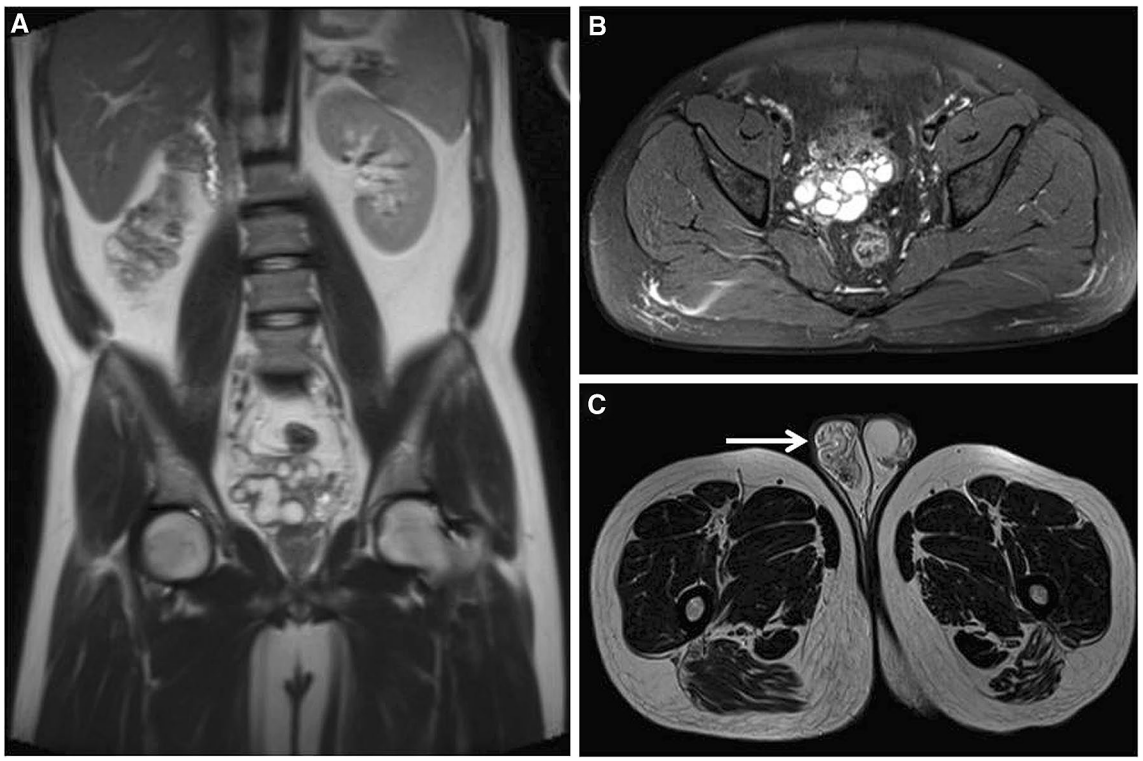

Fig. 3 MR images of Zinner's syndrome. T2-weighted coronal image (**a**) shows absence of the right kidney. Multiple cysts are seen in the right seminal vesicle (**b**). T2-weighted axial image (**c**) shows the right-sided tortuous vas deferens (*white arrow*) likely secondary to an obstructed ejaculatory duct

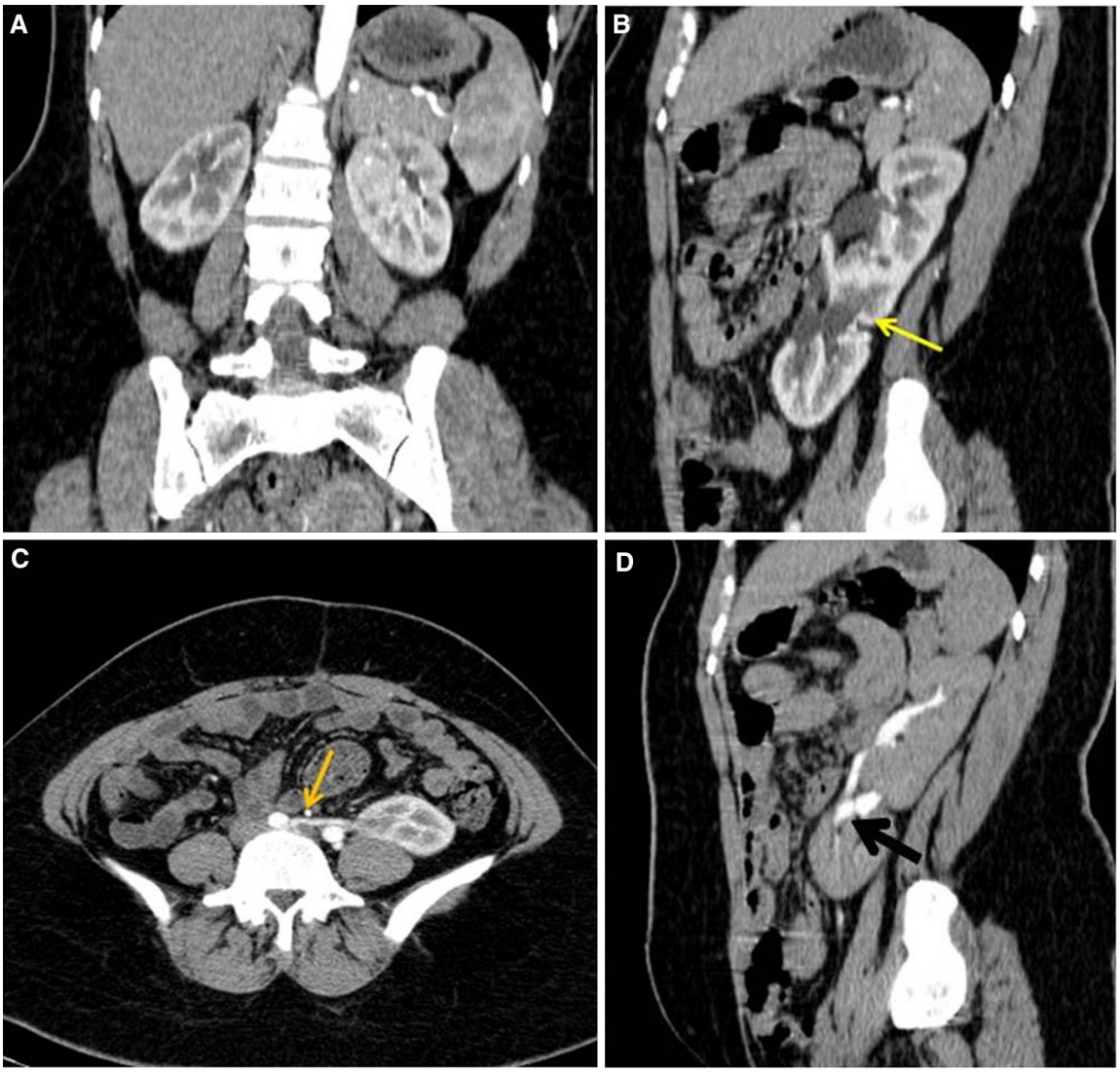

Fig. 4 Supernumerary kidney. Coronal reformatted computed tomography (CT) image (**a**) shows presence of both the kidneys in the renal fossa. Sagittal reformatted CT image (**b**) shows a third kidney, which is seen fused with the left kidney at its inferior pole (*yellow arrow*). A branch from the right iliac artery is seen supplying the supernumerary kidney (*yellow arrow*) in the axial post contrast image (**c**). Delayed sagittal reformatted CT image (**d**) shows the separate collecting system (*black arrow*) of a supernumerary kidney

organ with a separate arterial supply, venous drainage, collecting system, and distinct encapsulated tissue [9] (Fig. 4).

