

Multidetector CT urography of renal fusion anomalies

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ABSTRACT

Renal fusion anomalies, in which both kidneys are fused together in early embryonic life, are rarely encountered. Once a fused kidney is diagnosed or suspected, further laboratory and imaging evaluation should be performed to assess the status of the kidneys and to look for treatable causes of renal pathology. The early diagnosis of potential complications that can accompany this anomaly must be made in order to prevent permanent renal damage. The advantage of multidetector computed tomographic (MDCT) urography is its ability to depict the normal urinary tract anatomy, including both the renal parenchyma, and collecting structures and ureters. MDCT urography is helpful to screen for the presence of stones, hydronephrosis or masses. Additionally, it provides information about the vascular supply of the fused kidneys. Therefore, MDCT urography enables a comprehensive evaluation of patients with renal fusion anomalies in a single examination. Especially three-dimensional reformatted images can provide good delineation of congenital fusion anomalies of the kidney. In this study we report our experience with MDCT urography for the anatomic demonstration of renal fusion anomalies.

Key words: • multidetector computed tomography
• renal fusion • horseshoe kidney • crossed fused renal ectopia • cake kidney

Renal fusion anomalies, of the kidney, in which both kidneys are fused together in early embryonic life, are rarely encountered. They may be partial or complete. Partial renal fusion is represented by the horseshoe kidney and crossed renal ectopia with fusion. Cake kidney is an anomaly characterized by the complete fusion of both kidneys; it accounts for only 2% of fused kidneys (1). Renal fusion anomalies occur predominantly in males (2). The early diagnosis of complications that can accompany this anomaly must be made to prevent permanent renal damage. In this study we report our experience with multidetector computed tomographic (MDCT) urography for the anatomic demonstration of renal fusion anomalies.

Embryology

In the embryo, the two masses of metanephrogenic tissue lie within the pelvis. Each developing kidney reaches its definitive position in the lumbar region following complicated movements involving ascent, lateral migration, axial deflection, and internal rotation. The nephrogenic blastemas are squeezed together between the umbilical arteries at the beginning of the cranial migration of the ureteral buds, which may cause their fusion. Fused kidneys are usually prevented from ascending to their normal position and remain in an ectopic position. In all fused kidneys, the arterial supply and venous drainage are grossly abnormal. This reflects the primitive arrangement variably seen in ectopic kidneys, often because of their very limited rostral migration (2).

Multidetector computed tomographic urography: imaging technique

MDCT urography was performed with a 16-row multislice CT (Light-speed Ultra, General Electrical Medical Systems, Milwaukee, Wisconsin, USA). Scans were obtained in three phases: unenhanced, nephrographic phase, and pyelographic phase. The patient is positioned supine and receives a supplemental infusion of 250 mL normal saline immediately following the injection of contrast medium. Initial unenhanced images are obtained from the diaphragm to the symphysis pubis with 2.5 mm collimation and 1.25 mm reconstruction interval. The nephrographic phase was acquired with 2.5 mm collimation 100 s after administration of 100 mL of iodinated contrast agent (Iodixanol, Visipaque 320 mgI/mL, GE Healthcare, Milwaukee, Wisconsin, USA) at a rate of 3 mL/s. The pyelographic phase was acquired after a 10 min delay with 1.25 mm collimation. In some patients, delayed pyelographic phase images were acquired after 45 min or 60 min delay with 2.5 mm collimation. For 3-dimensional image reconstruction, the raw CT data were processed on a separate workstation (Advanced Workstation 4.2, GE Medical Systems, Wisconsin, USA) with multiplanar reformatting, maximum intensity projection, and volume rendering.

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Figure 1. a, b. Axial MDCT images (a, b) show horseshoe kidneys that are fused with functioning renal parenchymal (a) and fibrous tissue isthmus (white arrow, b) at their lower poles. The isthmus lies anterior to the aorta just below the origin of the inferior mesenteric artery (black arrow, a). A cortical cyst is also seen in the left kidney (b). (A, aorta; C, cyst.)

