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Original Research Article

CROSSED RENAL ECTOPIA WITHOUT FUSION: A MULTIDETECTOR COMPUTED TOMOGRAPHY STUDY

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ABSTRACT

Background: Congenital positional, rotational and fusion anomalies of the kidney are frequently encountered. Crossed renal ectopia is a condition in which the kidney is located on the side contralateral to its ureteral insertion into the urinary bladder. Crossed renal ectopia without fusion is a very rare anomaly sporadically reported in the literature. We have attempted to analyse such renal anomalies in a large series of patients.

Materials & Methods: Contrast enhanced multidetector computed tomographic scans of 960 patients (491 males and 469 females, age range 4-90 years) were reviewed .

Observations: Crossed renal ectopia without fusion was detected in three patients (0.31%; 1 in 320 cases). All three patients were males and left- to-right renal ectopia was noted in two cases and right-to-left ectopia in one case, in whom the ectopic right kidney was exhibiting multicystic dysplasia. An interesting observation was vascularisation of crossed ectopic unfused left kidneys by branches arising from the right common iliac artery.

Conclusion: Crossed renal ectopia without fusion is an extremely rare anomaly and may remain asymptomatic without being detected. Multidetector computed tomography angiography is an excellent imaging modality to detect renal positional and rotational anomalies. To the best of our knowledge the present study is the first study to detect renal anomalies in a large series of cases.

KEY WORDS: Renal ectopia, Renal anomalies, Renal positional anomaly.

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INTRODUCTION

Normally kidneys are positioned in the upper part of posterior abdominal wall in the paravertebral gutters. Simple renal ectopia occurs when one or both the kidneys are located in an abnormal position in the lumbar region, iliac fossa or pelvic cavity. Condition of a kidney located on the side opposite to the side of its ureteral insertion into

the bladder is called as crossed ectopia. In this condition the ureter from the ectopic kidney characteristically crosses the midline [1]. Mc Donald and Mc Clellan classified the crossed renal ectopia (CRE) into four types: crossed ectopia with fusion, crossed ectopia without fusion, solitary crossed ectopia and bilateral crossed ectopia [2]. Crossed ectopic kidneys are fused with their orthotopically located ipsilateral mates in 90 % of cases. Crossed fused renal ectopia is noted in 1 out of 7500 autopsies whereas, crossed ectopia without fusion is ten times more rarer, observed in 1 out of 75000 autopsies [3]. The left kidney crossing to the right side is three times more common and males are affected more than the females in a ratio of 2:1.

Crossed ectopic kidneys exhibit signs of malrotation with the hilum facing anteriorly or laterally. Their vascularity also shows abnormalities. In many cases the anomaly remains asymptomatic and detected incidentally at autopsy, surgery or radiological investigations for unrelated problems. Sometimes it may cause vague abdominal pain and may be associated with hematuria, renal calculi, hydronephrosis and recurrent urinary tract infections (UTI). The other abnormalities involving the ectopic kidney include cystic dysplasia, pelviureteric junction (PUJ) obstruction and vesicoureteral reflux (VUR). Anomalies involving genitourinary, skeletal and other organ systems can also be associated with crossed renal ectopia. The presence of crossed renal ectopia without fusion can be detected by ultrasonogrphy (USG), intravenous urography (IVU), renal scintigraphy, computed tomography (CT) and magnetic resonance imaging (MRI). Multidetetor row computed tomography (MDCT) angiography offers a cheap, non-invasive imaging modality providing exquisite anatomical details and excellent information about the anomaly and its associated conditions. We present here anatomical and radiological features of three cases of unfused type of crossed renal ectopia detected incidentally during MDCT angiography.

MATERIALS AND METHODS

The present study was carried out in a single diagnostic centre during the period from October, 2012 to July 2015. The study group includes 960 patients (491 males and 469 females, age range 4-90 years) who underwent contrast enhanced MDCT angiography for evaluation of suspected hepatobiliary, pancreatic, renal pathologies, abdominal pain and other abdominal pathologies. All the scans of the patients who had undergone previous abdominal surgery or were detected with any intra-abdominal pathology which is likely to distort the anatomy of the region concerned and poorly enhanced scans were excluded from the study. The imaging centre routinely obtains written informed consent from all the patients before contrast injection. All the patients underwent contrast enhanced computed tomography by a 64 channel scanner (GE Optima 660) and received 90-100 mL of non-ionic contrast (Omnipaque 300 mg I/mL) at the rate of 5 mL/s intravenously. Sections of 0.625 mm thickness were obtained from diaphragm to upper part of thigh and analysed in a separate work station (AW Volume share 4.5). After analysing axial, coronal and sagittal scans, volume rendered (VR), maximum intensity projections (MIP) and multiplanar reformatted images (MPR) were obtained.