Abnormalities of shape

Horseshoe kidney

The horseshoe kidney (HSK) represents one of the most frequent renal malformations with an incidence of 0.25 % in the general population, being more frequent in men (2.3:1) than women [1]. It consists of the fusion of lower poles which are more medially located than the upper poles and anterior to the great vessels at the level of the third to fifth vertebra, joined by an isthmus that can be composed of renal parenchyma (Fig. 5) or fibrous tissue at the junction of the aorta and inferior mesenteric artery. The majority of HSKs lies in a lower position than normal kidneys and are supplied by renal arteries derived from the abdominal aorta below the isthmus or by vessels originating from the common iliac arteries [1]. USG is initial imaging modality for diagnosis of HSKs. MDCT and MRI are comprehensive imaging modalities to detect HSK and its complications. Renal scintigraphy using technetium-labelled radionuclides may be performed to assess for suspected ureteropelvic junction (UPJ) obstruction. HSKs are more prone for injury in blunt trauma, varying degrees of UPJ obstruction with resultant infection and stones [10].

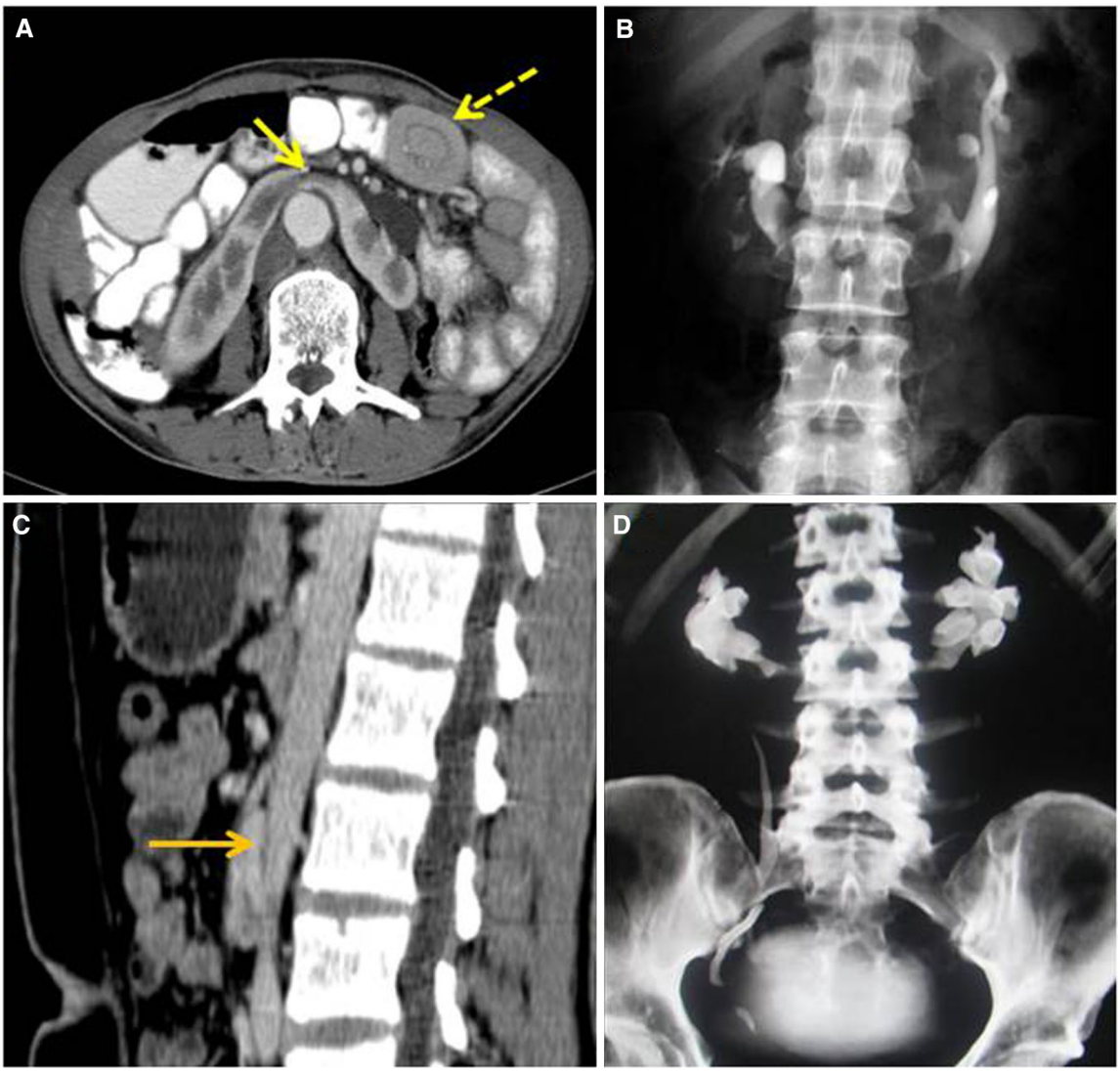

Fig. 5 Horseshoe kidney. CT (**a**) and intravenous urography (IVU; **B**) images show fusion of the lower ends of both the kidneys across the midline by a parenchymal isthmus (*yellow arrow*). Also note the transient intussusception (*dashed arrow*). A sagittal reformatted (**c**) image shows the isthmus at the junction of the inferior mesenteric artery and aorta (*orange arrow*). Post-contrast VRT (**d**) image demonstrates that the patient had a single ureter

Pancake kidney

Looney and Dodd were the first to describe pancake kidney (fused pelvic kidney). Pancake kidney is a rare fusion anomaly of the kidneys characterized by the presence of a displaced, lobulated pelvic renal mass of dual parenchymatous systems without an intervening septum. Each kidney has its own collecting system and anteriorly placed short ureters entering the bladder normally [11]. The presence of a pancake kidney may predispose a patient to recurrent urinary tract infections and stones, likely due to a rotation anomaly of the collecting system and short ureters which results in obstruction and stasis. The diagnosis is always incidental and most cases of pancake kidney are asymptomatic (Fig. 6). In earlier days, IVU was the usual method of detection, which is now replaced by USG, CT scan and radionucleotide scanning [11].