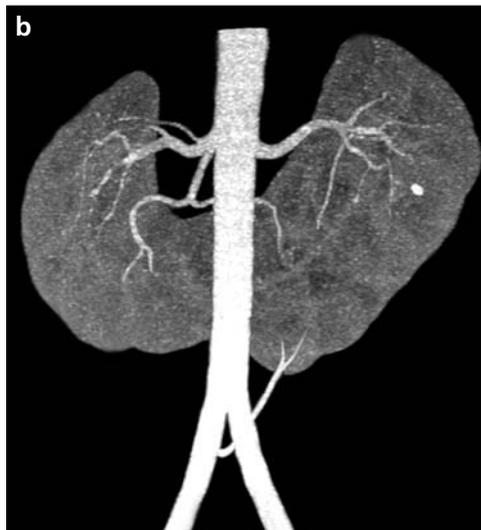
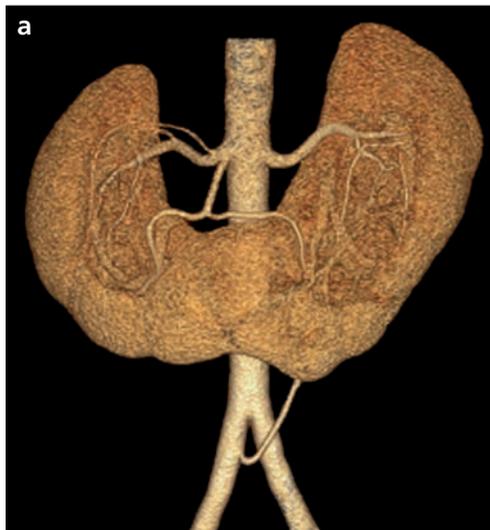


Figure 2. a, b. Coronal volume rendering (a) and maximum intensity projection (b) MDCT images show the arterial supply of the horseshoe kidney consisting of two main renal arteries arising from the aorta with the addition of one accessory left renal artery arising from the right iliac artery. The right renal artery has prehilum branching. The isthmus receives a branch from the right renal artery. A stone in the left kidney is also seen (b).

Horseshoe kidney

Horseshoe kidney is the most common renal fusion anomaly. It consists of two distinct functioning kidneys lying vertically on either side of the midline, connected at their lower poles by an isthmus of functioning renal parenchyma or, rarely, fibrous tissue that crosses the midline of the body (Fig. 1). In rare instances, the upper poles may be the site of fusion (2). It accounts for 90% of all renal fusion anomalies and

occurs in approximately 0.25% of the population (1).

Horseshoe kidneys may be found at any location along the path of normal renal ascent from the pelvis to the midabdomen. The isthmus usually lies anterior to the great vessels, at the level of the third to fifth lumbar vertebra, just below the origin of the inferior mesenteric artery from the aorta (Fig. 1). Rarely, it is posterior to these vessels or runs between them (2).

The blood supply to the horseshoe kidney can be quite variable. In 30% of cases, it consists of one renal artery to each kidney, but it may be atypical with duplicate or even triplicate renal arteries supplying one or both kidneys. The isthmus and adjacent parenchymal masses may receive a branch from each main renal artery, or they may have their own arterial supply from the aorta, inferior mesenteric artery, iliac arteries, or sacral arteries (3) (Fig. 2).

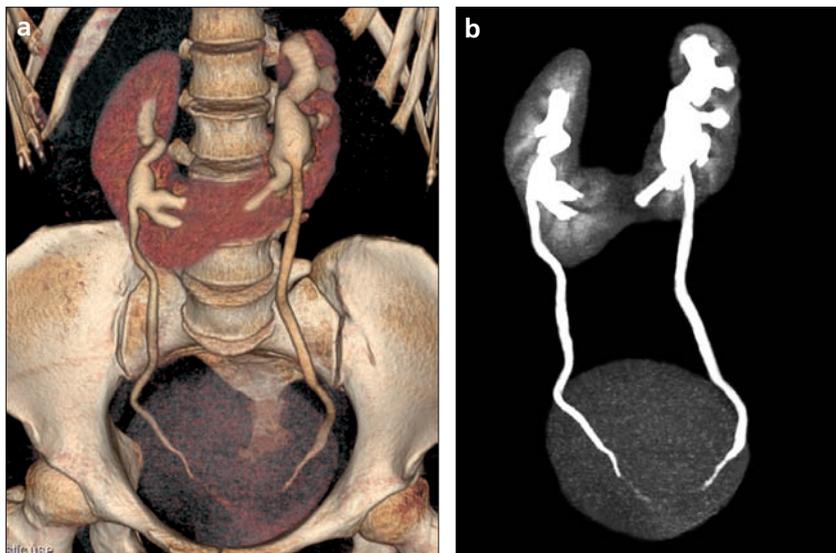


Figure 3. a, b. Coronal volume rendering (a) and maximum intensity projection (b) MDCT images in the pyelographic phase show horseshoe kidney. Two distinct kidneys lying vertically on either side of the midline are connected at their lower poles by an isthmus of functioning renal parenchyma. Both ureters enter the bladder in normal relationship. Mild hydronephrosis in the left kidney is also seen.