OBSERVATIONS

Out of 960 patients examined, we detected 3 cases of crossed unfused renal ectopia (0.31%; 1 in 320 cases). All three patients were males and left- to-right renal ectopia was noted in 2 cases and right-to-left ectopia in one case.

Case 1: Fourteen year old male patient with vague abdominal pain and a palpable mass in the right iliac fossa was referred to our centre for MDCT evaluation which revealed left to right crossed ectopia without fusion. The left kidney was ectopic, lying in front of right common iliac artery well below the lower pole of right kidney with posterolaterally directed hilum (Figure-1, 3-A) It is located just to the right of midline anterior to L4, L5 vertebrae. (Figure-2) The left ureter passing behind the left kidney crosses the midline at the level of sacral promontory to enter the bladder on the left side (Figure-2). The main left renal artery arising from aortic bifurcation is seen entering the renal parenchyma after crossing its anterior surface. The accessory LRA

arises from right common iliac artery and passes posteriorly. The left renal vein emerging posteriorly ascends parallel and to the right of right common iliac artery to drain into inferior vena cava (IVC) (Figure-1, 3-B). The right renal vessels were normal.

Fig. 1: Volume rendered (VR) MDCT image of crossed renal ectopia without fusion in a 14 year old male patient showing arterial supply. **[A]** Right kidney (RK) is normally located and is supplied by a single right renal artery (RRA). The left kidney (LK) is crossed and ectopic lying anterior to right common iliac artery and malrotated with hilum facing posterolaterally. **[B]** The left kidney (LK) is supplied by two left renal arteries (LRA). The main LRA is arising from anterior aspect of aortic bifurcation and descends anterior to left kidney and enters anterior aspect of renal parenchyma. The accessory LRA arises from right common iliac artery and passes posteriorly. The left renal vein (LRV) emerges from the hilum and ascends posterior to left kidney and to the right of right common iliac artery to drain into inferior vena cava.

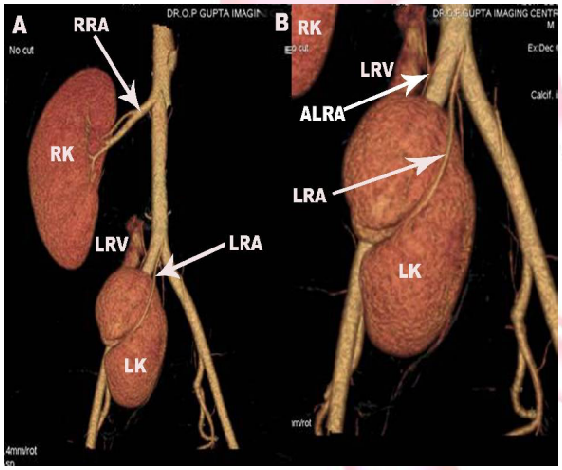


Fig. 2: MDCT urography of a 14 year old male patient showing orthotopic right kidney (RK) drained by right ureter (RU) opening into urinary bladder ipsilaterally. The crossed ectopic left kidney (LK) lies to the left of midline in front of lumbosacral junction with hilum facing posterolaterally. The left ureter (LU) is seen crossing the midline in front of sacral promontory to open on the left side of bladder.

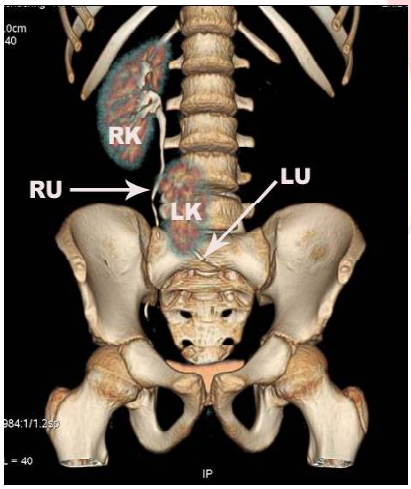


Fig. 3: MDCT sagittal **[A]** and axial **[B]** images of a 14 year old male patient. **[A]** Sagittal section showing crossed ectopia of the left kidney (LK) and normally located right kidney (RK). **[B]** Axial image showing posterolaterally directed hilum of left kidney (LK) with left renal vein (LRV) emerging. Note that the LK is lying anterior to right common iliac artery (RCIA). LCIA= left common iliac artery. LRA=main left renal artery. The accessory LRA is indicated by zig-zag arrow.