Abnormalities of site

Ectopic kidney is described as abnormal localization of a kidney due to a developmental anomaly and it occurs as a result of halt in migration of kidneys to their normal locations during the embryonal period. Abnormality of the ureteral bud or metanephric-blastema, genetic abnormalities or anomalous vasculature act as a barrier to ascension. Renal ectopia is divided into ipsilateral renal ectopia and crossed renal ectopia [5, 8].

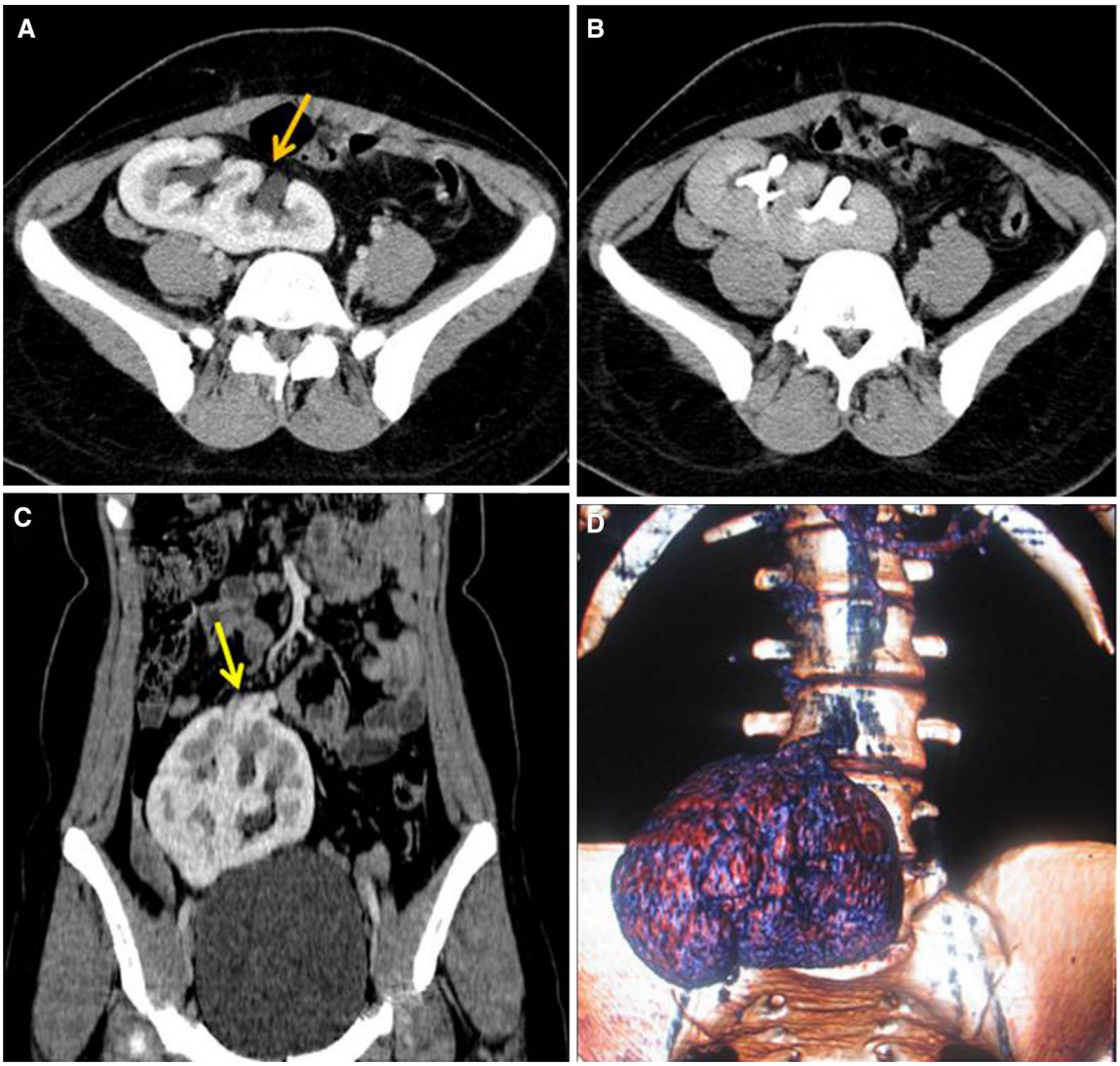

Fig. 6 Pancake kidney. Axial CT images (**a**, **b**) showing both the kidneys are low lying and fused. Coronal reformatted CT image (**c**) and VRT (**d**) image show both the kidneys are fused enface and low lying

Ipsilateral renal ectopia means that the kidney is on the same side of the body as the orifice of its attendant ureter. Crossed renal ectopia is defined as a kidney located on the opposite side from which its ureter inserts into the bladder. The ectopic kidney has clinical significance owing to its atypical location, malrotation and vascular particularities [12].

Ipsilateral renal ectopia

Ipsilateral renal ectopia is further divided into cranial and caudal ectopia according to whether it is above or below the normal position. Cranial renal ectopia is usually intrathoracic, and caudal renal ectopia is classified as abdominal and pelvic (sacral) type [6]. An abdominal kidney is located above the iliac crest but below the level of the L2 vertebral body. The pelvic kidney is most common (Fig. 7) and located below the iliac crest [5, 6]. Most ectopic kidneys are clinically asymptomatic, few of them are associated with complications like hydronephrosis, ureteropelvic junction or ureterovesical junction obstruction and reflux [6]. A pelvic kidney must be differentiated from nephroptosis, which is mobile and a low-lying kidney due to lack of supportive fascia [6].

Pelvic ectopia See Fig. 7.