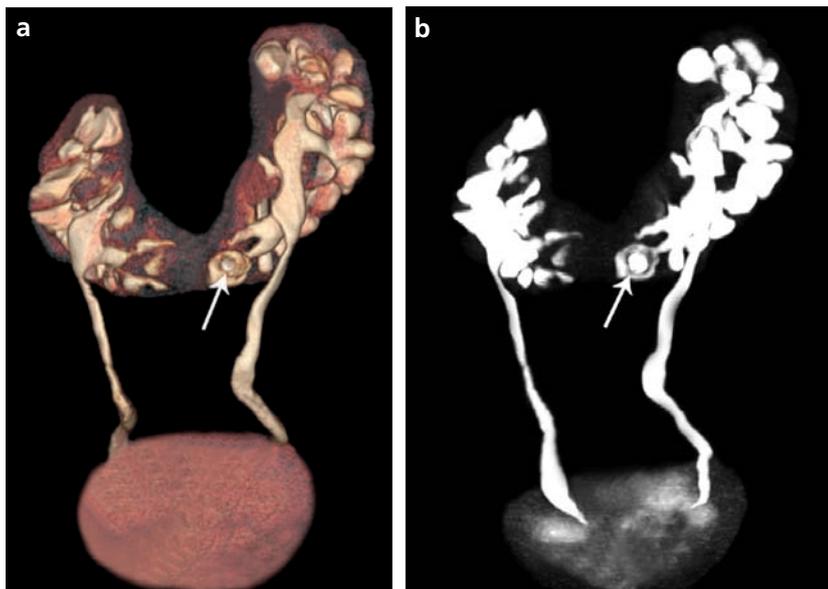


Figure 4. a, b. Coronal volume rendering (a) and maximum intensity projection (b) MDCT images in the pyelographic phase show hydronephrosis of both kidneys due to upper ureteral angulation that causes stasis of urine. A left renal calculus is seen (arrow).

The renal axis and orientation of the pelvicalyceal system are abnormal, with the kidney appearing more vertical than normal, the lower poles lying more medial than the upper poles, and the renal pelvises located more anteriorly or laterally than normal. The ureter may insert high on the renal pelvis and pass anteriorly over the lower poles and isthmus, probably as the result of incomplete renal rotation. Despite upper ureteral angulation, the lower ureter usually enters the bladder

normally and is rarely ectopic (2) (Fig. 3). Because of their abnormal course, the ureters are prone to be obstructed, precipitating hydronephrosis, infection, and stone formation (2, 4).

Nearly one-third of patients with horseshoe kidney remain asymptomatic, and the horseshoe kidney is an incidental finding during radiological examinations. When symptoms are present, they usually are related to hydronephrosis, infection, or calculus formation (2, 4) (Figs. 4–6). The most

common symptom that reflects these conditions is vague abdominal pain that may radiate to the lower lumbar region. The Rovsing sign, consisting of abdominal pain, nausea, and vomiting on hyperextension of the spine, is rare. Signs or symptoms of urinary tract infection occur in 30% of patients. In children, urinary tract infection is the most common presentation. Ureteropelvic junction obstruction causing significant hydronephrosis occurs in one-third of individuals (2). Stones are thought to be due to associated hydronephrosis or ureteropelvic junction obstruction, causing stasis of urine (5).

Associated genitourinary anomalies in horseshoe kidney are common and occur in as many as two-thirds of patients. Vesicoureteral reflux, ureteral duplication, ectopic ureterocele, retrocaval ureter, cystic disease including multicystic dysplasia and adult polycystic kidney disease, hypospadias, undescended testes, bicornuate uterus, and septate vagina may be present (2). Horseshoe kidney has been reported to be associated with increased risk for renal neoplasms such as Wilms tumors, renal carcinoids, and transitional cell carcinoma. Renal cell carcinoma is the most common tumor reported with horseshoe kidney, although its reported incidence is not higher than that in the normal population (6).

The horseshoe kidney is also associated with congenital anomalies outside the genitourinary system. The organ systems most commonly affected include skeletal (hemivertebrae with scoliosis, rib defects, clubfoot, congenital hip dislocation), cardiovascular (ventriculoseptal defects), gastrointestinal (anorectal malformation, malrotation, and Meckel diverticulum), and central nervous systems (neural tube defects) (2).

Crossed fused renal ectopia

Crossed fused renal ectopia is the second most common fusion abnormality of the kidney, with an estimated incidence of approximately 1:1300–1:7500 (7). In crossed fused ectopia, one kidney crosses over to opposite side, and the parenchyma of the two kidneys fuse. Most commonly, the upper pole of the inferiorly positioned crossed ectopic kidney is fused to the lower pole of the superior, normally positioned kidney. The ureter of the ectopic kidney crosses the midline and enters the

