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Radiology

PROBLEM OF FUSED KIDNEYS - DIAGNOSED WITH MDCT: OUR SINGLE CENTER EXPERIENCE

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ABSTRACT

Introduction: Multi-detector CT scan (MDCT) can precisely detect the presence of fusion anomaly and its associations. Our retrospective cross sectional study aims to analyze the morphological appearances of renal fusion anomalies; assessment of frequency, clinical presentation and clinical significance of associated conditions with MDCT to provide valuable information for proper management and aid in surgery; if indicated.

Methods and Material: Out 3040 patients who underwent CT abdomen at our centre from 1st January 2014 to 31th March 2017; about 56 of them had renal fusion anomalies. Type of Fusion anomaly was described along with its anatomy and other associated abnormalities depending upon CT scan findings. Possible complications and management were discussed and literature was reviewed.

Results: Out of these 56 patients; 40 patients showed horse shoe kidney and 14 patients showed crossed renal ectopia. However one patient showed crossed ectopia without fusion and two pan cake kidneys were noted. About 42 patients showed various complications like calculus, hydronephrosis and pelvi-ureteric junction obstruction (PUJO). Remaining 7 patients showed associated abnormalities like small contracted kidney with ureterocele, ureteric stricture, renal cysts, renal mass and bladder mass lesions. Two renal masses were seen in Horse shoe kidney and pan cake kidney each. Bladder mass lesion was seen in horse shoe kidney patient.

Conclusions: MDCT urography provides better evaluation of such anomalies in a single examination. Three-dimensional reformatted images can provide particularly good Delineation of such anomaly along with its vascular supply information; which is very important to the surgeon.

KEYWORDS : Fusion Anomalies, Horse Shoe Kidney, Crossed Fused Ectopic Kidney, CT Scan

Introduction

About 30-40% developmental anomalies are seen in genito-urinary system [1]. Common partial fusion anomalies include Horse-shoe kidney and crossed renal ectopia. Complete fusion is seen in pan cake kidney.

As most of such anomalies are asymptomatic, most of them remain undetected hence the true incidence of this anomaly remains unknown. Fusion anomalies are also predisposed to various complications as hydronephrosis, infection, calculus and neoplasm. As fusion anomalies frequently are associated with mal- rotation, abnormal position or abnormal blood supply; their thorough knowledge is very important to the operating doctor when needed.

Material and Method

Our retrospective cross-sectional study comprised of 3040 patients diagnosed with renal fusion anomalies on CT scan at Department of Radio-diagnosis, Institute of Kidney Disease and Research Centre, Dr H L Trivedi Institute of transplantation Sciences, Ahmedabad, Gujarat. All of them underwent CT scan on Seimens Somatom 64 Slices CT scanner with Injection of 350 ml Iohexol in 60 mg/kg dose with prior written consent. Rapid injection of 70 – 100 ml bolus of 300-400mg/ml contrast at the rate of 3.5 mL/sec followed by 20 ml saline at rate of 2.8 mL/sec was infused. The region included was from dome of diaphragm up to termination of common iliac arteries. Slice thickness was 5 mm and the scans were reconstructed at 0.6 mm thickness. Arterial, venous and delayed phases were obtained at 10 seconds, 60 seconds and 7 to 10 minutes respectively. Non enhanced images were examined for presence and position of kidney; type of fusion anomalies and any calculus. Contrast enhanced images were used to confirm the type of fusion anomaly, presence of any associated parenchymal abnormality. Excretory phases were used to comment upon the status of PC system. Arterial and venous supply are also examined in a Multi planar and curved planar reformations (MPR and CPR), maximum intensity projection (MIP), and volume rendering (VR) techniques were used for post processing of images. Both renal veins and IVC were examined in terms of their drainage patterns and presence of anatomical variations, if present.

Results

Out of 3040 patients studied; 56 patients were found to have fusion anomalies. The age varied from 3 years to 72 years (mean age: 33.93 ± 18.71 years). Out of 56, 34 were males and 22 were females. About 40 (24 males and 16 females) of them showed Horse shoe kidney and 15 (9 males and 6 females) showed crossed renal ectopia. However one patient showed crossed ectopia without fusion. Only two cases of pan cake kidney were noted.