Intrathoracic ectopia A thoracic kidney is an extremely rare renal ectopia that occurs due to ascension of the kidney above lumbar vertebra, with about 200 cases published [13]. It is common on the left [6]. Most are congenital and diagnosed by chance during chest radiography or at surgery. An acquired thoracic kidney is usually related to diaphrag-

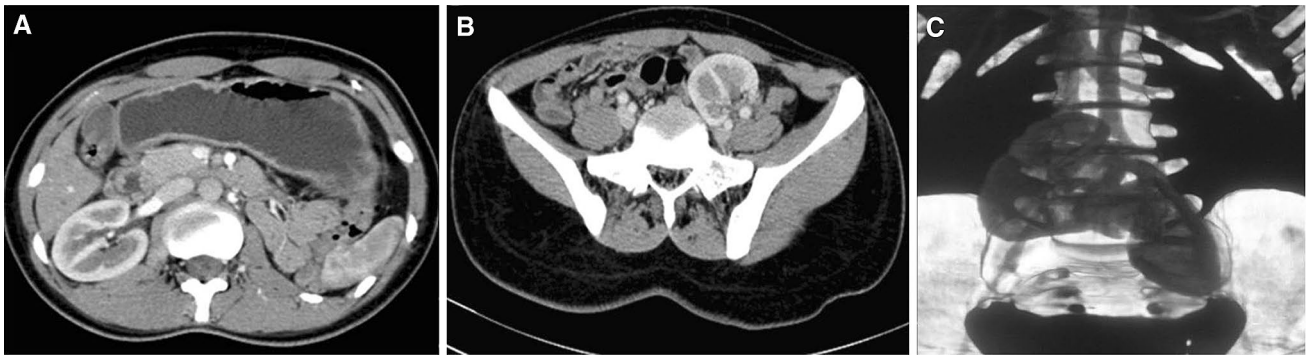

Fig. 7 Pelvic ectopic kidney. Post-contrast axial image (**a**) shows the left renal fossae is empty, the left kidney is low-lying in the abdomen (**b**) Coronal VRT image of a different patient (**c**) showing bilateral pelvic/sacral kidneys

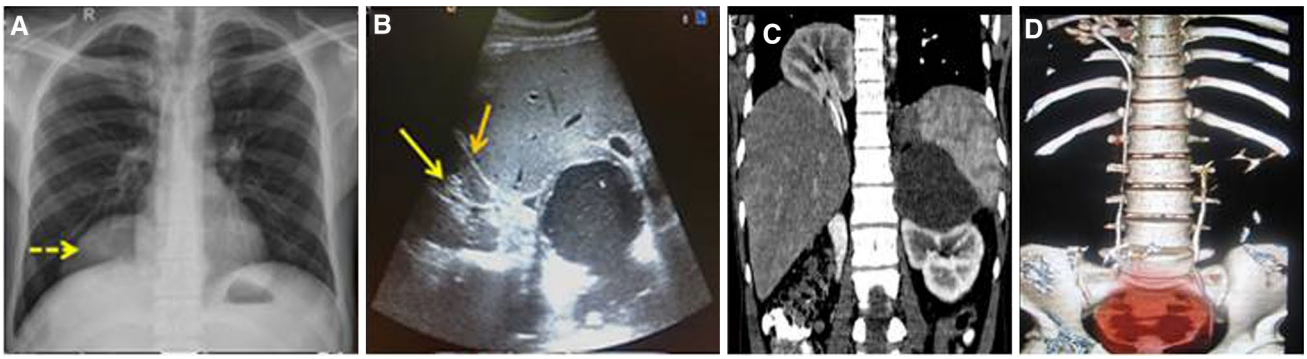

Fig. 8 Thoracic ectopic kidney. Chest radiograph PA view (**a**) shows a soft tissue density shadow (*dashed arrow*) in the right cardiophrenic region. A sagittal ultrasound image (**b**) shows the right kidney (*yellow arrow*) above the dome of the diaphragm; the patient also had pseudocysts of the pancreas. Coronal reformatted CT image (**c**) showing right thoracic ectopia where in the kidney is lying well above the liver. Coronal VRT image (**d**) shows the long course of the right ureter

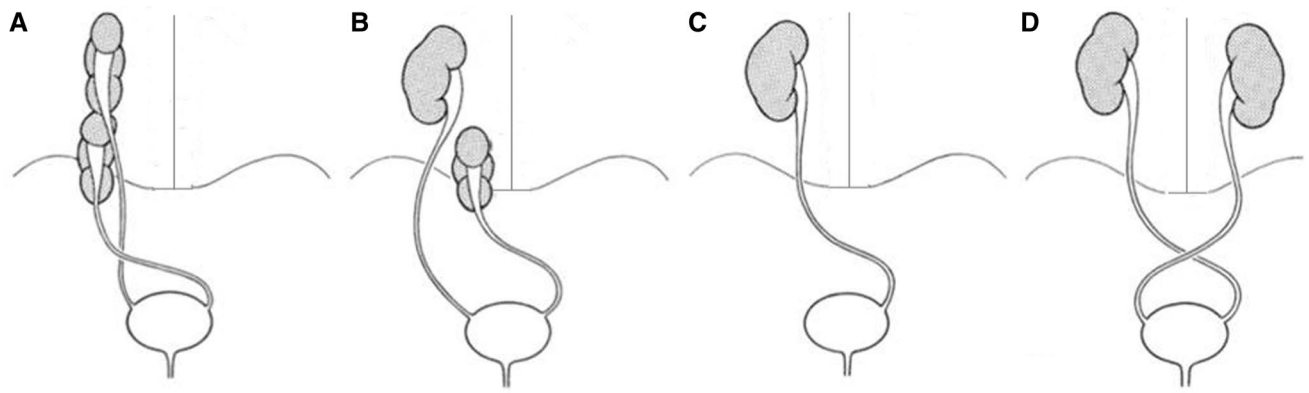

Fig. 9 Diagrammatic representation of four types of crossed renal ectopia. **a,** Type A: with fusion; **b,** Type B: without fusion; **c,** Type C: solitary crossed; **d,** Type D: bilaterally crossed (15)

matic rupture. A congenital thoracic kidney may be caused by accelerated cranial migration of the embryonic kidney or delayed diaphragm formation (13]. Blood supply to the intrathoracic kidney arises from a renal branch of the aorta in its usual location, rarely with an anomalous superior origin of the renal artery. A thoracic kidney is always accompanied by an elongated ureter, while the presence of other abnormalities is not constant (13] (Fig. 8).

Crossed fused renal ectopia

Crossed fused renal ectopia (CRE) is the second most common fusion abnormality of the urinary tract after horseshoe kidney with a male predominance of 3:2. It is due to abnormal development of the ureteric bud and metanephric blastema during the four to eight weeks of gestation. CRE is defined as a kidney located on the opposite side from which its ureter inserts into the bladder. The ureter from each kidney is usually orthotopic and the trigone of the bladder is normal. Associated anomalies involving the skeletal and genital systems are frequent in solitary CRE [6, 14]. Abdominal radiograph can suggest the CRE, if one renal shadow is not visualized and the opposite renal outline is enlarged or when stones occur in an unusual location. USG can determine the presence of two kidneys on the same side of the abdomen and also the absence of a kidney in the contralateral normal position [1]. Contrast-enhanced CT scan, MRI and renal scientigraphy are the other methods for the anatomical imaging of CRE [1, 2].

Anatomically, four types of crossed renal ectopia have been described (Fig. 9): Type A: with fusion (Fig. 10); Type B: without fusion (Fig. 11); Type C: solitary crossed; Type D: bilaterally crossed. This anomaly is thought to result because of an abnormally situated umbilical artery that prevents normal cephalic migration [2, 6–14].