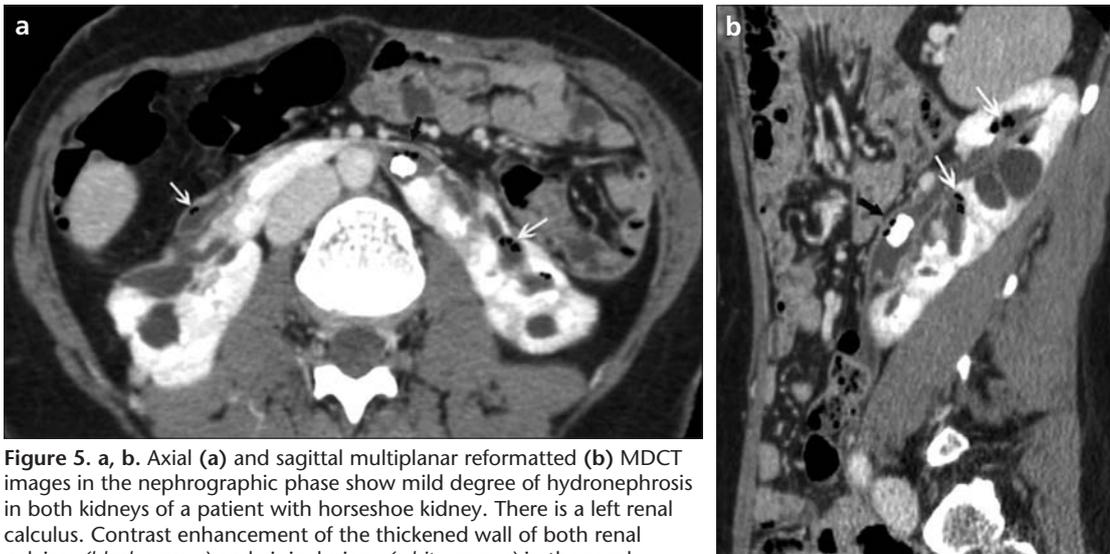


Figure 5. a, b. Axial (a) and sagittal multiplanar reformatted (b) MDCT images in the nephrographic phase show mild degree of hydronephrosis in both kidneys of a patient with horseshoe kidney. There is a left renal calculus. Contrast enhancement of the thickened wall of both renal pelvises (black arrows) and air inclusions (white arrows) in the renal collecting systems caused by infection are seen.

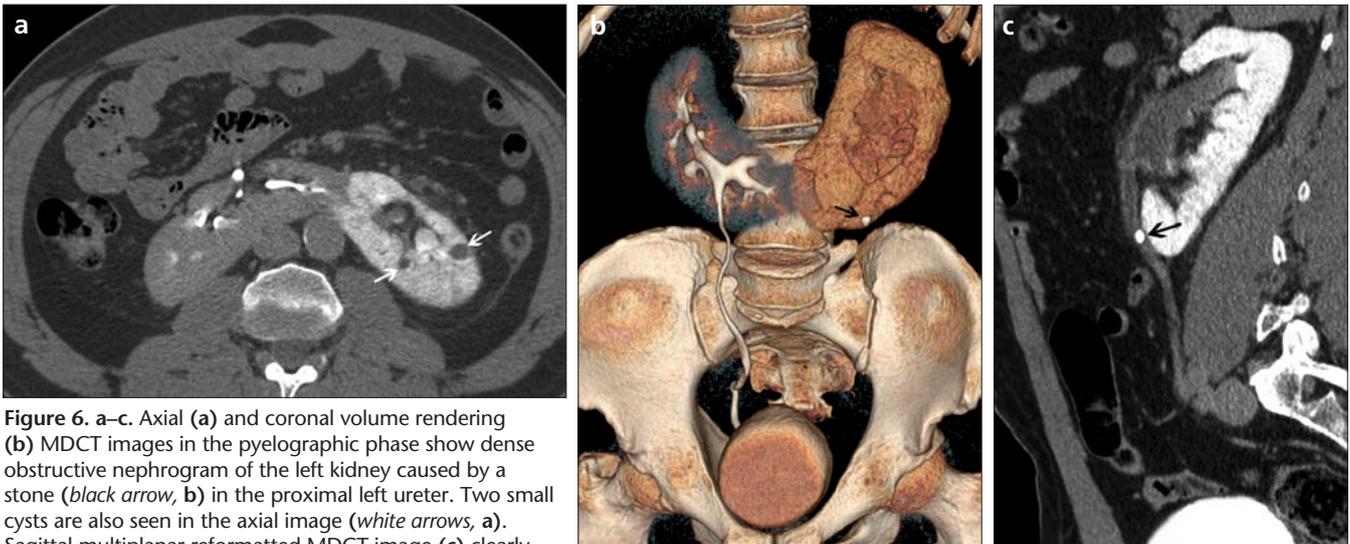


Figure 6. a–c. Axial (a) and coronal volume rendering (b) MDCT images in the pyelographic phase show dense obstructive nephrogram of the left kidney caused by a stone (black arrow, b) in the proximal left ureter. Two small cysts are also seen in the axial image (white arrows, a). Sagittal multiplanar reformatted MDCT image (c) clearly demonstrates the left ureteral stone (black arrow).

bladder on the opposite side (Fig. 7). The left kidney is most frequently ectopic (crossing to the right side of the abdomen) (2) (Fig. 8).