About 8 out of 56 patients showed no evidence of complications and 42 patients showed various complications like calculus, hydronephrosis and pelvi-ureteric junction obstruction (PUJO). Remaining 7 patients showed associated abnormalities like small contracted kidney with ureterocele, ureteric stricture, renal cysts, renal mass and bladder mass lesions. Two renal masses were seen in Horse shoe kidney and pan cake kidney each. Bladder mass lesion was seen in horse shoe kidney patient.

According to our study; prevalence rate of Fusion anomalies was 56/3040 (1.84%) which was more in males (60%). The anomalies were detected in a varied age group depending upon the time of presentation. The incidence of Horse shoe kidney according to our study was 71.43 % and that of crossed renal ectopia was 25 % in total anomalies and 1.32% and 0.46% respectively in total population [Table 1].

Table 1: Table showing number incidence of various fusion anomalies in total population as Well as in total fusion anomalies

	N=3040 patients	N= 56 patients
HSK	40 (1.32%)	40 (71.43%)
CRE	14 (0.46%)	14 (25%)
Pan cake	2 (0.07%)	2 (3.57%)

Pan cake kidney which is very rare anomaly showed about 3.57% incidence rate. Out of 14 crossed fused kidneys; 10 were left crossed fused ectopia and 4 of them were right crossed fused ectopia. Incidence of complications was about 92.5 % in patients with horse shoe kidney; 71.43% in that of crossed renal ectopia and the pan cake fusion showed complication in both the cases [Table 2].

Table 2: Table showing incidence of various fusion anomalies along with their

Type of fusion	Male	Female	Age (years)	Complications
HSK	24 (60%)	16 (40%)	36.70±19.66	37 (92.5%)
CRE	8 (57.14%)	6 (42.8%)	28.64±14.63	10 (66.67%)
Pan cake	2 (3.57%)	0 (0%)	15.50±3.54	2 (100%)

Complications

Variety of complications was seen within the observed kidneys with complications like Calculus formation, Hydronephrosis, PUJ obstruction and mass lesions [Table 3].

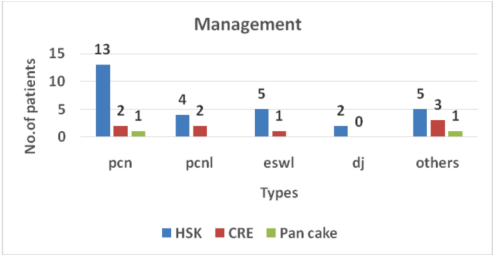
Table 3 : Table showing various complications in various types of fusion anomalies

Complications	HSK (N=40)	CRE (N=14)	Pan cake (N=2)
No	5 (12.5%)	6 (42.86%)	-
HN	7 (17.5%)	3 (21.43%)	-
Cal	23 (57.7%)	5 (35.71%)	1 (50%)
PUJO	3 (7.5%)	-	-
Mass	2 (5%)	-	1 (50%)

Various Complications were offered treatments like Per cutaneous nephrostomy, per cutaneous lithotomy, ESWL, DJ stenting and other according to the presence of complications. About 11 patients with HSK and 6 patients with CRE needed no treatment in absence of complications [Table 4].

Table 4: table and graph showing various management options used in fusion anomalies

Management	HSK (N=40)	CRE (N=14)	Pan cake (N=2)
Pcn	13 (32.5%)	2 (13.33%)	1 (50%)
Pcnl	4 (10%)	2 (13.33%)	-
Eswl	5 (12.5%)	1 (6.67%)	-
Dj	2 (5%)	-	-
others	5 (12.5%)	3 (21.43%)	1 (50%)
NA	11 (27.5%)	6 (40%)	-



Discussion

Various complicated processes like ascent, lateral migration, axial deflection, and internal rotation are responsible for renal development. Fused kidneys are usually seen in ectopic position as they are restrained from ascent. Fusion of kidneys is usually associated with abnormal vascular supply [2].