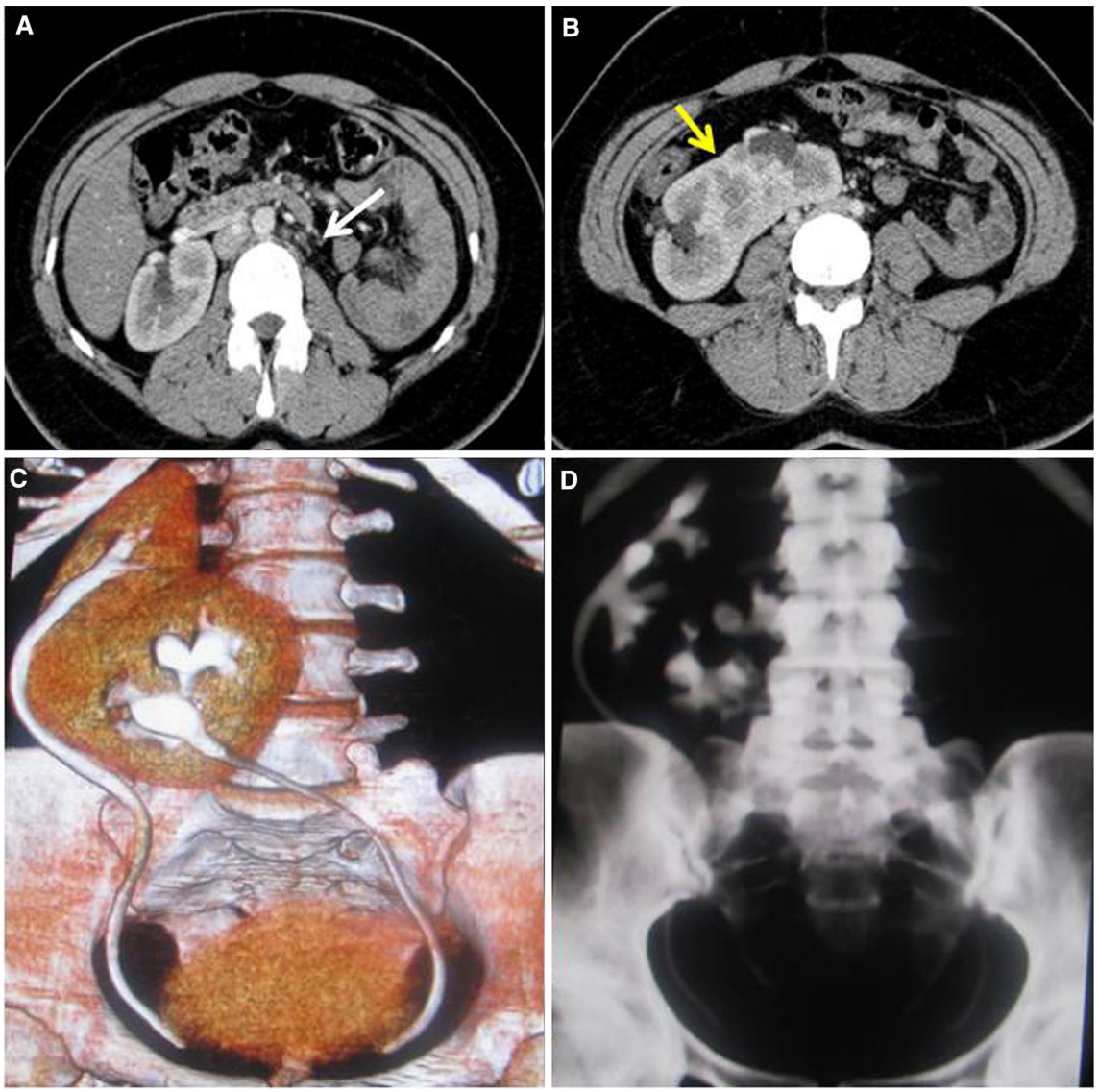

Fig. 10 Cross fused kidney (Type A crossed ectopia). Axial post contrast image (**a**) shows left renal fossa is empty (*white arrow*). A lower section axial image (**b**) shows the left kidney has crossed across the midline and fused to the inferior pole of right kidney (*yellow arrow*). A coronal VRT image (**c**) and coronal MIP image (**d**) showing the ureter of the ectopic kidney (crossed kidney) crossing to the contralateral side and inserting into the urinary bladder