McDonald and McClellan classified crossed ectopic kidney into four types: crossed renal ectopia with fusion (85%), crossed renal ectopia without fusion (10%), solitary crossed renal ectopia, and bilaterally crossed renal ectopia (8). Six variations of crossed fusion have been described. In decreasing order of frequency, they are: type 1, inferior crossed fused ectopia; type 2, sigmoid or S-shaped kidney; type 3, unilateral lump kidney; type 4, unilateral disc kidney; type 5, L-shaped kidney; type 6, superior crossed fused ectopia (2).

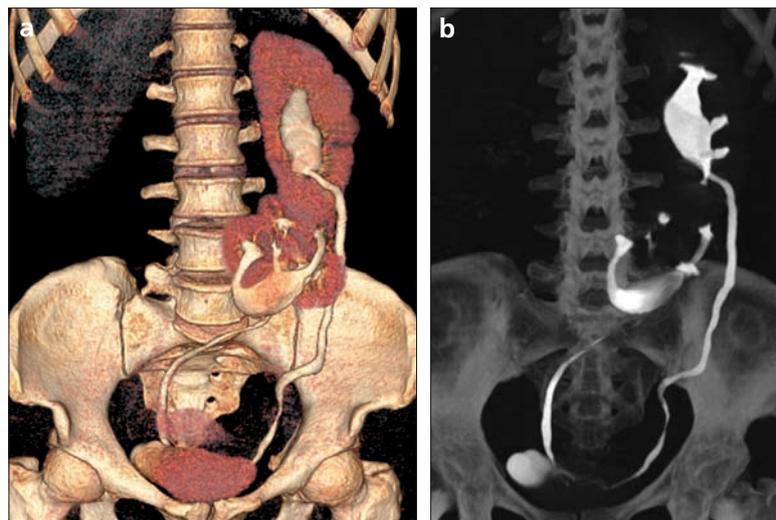


Figure 7. a, b. Coronal volume rendering (a) and maximum intensity projection (b) MDCT images show crossed fused renal ectopia on the left side. The ureter of the right ectopic kidney crosses the midline and enters the bladder on the opposite side.

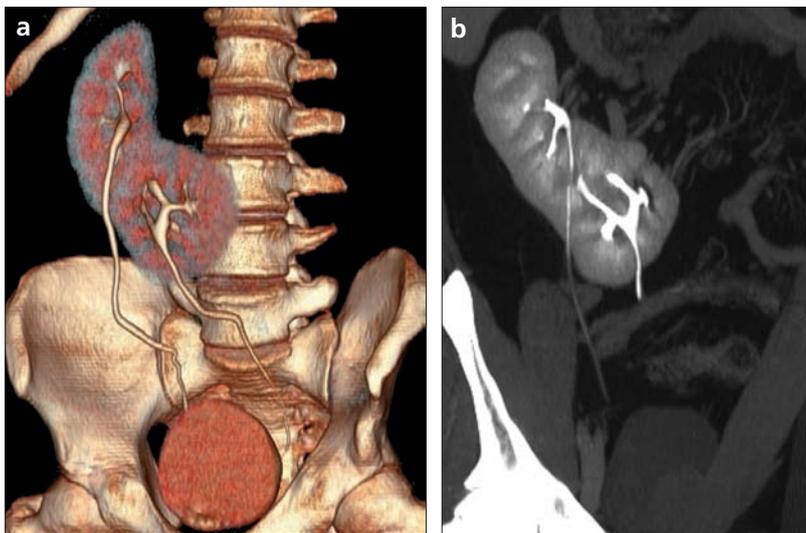


Figure 8. a, b. Coronal volume rendering (a) and oblique maximum intensity projection (b) MDCT images show crossed fused renal ectopia on the right side.

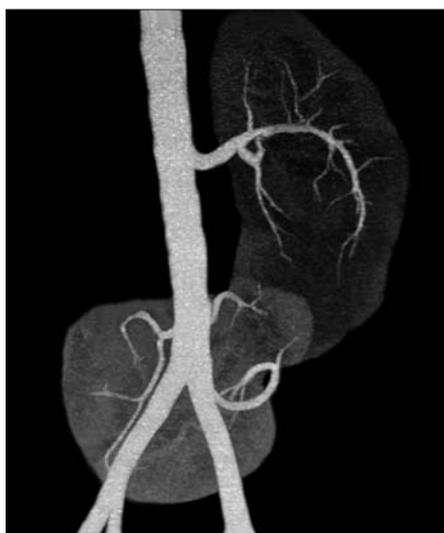


Figure 9. Coronal volume rendering MDCT image shows the arterial supply of the crossed fused ectopic kidneys consisting of one left renal artery arising from upper aorta and three right renal arteries, two of which arise from the lower aorta and one of which arises from the contralateral iliac artery. A poor nephrogram is also seen in the left kidney, caused by ureteropelvic obstruction.