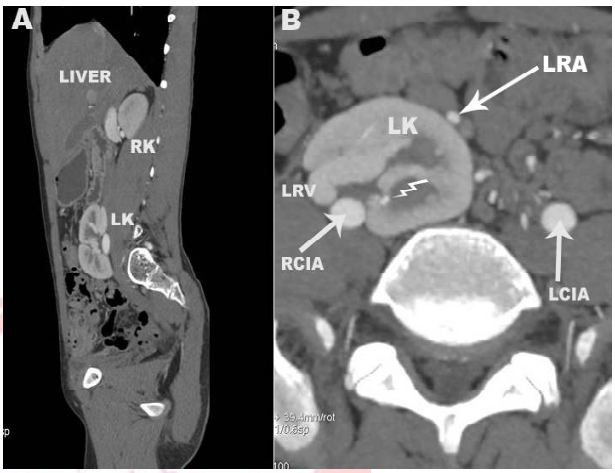
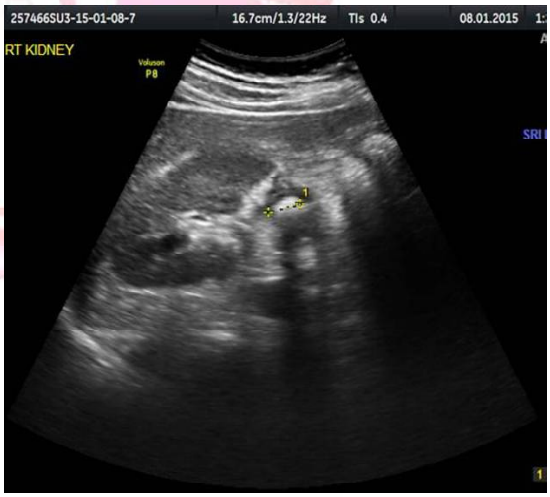


Fig. 4: USG of a 39 year old male patient showing right kidney with a calculus measuring 1.33 cm



Case 2: Male patient, 39 year old, with complaint of renal colic, underwent USG and subsequent MDCT evaluation. USG revealed a small stone (1.33 cm) in the normally placed right kidney (Figure-4). Crossed ectopic left kidney without fusion was situated below the right kidney anterior to right common iliac vessels. Right kidney was supplied by two arteries and the ectopic left kidney by four arteries, one from anterior aspect of aorta above its bifurcation, another from left common iliac artery and two small arteries entering the upper pole from right common iliac artery (Figure 5- A, B) The left ureter crossed the midline to open into urinary bladder on the left side (Figure-6 A, B).

Fig. 5: MDCT-VR images **[A]** anterior view and **[B]** posterior view of a 39 year old male patient showing arterial supply. The orthotopically located right kidney (RK) is supplied by two right renal arteries (R1, R2). R1 arises just below the origin of superior mesenteric artery and R2 arises opposite to origin of inferior mesenteric artery and both enter anteromedially directed hilum. The crossed ectopic left kidney (LK) is supplied by four left renal arteries, one originates from aorta just above its bifurcation (L1) and enters the upper pole; another (L2) arises from left common iliac artery (LCIA) which crosses the midline to enter anteriorly placed hilum. Two small left renal arteries (*) originate from right common iliac artery (RCIA) to enter the upper pole.