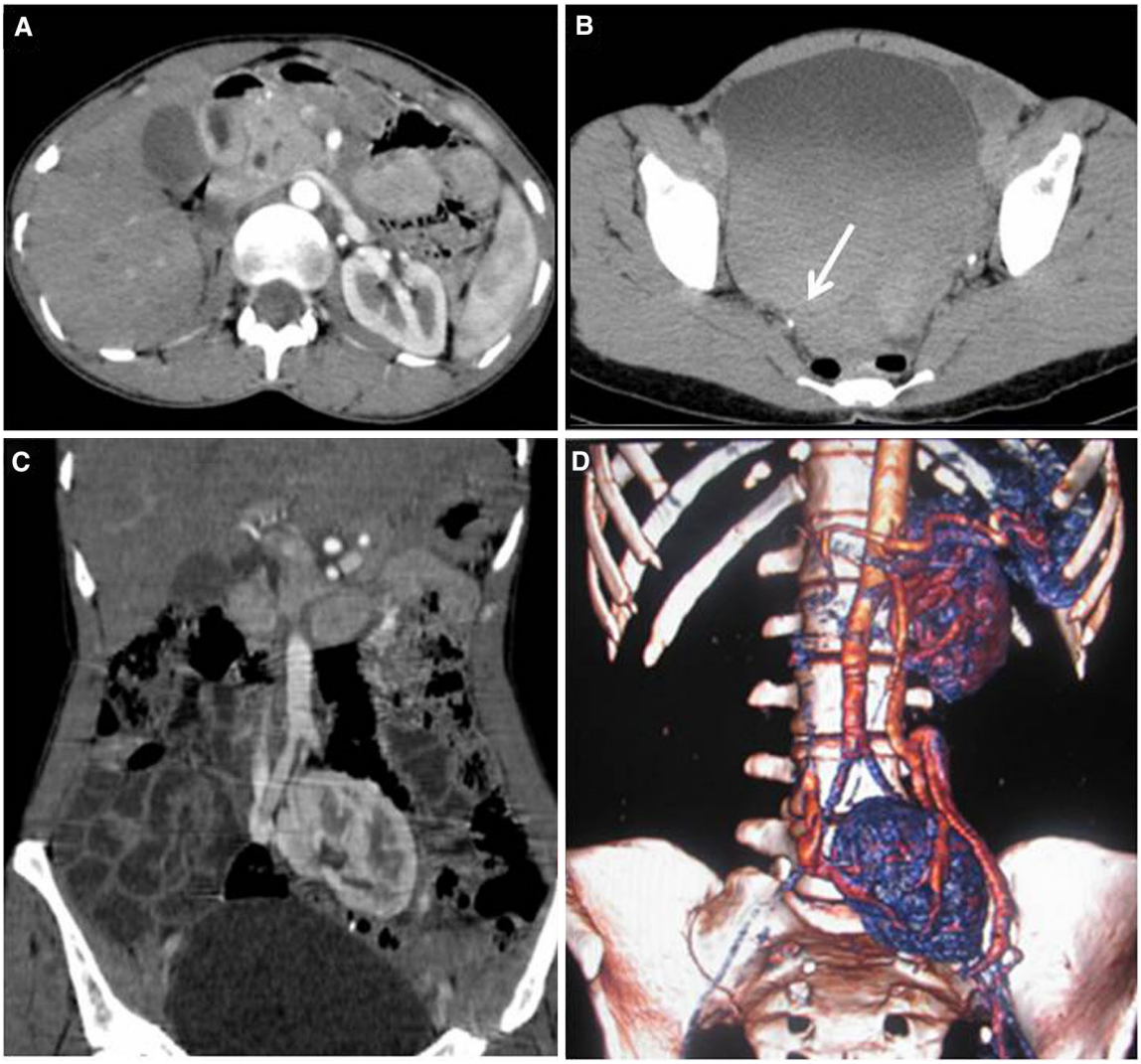

Fig. 11 Cross unfused kidney (Type B crossed ectopia). An axial post contrast CT image (**a**) shows empty right renal fossa. An axial image (**b**) shows the ureter of the ectopic kidney crossing to the contralateral side. A coronal reformatted image (**c**) shows the crossed right kidney across the midline and seen lying inferior to the left kidney. A coronal VRT image (**d**) shows both the kidneys are seen to the left of midline

Abnormalities of rotation

Usually the kidney and renal pelvis rotate ventro-medially during ascent. When this process is not exact, the conditions, known as malrotation (Fig. 12), including nonrotation, incomplete rotation, and reverse rotation, occur. In nonrotation, the renal pelvis presents itself ventrally in relation to the kidney. The pelvis lies between 30° and 90° from the horizontal in incomplete rotation [1, 5, 6]. In reverse rotation, the hila faces laterally. Magnetic resonance urography (MRU) can better delineate the renal outline and axis. USG may suggest malrotation if the renal pelvis is directly anterior to the calyces and the renal parenchyma. The malrotation type is very well delineated on CT and MRI [1].

Abnormalities of contour– hypertropied column of bertin

The column of Bertin, originally described by Bertin as a septum, is a thickened aggregate of the cortical tissue instead of the usual, thin cortical septum that separates two pyramids. On USG it is defined as a round or oval mass with an echogenicity equal to or slightly greater than that of normal adjacent cortex extending from the renal cortex to the renal sinus, can mimic low or iso-echogenic renal tumor (Fig. 13). It is the most common cause of renal pseudotumors [4, 6]. Because of the nonspecific nature of the previously described sonographic findings, patients with an atypical sonographic appearance or unusually large and prominent columns (septa) of Bertin are confirmed by

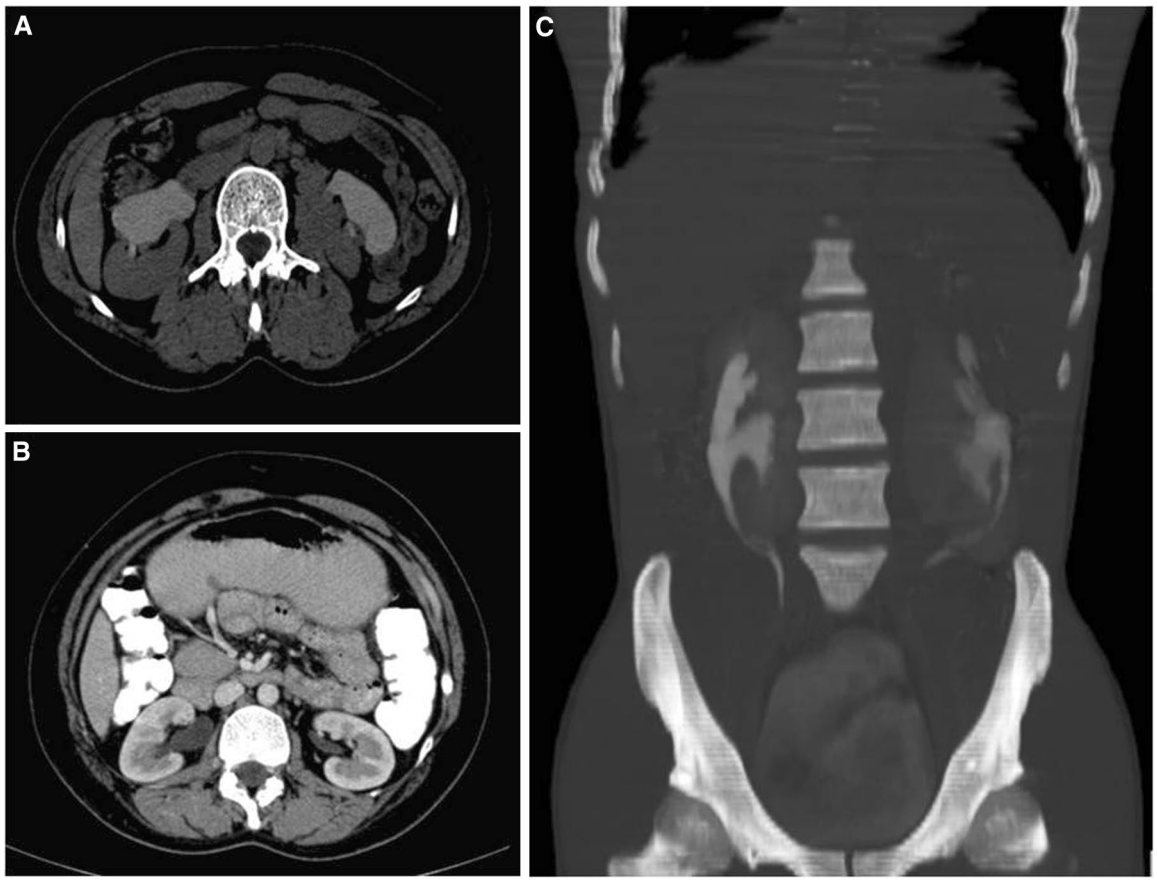

Fig. 12 Malrotation. An axial post contrast image (**a**) shows a nonrotated renal pelvis. An axial image (**b**) shows incomplete rotation where the pelvis faces medially. A coronal MIP image (**c**) shows reverse rotation where the pelvis is rotated laterally

showing its enhancement on contrast-enhanced CT, MRI, or contrast-enhanced sonography (perfusion) to be similar to that of surrounding cortex [4, 6]. Prominent columns (septa) of Bertin are usually followed up with USG

Other congenital pseudotumours include a dromedary hump, a junctional parenchymal defect, a hilar lip and fetal lobulation [6].