Figure 10. Oblique multiplanar reformatting MDCT image in the nephrographic phase shows a mild degree of hydronephrosis, contrast enhancement of thickened wall of the renal pelvis (white arrow) and a stone (black arrow) in the lower pole of the left superior kidney. In addition, stranding of the fat in the vicinity of the inflamed renal pelvis in the left superior kidney is seen.

Blood supply to the ectopic kidney most frequently arises from the vessels on the ipsilateral side but occasionally arises from the contralateral side. Both the normally located kidney and its fused mate commonly have aberrant arterial anatomy; the arterial supply is from the upper abdominal aorta in 25% of cases and from the lower aorta or iliac arteries in the remainder. While the total number of arteries ranges from one to six, most commonly there are two to four major arteries to the two kidneys (2, 9) (Fig. 9).

Cross fused renal ectopia is typically asymptomatic and is diagnosed as an incidental finding when the patient is examined for other medical diseases. When symptoms do occur, the most common symptoms reported are abdominal or flank pain, a palpable mass, hematuria, dysuria, and urinary tract infections (2, 10). The urological conditions associated with this anomaly are: nephrolithiasis (Fig. 10), ureteropelvic junction obstruction (Fig. 11), hydronephrosis (Fig. 12), reflux, and ectopic ureteroceles (2). The abnormal position of the kidney can lead to poor outflow and a predisposition to hydronephrosis, calculi, and infection. Tumors in crossed ectopic kidneys have been reported sporadically (11). Pediatric patients most often present with multiple congenital anomalies, especially of the skeletal anomalies of the bony pelvis, and vertebrae. Other anomalies associated with this condition are cardiovascular, gastrointestinal, and other genitourinary anomalies, as previously mentioned (2, 10).

Cake kidney (complete renal fusion)

Cake kidney is a rare congenital anomaly of the genitourinary system, with fewer than 30 cases described in the literature (12). The term cake kidney or fused pelvic kidney was defined by Glenn as an anomaly in which “the entire renal substance is fused into one mass, lying in the pelvis, and giving rise to two separate ureters which enter the bladder in normal relationship” (1) (Fig. 13). In rare instances, such kidneys possess one ureter (13). The fused kidney occupies prevertebral or presacral space (1) (Fig. 14).

The cake kidney may remain asymptomatic and be detected at autopsy. It may become infected or may cause local pain from dragging on the renal vessels by the weight of the organ (1, 14).

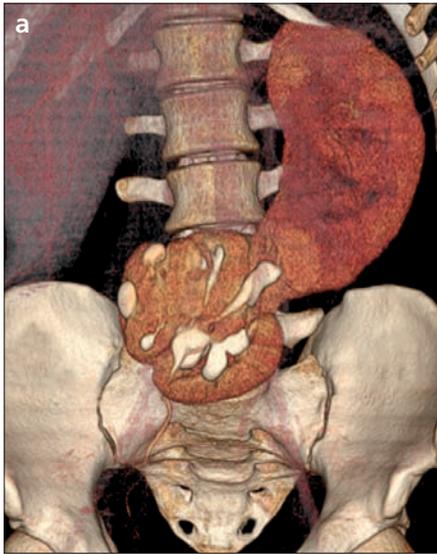


Figure 11. a–c. Coronal volume rendering MDCT image in the pyelographic phase (a) shows crossed fused renal ectopia on the left side. The pyelogram of the left superior kidney was not seen. Axial (b) and sagittal multiplanar reformatted (c) MDCT images in the pyelographic phase show severe hydronephrosis in the left kidney caused by ureteropelvic junction obstruction.

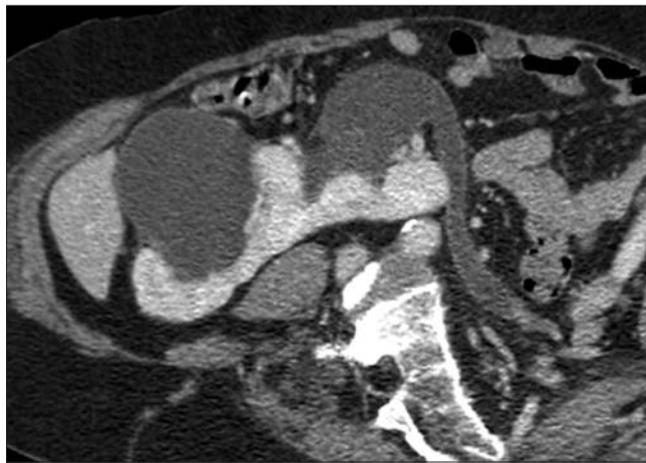


Figure 12. Oblique multiplanar reformatted MDCT image in the nephrographic phase shows crossed fused renal ectopia on the right side with severe hydronephrosis.