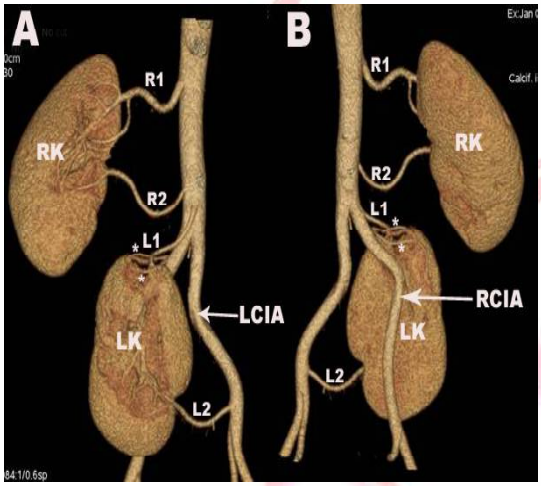
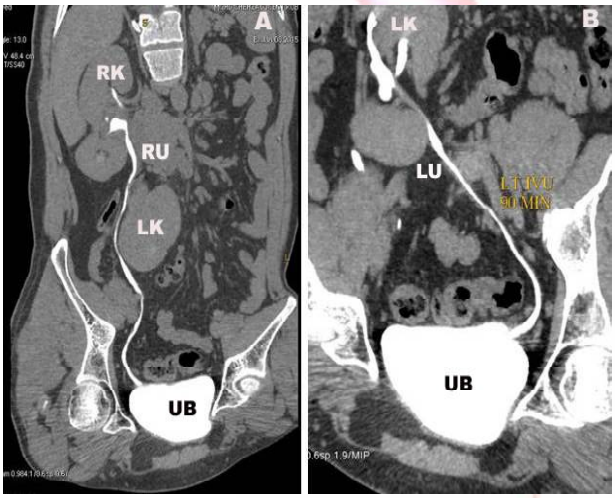
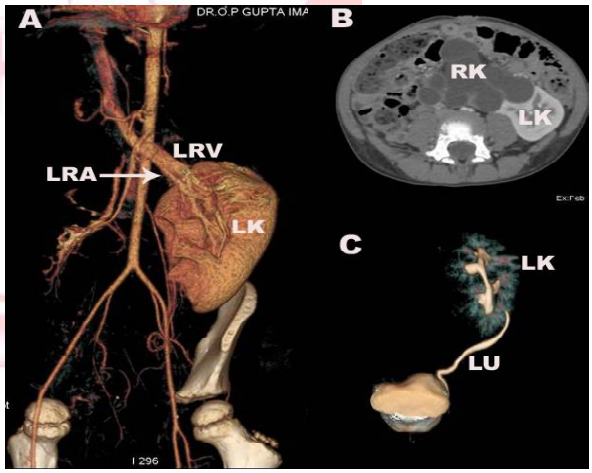


Fig. 6: MDCT oblique coronal multiplanar reformatted images of a 39 year old male patient. **[A]** Shows normally positioned right kidney (RK) drained by right ureter (RU) which passes to the right of ectopic left kidney to drain ipsilaterally into urinary bladder (UB).**[B]** Shows the left ureter (LU) from the ectopic left kidney (LK) crossing the midline to open on the left side of bladder (UB)



Case 3: A 4 year old boy with a palpable mass underwent MDCT evaluation. The left kidney was placed at a slightly lower level with left renal artery having a downward course. The left renal vein was having a normal preaortic course to drain into IVC.(Figure-7A) The right kidney was totally dysplastic having multiple cysts and was placed in the midline anterior to distal aorta and its bifurcation.(Figure-7B) Some cysts of the right kidney protrude into left kidney but both kidneys have their own distinct renal fascia. Since the right kidney was non-functional, the right ureter could not be visualized. Left ureter was opening into bladder normally.(Figure-7C) Because the right kidney was ectopic and was not fusing with the left kidney, we consider it as crossed ectopia without fusion.

Fig. 7: **[A]** VR image of a 4 year old boy showing left kidney (LK) positioned at a lower level with left renal artery (LRA) from aorta entering anteriorly placed hilum and left renal vein (LRV) having its normal preaortic course. **[B]** Axial image showing dysplastic ectopic right kidney (RK) positioned in the midline anterior to aortic bifurcation, protruding into but not fusing with left kidney (LK) **[C]** MDCT urography showing left ureter (LU) draining into urinary bladder ipsilaterally. Right ureter is not visualized because the dysplastic right kidney is non-functional.