Abnormalities of calyx

Calyceal diverticulum refers to a urine-containing cystic cavity within the renal parenchyma [6]. Often found incidentally on radiological imaging, they are generally benign and usually asymptomatic, although complications include infection and stone formation. More importantly, calyceal diverticula may mimic other potentially more serious pathology on imaging such as renal tumor or abscess [4]. Diagnosis is best made by MDCT urography and also by IVU. On MDCT urography, calyceal diverticulum has the appearance of a contrast-filled cystic cavity which communicates with the renal collecting system. On ultrasound, calyceal diverticula appear to have a similar appearance and an echotexture as cysts; hence, they are difficult to differentiate from cysts unless filled with stones [6] (Fig. 14).

Abnormalities of ureter duplications

Ureteral duplication is one of the most common anomalies of the urinary tract (Fig. 15). It can be complete or incomplete [15]. Partial/incomplete duplication results from the branching of the ureteral bud before it connects with the metanephric blastema. They can be further classified as Y or V duplication. Complete duplication occurs when two separate ureteric buds arise from the Wolffian duct [6]. Complete duplication is most often associated with vesicoureteral reflux, ectopic ureterocele, or ectopic ureteral insertion [15].

In the complete type of duplication, although the lower-moiety ureter usually inserts into the trigone, the

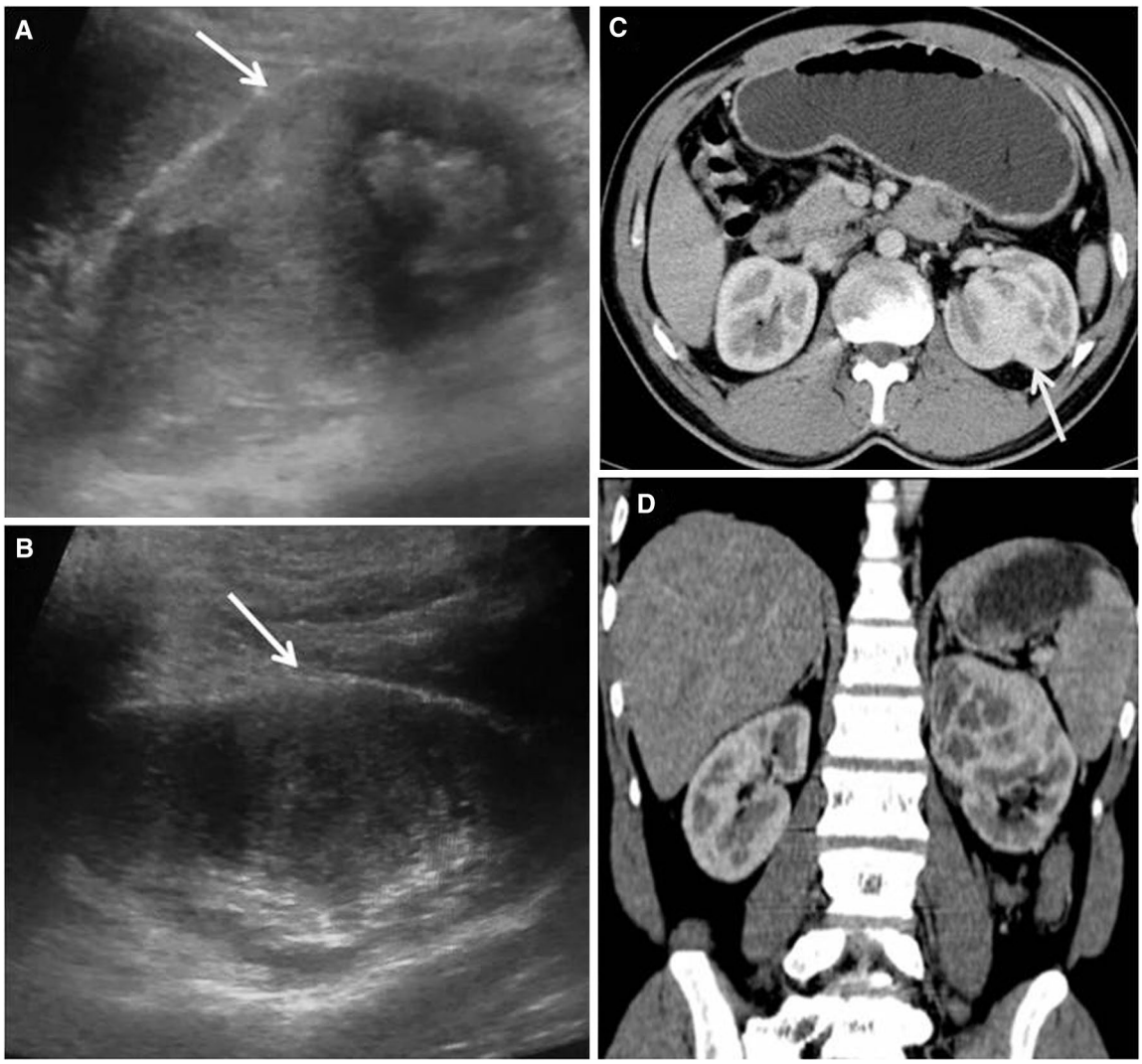

Fig. 13 Hypertrophied column of Bertin. Sagittal ultrasound images (**a**, **b**) of the left kidney show an iso-echoic cortical mass in the mid pole (*white arrow*). Post-contrast axial and coronal CT images show that the so-called mass on the ultrasound was just a thickened column of cortical tissue (*white arrows*) with similar enhancement to that of the adjacent cortex

upper-moiety ureter usually inserts into the inferior and medial portion of the urinary bladder. This pattern is known as the Weigert-Meyer rule. In case of partial duplication with distal obstruction, the peristalsis down in one ureter may force urine via reflux up the other which is known as the "yo-yo" phenomenon [6, 15]. Complications of a double collecting system include obstruction, stone formation, ureterocele, and vesicoureteral reflux [6, 15]. Imaging modalities commonly used are IVU and voiding cystourethrography, as well as USG, CT, and MRI. Abnormalities of ureteral duplication are well demonstrated by IVU and vesicoureteral reflux is diagnosed by voiding cystourethrography; however, in recent years, MDCT urography and MRI are helpful [15].