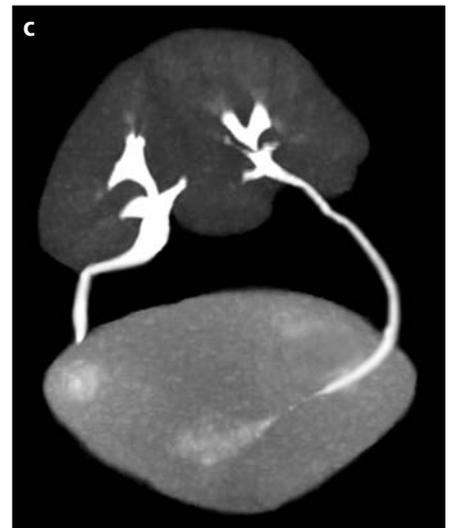
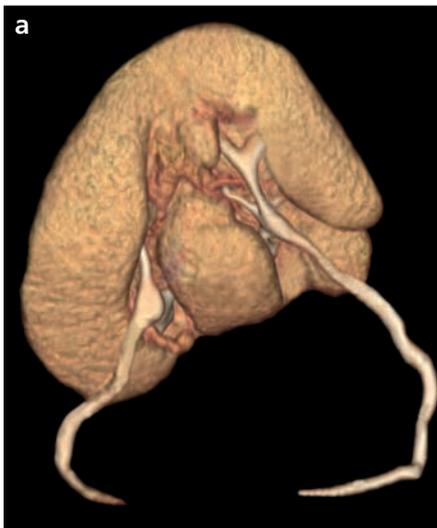


Figure 13. a–c. Anterior (a) and posterior (b) coronal volume rendering MDCT images in the nephrographic phase show cake kidney, in which entire renal substances of two kidneys are fused into one mass. Coronal maximum intensity projection MDCT image in the pyelographic phase (c) shows cake kidney overlying the bladder, drained by two separate ureters which enter the bladder in normal relationship.



Figure 14. Axial MDCT image shows cake kidney lying in the midline presacral area (K, kidney; S, sacrum).

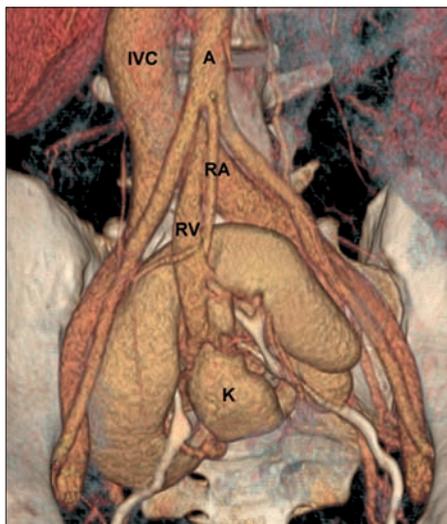


Figure 15. Coronal volume rendering MDCT image in the nephrographic phase demonstrates both the renal artery and vein. The renal artery arises at the aortic bifurcation and the renal vein opens into left common iliac vein. In addition, both ureters are also seen. (A, aorta; IVC, inferior vena cava; RA, renal artery; RV, renal vein; K, kidney.)

The majority of diagnosed cases have been reported to present concomitant anomalies in other organs, such as abnormal testicular descent, tetralogy of Fallot, vaginal absence, sacral agenesis, caudal regression syndrome, spina bifida, and anal abnormalities (13).

The vascular supply of the cake kidney is consistent with its arrested migration. In the developing kidney the vascular supply is re-established progressively as it migrates superiorly to reach its normal position. If the migration is arrested, the temporary blood supply will become permanent. Therefore, cake kidney derives its blood supply from the aorta near the bifurcation or from the common iliac vessels. Venous drainage is usually into the distal inferior vena cava or the common iliac veins (Fig. 15). This anomalous blood supply is at increased risk for vascular compromise due to pelvic trauma, vascular disease, pregnancy, or space-occupying lesion (15).

Conclusion

Once a fused kidney is diagnosed or suspected, further laboratory and imaging evaluation should be performed to assess the status of the kidneys and to look for treatable causes of renal pathology. The early diagnosis of complications that can accompany this anomaly must be made to prevent permanent renal damage.

To date, the diagnosis of renal fusion anomalies has been made by ultrasonography, excretory urography, and CT. Ultrasound is often the initial procedure performed in the work-up of the patient. Excretory urography is often

useful, but the absence of fusion may not be apparent unless the two renal masses are widely separated. CT allows accurate diagnosis. The advantage of MDCT urography is its ability to depict normal urinary tract anatomy, including the renal parenchyma and collecting structures and ureters. Congenital anomalies of the collecting system and ureters can be visualized better with MDCT urography than with conventional CT. Congenital anomalies of renal position, number, and form are easily depicted with MDCT urography, including renal ectopia, malrotation, and fusion anomalies. MDCT urography is helpful to screen for the presence of stones, hydronephrosis, or masses. Additionally, it provides information about the vascular supply of the fused kidneys. Therefore, MDCT urography has the potential to provide a comprehensive evaluation of patients with renal fusion anomalies in a single examination.