DISCUSSION

Crossed renal ectopia is a rare congenital anomaly in which one kidney is transpositioned to the contralateral side with its associated ureter opening into the urinary bladder in its normal position after crossing the midline. In this condition both the kidneys are located on one side of the midline. In 90% cases the crossed ectopic kidney fuses with the normally positioned kidney and in the rest it may remain unfused [1]. The crossed renal ectopia may remain asymptomatic, being detected incidentally at autopsy, surgery and during screening procedures for unrelated conditions. In crossed renal ectopia without fusion the ectopic kidney generally lies at a distance inferior to its orthotopic mate, oriented horizontally or

diagonally with its own Gerota's fascia. Since ectopic kidney is also malrotated, the position of hilum is abnormal. The left kidney crossing to the right side is three times more common and males are affected more than the females in a ratio of 2:1[1]. On the contrary, it is noted that crossed renal ectopia without fusion is twice as common on the right as on the left and is more frequent in male than female patients (1.4:1) [3,4,5]. We have reported here unfused crossed renal ectopia in three male patients, two cases of left-to-right ectopia and one case of right-to-left ectopia.

Table 1: Crossed Renal Ectopia without Fusion: Case Reports.

AUTHOR	SEX	AGE	MODALITY	TYPE OF ECTOPIA	SYMPTOMS	ASSOCIATED ANOMALIES	REMARKS
Winram 1959 [6]	M	46	Retrograde pyelography, Aortography	Right-Left	Hematuria; RK- non functioning		Right nephrectomy
Miyakita et al 1985 [7]	F	26	IVP, Retrograde Pyelography, aortography	Not reported	No complaints	Infertility	Hydrosalphinx
Miyakita et al 1985 [7]	M	28	IVP, Retrograde Pyelography, aortography	Not reported	No Complaints	Hypospadias	
Virmani & Pathania, 1987 [8]	M`	28	IVP	Left-Right			
Al Mugueiren 1997 [9]	F	2	USG,IVP,CT	Right-Left	LK-malrotated RK- multicystic - Dyspalsia		RK nephrectomy
Mansberg et al 1999 [4]	M	10	USG, Retrograde Pyelography, Scintigraphy	Left-Right	Non-functioning LK	Ectopic LU insertion into prostatic utricle	Left Nephrectomy
Yano et al 2003 [10]	M	63	CT, Aortography, Scintigraphy	Left-Right		Abdominal aortic aneurysm	Aneurysemectomy
Nursal, 2005 [5]	M	4	USG, Scintigraphy	Right-Left	Recurrent UTI	Hydrocephalus, Meningomyelocele, scoliosis, right pes equinovarus	
Nursal, 2005 [5]	M	10	USG, Scintigraphy	Left-Right		Anal atresia	
Belekar et al 2008 [11]	M	55	USG,IVU, DTPA scan	Left-Right	LK- calculi, Hydronephrosis non functioning		Left nephrectomy
Narci et al. 2010 [12]	M	7 mo.	USG,MRI,DMSA, DTPA scans, Pyelogaphy	Left-Right	RK- ureterocele Hydronephrosis multicystic dysplasia,		Left nephroureterectomy & Right excision of ureterocele
Jorwekar et al 2011 [13]	F	8	CT, IVU, DTPA scan	Left- Right	Hydronephrosis, non-functioning LK		Nephrectomy
Ramaema et al 2012 [14]	M	16	CT	Left-Right	Acute abdominal pain		
Lodh et al 2013 [15]	M	7	IVP, DTPA scan	Left-Right	Hydronephrosis of LK	Absent left testis	Left Nephrectomy
Khandelwal et al 2013 [16]	M	50	USG, IVU, CT, MCU, Cystoscopy	Left-Right	LK- Dilated pelvicalyces & Calculus. Malrotated RK		
Akcimen et al 2014 [17]	M	64	USG, CT	Left-Right	Left side flank pain		Nephrolithiasis
Prasad & Thomas, 2014 [18]	M	45	CT	Left-Right	LK- hilum facing posteriorly		Mass in stomach-GIST
Jindal et al 2014 [19]	M	40	USG, IVP, CT	Left-Right	LK- hydronephrosis		
Rodrigues et al 2014 [20]	F	65	IVU, USG	Left-Right	Abdominal pain Abdominal mass		
Murathati et al 2015 [21]	M	8	USG, CT	Left-Right	Hematuria Right flank pain		Grade 3 renal injury due to blunt trauma
Ratola et al 2015 [22]	F	11	USG, MRI	Left-Right	Abdominal pain, anorexia.		