Conclusions

Imaging plays an important role in diagnosing renal anomalies and their complications. USG is, generally, the initial diagnostic study of choice for evaluating renal anomalies and their complications. However, MDCT is a useful imaging method that allows assessment of congenital anatomic variants of the kidney and ureter, providing information that

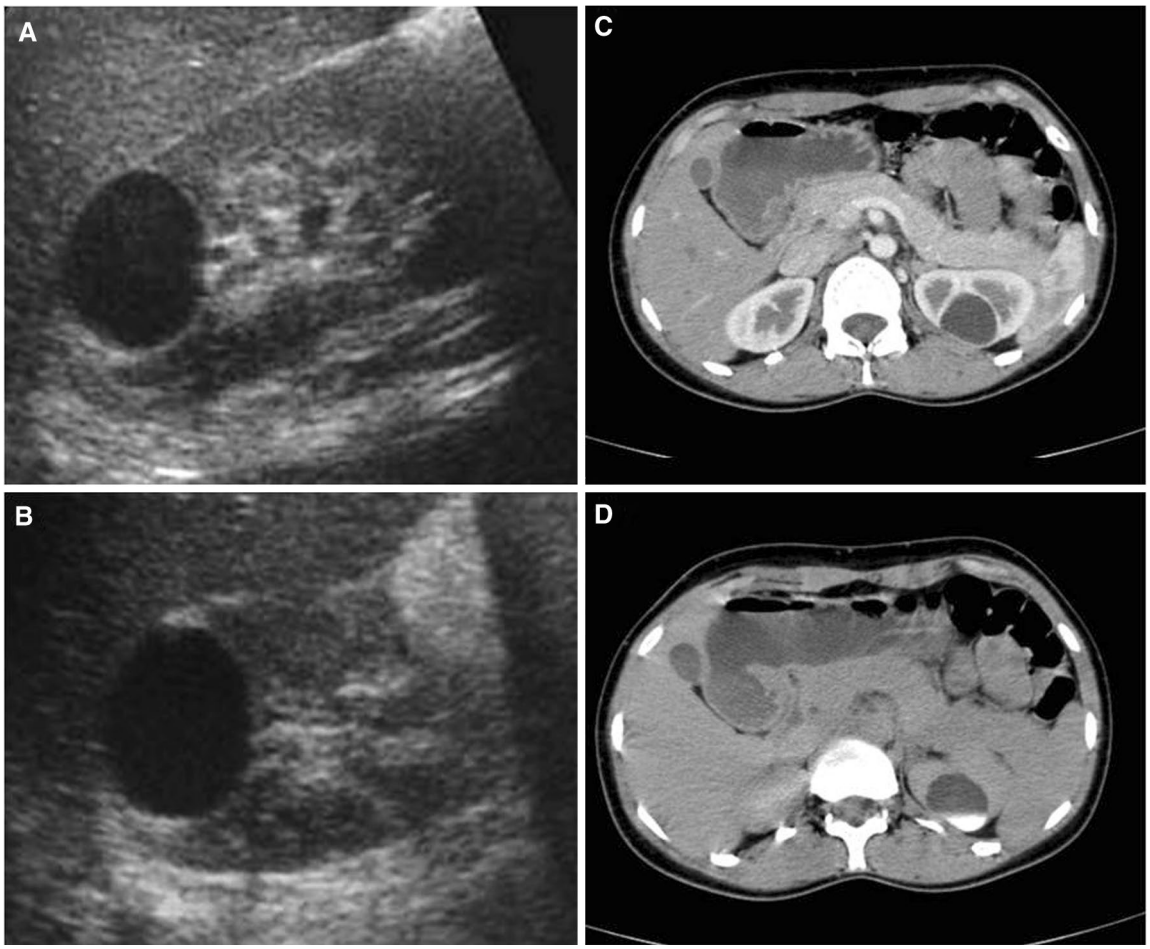

Fig. 14 Calyceal diverticulum. Sagittal (**a**) and transverse (**b**) ultrasound images of the left kidney show an anechoic cystic lesion in the upper pole. Post-contrast nephrographic and delayed-phase axial CT images **(C, D)** show there is filling of the contrast within the cystic structure

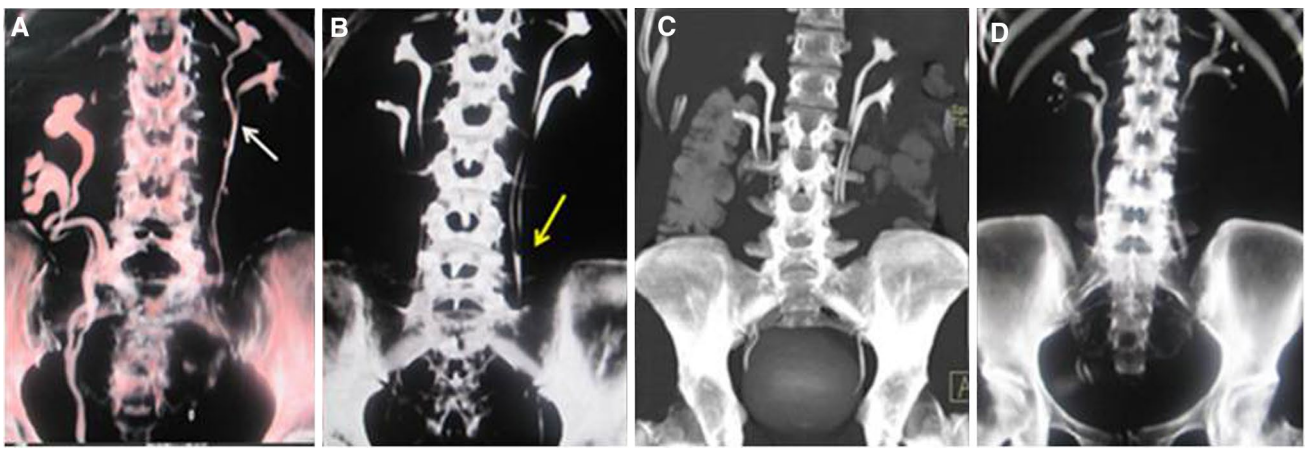

Fig. 15 Ureteral duplications: A coronal VRT image (**a**) shows a left-sided, Y-type partial duplication (*white arrow*). A coronal VRT image (**b**) shows a left-sided, V-type partial duplication (*yellow arrow*) with right-sided complete duplication. A full MIP coronal image (**c**) shows bilateral complete duplication. A coronal VRT image (**d**) shows right-sided complete duplication

would otherwise require multiple other imaging methods. As cross-sectional imaging with CT has its own limitations with regard to contrast material and radiation exposure, wise use of the modality is a must. Familiarity of variable imaging features of renal anomalies and their complications will provide early diagnosis and proper management.

Compliance with ethical standards

Conflict of interest None.

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