The main limitations of MDCT urography are the increased risk of substantial radiation exposure and the need for iodinated contrast agents (16–18). Magnetic resonance imaging (MRI) is an acceptable alternative modality in patients with contraindications to the use of iodinated contrast agents or when radiation dose is an issue, as in imaging of pediatric patients, pregnant women, and repeated follow-up examinations. However, MRI is not sensitive in detection of urolithiasis, and its spatial resolution is inferior to that of MDCT. Less common availability and higher cost are main limitations of MRI (19, 20).

In conclusion, the diagnosis of fused kidney is not necessarily associated with a poor prognosis. It requires long-term follow-up of renal function, early detection of complications, and exclusion of concomitant congenital anomalies. MDCT urography enables a comprehensive evaluation of patients with renal fusion anomalies in a single examination. Three-dimensional reformatted images can provide particularly good delineation of congenital fusion anomalies of the kidney.

References

1. Kaufman MH, Findlater GS. An unusual case of complete renal fusion giving rise to a 'cake' or 'lump' kidney. *J Anat* 2001; 198:501–504.
2. Bauer SB. Anomalies of the upper urinary tract. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ, eds. *Campbell's urology*. 8th ed. Philadelphia: WB Saunders, 2002; 1898–1906.
3. Yoshinaga K, Kodama K, Tani I, Toshimori K. Morphological study of a horseshoe kidney with special reference to the vascular system. *Anat Sci Int* 2002; 77:134–139.
4. Castro JE, Green NA. Complications of horseshoe kidney. *Urology* 1975; 6:344–347.
5. Evans WP, Resnick MI. Horseshoe kidney and urolithiasis. *J Urol* 1981; 125:620–621.
6. Hohenfellner M, Schultz-Lampel D, Lampel A, Steinbach F, Cramer BM, Thurhoff JW. Tumor in the horseshoe kidney: clinical implications and review of embryogenesis. *J Urol* 1992; 147:1098–1102.
7. Rinat C, Farkas A, Frishberg Y. Familial inheritance of crossed fused renal ectopia. *Pediatr Nephrol* 2001; 16:269–270.
8. McDonald JH, McClellan DS. Crossed renal ectopia. *Am J Surg* 1957; 93:995–999.
9. Bailey SH, Mone MC, Nelson EW. Transplantation of crossed fused ectopic kidneys into a single recipient. *J Am Coll Surg* 2002; 194:147–50.

10. Boyan N, Kubat H, Uzum A. Crossed renal ectopia with fusion: report of two patients. *Clin Anat* 2007; 20: 699–702.
11. Gerber WL, Culp DA, Brown RC, Chow KC, Platz CE. Renal mass in crossed-fused ectopia. *J Urol* 1980; 123:239–244.
12. Calado AA, Macedo A Jr, Strougi M. Cake kidney drained by single ureter. *Int Braz J Urol* 2004; 30:321–322.
13. Goren E, Eidelman A. Pelvic cake kidney drained by single ureter. *Urology* 1987; 30:492–493.
14. Ignjatovic I, Stojkovic I, Dinic LJ, Jovanovic M, Ivanovic D. Bilateral fused pelvic kidney ectopia with a single draining ureter solved by craniolateral displacement of the kidney and Boari-modified bladder-calycal anastomosis with bladder augmentation. *Int J Urol* 2007; 14:552–554.
15. Brock JW 3rd, Braren V, Phillips K, Winfield AC. Caudal regression with cake kidney and a single ureter: a case report. *J Urol* 1983; 130:535–536.
16. Chow LC, Sommer FG. Multidetector CT urography with abdominal compression and three-dimensional reconstruction. *AJR Am J Roentgenol* 2001; 177:849–855.
17. Kawashima A, Vrtiska TJ, LeRoy AJ, et al. CT urography. *Radiographics* 2004; 24: S35–58.
18. Noroozian M, Cohan RH, Caoili EM, Cowan NC, Ellis JH. Multislice CT urography: state of the art. *Br J Radiol* 2004; 77: S74–86.
19. Beer AJ, Dobritz M, Zantl N, Weirich G, Stollfuss J, Rummeny EJ. Comparison of 16-MDCT and MRI for characterization of kidney lesions. *AJR Am J Roentgenol* 2006; 186:1639–1650.
20. Nikken JJ, Krestin GP. MRI of the kidney: state of the art. *Eur Radiol* 2007; 17:2780–2793.