Fusion anomalies of kidneys can be classified in partial or complete fusion. Horse-shoe kidney (HSK) and Crossed fused renal ectopia(CRE) are included in partial fusion and pan cake kidney is considered as complete fusion anomaly. Crossed fused ectopia is further sub classified in six subtypes according to the type of fusion.

1. Horse Shoe Kidney

It is the most common renal fusion anomaly; seen in 90% of all renal fusion anomaly [5]. It shows two distinct renal masses lying vertically on either side of midline with their lower poles connected by isthmus of renal parenchyma or fibrous band that crossing the midline of the body; giving the whole structure the shape of a 'Horse-shoe". Rarely, the upper poles may be fused in horseshoe kidney [2]. In 1564, Botallo described and illustrated the first Horseshoe kidney. In 1820, Morgangi described the first diseased horseshoe kidney [6].

Its incidence is estimated to be 0.25% of all population [7]. As it is asymptomatic, can be incidentally found or can present with any complication in any age group. Males are more commonly affected in a ratio of 2:1. Definite genetic predisposition is not proposed.

Description:

In most of the cases (95%); lower poles are fused. Usually the fused segment is functioning renal parenchyma with its own blood supply and thin fibrous band occasionally. The fused segment is seen at the level of L3-L4 vertebral body just below the origin of inferior mesenteric artery in pre-aortic location. Rarely, it is retroaortic in location or runs between the great vessels [2].

The calyces face medially due to malrotation and the axis of renal pelvis is seen in vertical or oblique lateral plane. Lower calyx may extend more medially to facilitate drainage of isthmus. The laterally lying ureter commencing high on pelvis; while crossing the isthmus, they drain in to bladder via their normal openings or ectopic site rarely [2].

Blood supply:

While lying in pelvis each of metanephric blastema acquires blood supply from distal part of abdominal aorta, common iliac, internal iliac and median sacral arteries. During ascent, the blood supply changes as the new arteries generate cranially and old arteries degenerate caudally. Due to prevention of ascent of horse shoe kidneys, additional and ectopic renal arteries are seen in HSK. Each of kidneys consists of one renal arterial supply in 30% cases. Three types of Horse shoe kidneys have been identified according to the blood supply according to the Papin's autopsy study including 139 horse shoe kidneys.

Group I consisting of 20% patients shows normal renal vascular anatomy.

Group II includes 66 % of them showing three to five renal arteries.

Group III consists of only 14 % patients where more than five renal arteries are seen [8].

Various other classifications are also proposed depending upon the level of origin of renal arteries.

In type I the renal artery arises from normal level.

In type II two normal renal arteries and one or more accessory renal arteries arising from distal aorta or iliac arteries are seen.

In type III all renal arteries arise from ectopic origin.

Graves studied the patterns of blood supply of Horse shoe kidney with the means of resin cast and described six types of kidneys depending upon the blood supply [Figure 1].

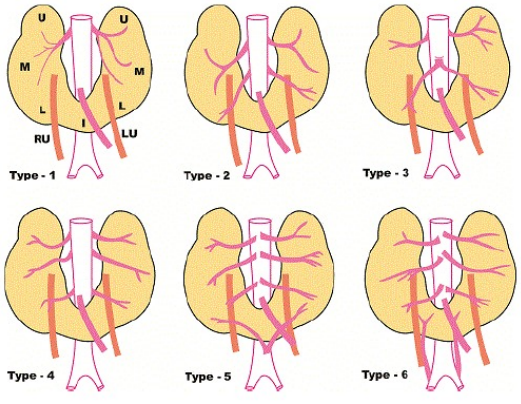


Figure 1: Grave's Classification of six arterial patterns of horse shoe kidney [9].

U,M,L- upper, middle and lower segments
I - Isthmus, RU – Right ureter, LU _ Left ureter

In type I may be normal with single artery supplying upper, middle and lower segments.

Type II is defined where upper and middle segments of each kidney are supplied by single artery and lower segment is supplied by a branch from aorta.

Type III is when the arteries to lower segments arise through a common trunk from aorta.

Type IV is classified when all three segmental arteries are separately arising from aorta.

Type V includes the variations in blood supply in isthmus where the arteries may arise above or below the isthmus which may be unilateral or bilateral and may arise independently from aorta or through a common trunk.