Total number of cases – 21; Males- 16; Females- 5; Left- to-Right ectopia- 16 cases; Right-to- Left – 3 cases; Not reported-2

Because of its rarity, CRE without fusion is sporadically reported in the literature and only 62 cases have been reported till1959 [1, 6]. We could collect 21 case reports of this anomaly reported in the literature till date and the observations of these case reports are summarized in Table-1. Analysis of these case reports indicate that this anomaly is four times more common in males than in females (16 males and 5 females; Age range- 7 months to 65 years) and left-to-right ectopia was seen in 16 cases and right-to left ectopia in 3 cases only. Our observations also suggest that this anomaly is more common in males and affects left kidney more frequently. Most common presenting symptoms observed in these case reports are abdominal pain, hydronephrosis, hematuria and calculi. (Table 1) Urogenital anomalies observed with CRE without fusion include hypospadias, infertility, anorchia and ectopic ureteric opening. [4, 7,15]. Anal atresia was present in a boy and in another, more severe anomalies like hydrocephalus, scoliosis, meningomyelocele and pes equinovarus were seen.[5] Multicystic dysplasia was also observed in two cases similar to our case [9, 12] Abdominal aortic aneurysm was reported in one case.[10]

Renal ectopia occurs due to defects in the normal process of ascent of the kidney which also affects the axial rotation resulting in the anomalous position of the hilum. In one of our cases the hilum was facing posterolaterally (Figure-3 B) and in another anteriorly.(Figure-5A) In the third case the position of the hilum could not be assessed because of the severe dysplasia affecting the ectopic right kidney. Because of a lower position, ectopic kidneys receive arterial supply from distal aorta close to its bifurcation, common iliac, internal iliac, inferior mesenteric or median sacral arteries. In two of our cases exhibiting left-to-right ectopia, multiple renal arteries arising from aortic bifurcation and common iliac arteries were supplying the ectopic kidneys. An interesting observation was that in both the cases the ectopic left kidney was supplied by branches from the right common iliac artery.(Figure-1 and 5) Ectopic origin of main renal artery or accessory renal arteries from contralateral side is rarely reported [23]. Origin of main or accessory renal arteries from common iliac arteries supplying an ectopic kidney has important surgical implications during abdominal aortic aneurysmal repair, pelvic surgeries, retroperitoneal lymph node dissection and endovascular repair.

The exact incidence of crossed renal ectopia without fusion in general population is unknown. It is estimated as 1 in 75000 autopsies [3]. Estimating the prevalence of renal ectopia in 12000 patients investigated by USG, Asghar and Wazir reported crossed renal ectopia without fusion in 2 cases only (0.01%) and crossed fused renal ectopia in 5 cases (0.04%) only [24]. The true incidence cannot be estimated because many cases remain asymptomatic and undiagnosed throughout life. The present study detected 3 cases out of 960 cases (0.31%) which is higher than that reported in the literature. This could be due to use of MDCT for evaluation which is considered as a more sensitive imaging modality. By using MDCT urography we have reported a higher incidence of horseshoe kidney (1.02%; 7 out of 682 cases) and crossed fused renal ectopia (3 out of 682 cases) [25,26]. Alternately the higher incidence could be due to racial differences in congenital renal anomalies.

The kidneys developing in the sacral region, undergo a complex process of ascent and rotation to reach their final destination in the renal fossae. Crossing over can occur during the cephalad migration resulting in the positional and rotational abnormality. Because the mechanisms responsible for normal ascent of the kidney during gestation are unknown, the cause of crossed ectopia is also not understood well and several theories have been put forward to explain this phenomenon. These include the classical mechanical theory (abnormally placed umbilical arteies mechanically obstructing the normal ascent), the ureteral theory (ureteric bud wandering to the opposite side), theory of abnormal rotation of caudal end of the fetus (causing displacement of nephrogenic anlagen to the contralateral side), the teratogenic theory and the genetic theory [1, 25].

It is also suggested that poor development of a metanephric blastema with a defect in the kidney tissue responsible for prompting the

kidney to ascend to its usual position could be a factor causing this anomaly [17]

CONCLUSION

Crossed renal ectopia without fusion is a very rare anomaly sporadically reported in the literature mainly as case reports. In many cases the anomaly remains asymptomatic and detected incidentally at autopsy, surgery or radiological investigations for unrelated problems. We have presented here three cases detected incidentally during review of MDCT scans of 960 patients. To the best of our knowledge the present study is the first such study to detect crossed renal ectopia without fusion in a large series of patients. MDCT angiography is a sensitive imaging modality to detect such renal anomalies.

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Conflicts of Interests: None

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