In type VI; the fused segment may be supplied by a vessel from common iliac artery commonly and internal iliac artery or median sacral artery rarely [9].

Venous drainage:

Patients having HSK also show variations in venous drainage. A study on 105 patients with Horse shoe kidney; renal vein anomalies were reported in 24 patients (22.9%) including 15 dual right RVs, 1 triple right RV, 5 dual left RVs, 4 circum-aortic and 1 retro-aortic left RVs. Two patients had both circum-aortic left RVs and double right RVs [30] Anomalies of IVC (3.9% - 5.7%) were found significantly more frequently in patients with Horse shoe kidney than those without it [10,11,12]. These IVC anomalies included pre-isthmic IVC, dual IVC, left IVC and azygos continuation of IVC. Such vascular anomalies associated with horse shoe kidney grant technical difficulties during surgery. Significantly higher incidence of anomalous SVC was also reported in such patients (4.2%) than otherwise (0.22%) [13]. Thorough knowledge of vascular anomalies is needed for proper diagnosis of images which can be confused with retroperitoneal masses or lymphadenopathy and to prevent complications.

Associated anomalies:

Horse shoe kidney is frequently associated with congenital anomalies in other systems like, skeletal (hemi vertebrae with scoliosis, rib defects, clubfoot, congenital hip dislocation), cardiovascular system (ventriculo-septal defects), gastrointestinal system (anorectal malformation, mal-rotation, and Meckel's diverticulum), and central nervous system (neural tube defects) [14]. 20% of patients with trisomy 18 and 60% of them with Turner's syndrome are associated with horse shoe kidney. 4 % of males with horse shoe kidneys show hypospadias and undescended and 7% of females show bicornuate uterus or septate vagina. Duplication of ureter (10%) and vesico-ureteric reflux (>50%) are also seen in patients with horseshoe kidney.

Clinical presentation:

Due to abnormal course, the ureters are prone to obstruction which precipitates hydronephrosis, infection and calculus formation [2, 15]. As almost one third of patients with horse shoe kidney are asymptomatic; it is incidentally found on radiological imaging. But when symptomatic, vague abdominal pain radiating to lower lumbar region is seen. Rarely may it present with characteristic "Rovsing sign" showing abdominal pain, nausea and vomiting on hyperextension of spine. Urinary tract infection is most common presentation in children. We found about 40 patients with horse shoe kidney out of which 5 patients had no complications; 7 patients (17.5%) showed presence of hydronephrosis; 23 patients (57.7%) had calculi formation and 3 of them showed evidence of Pelvi ureteric junction obstruction (7.5%) [Table 3]. Two patients with Horse shoe kidney showed presence of Mass lesion (5%). [Figure 2].

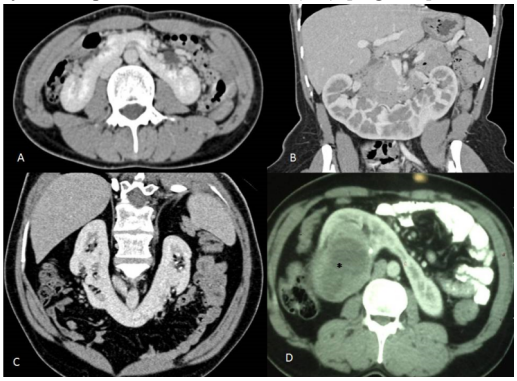


Figure 2: (A) Axial (B) coronal scans of CECT abdomen showing horse shoe kidneys
(C) Reformatted Coronal image of CECT abdomen showing horse shoe kidneys with multiple small calculi in all calyces of both kidneys.
(D) Axial contrast enhanced CT scan showing minimally enhancing mass lesion in Horse shoe kidney

Complications:

1. PUJ Obstruction:

1/3rd of patients with horse shoe kidney show uretero pelvic junction obstruction causing significant hydronephrosis [2].

2. Renal Calculi:

Concomitant formation of calculus (20 to 80% cases) and PUJ obstruction is seen [16,17].There is increased risk of development of xanthogranulomatous pyelonephritis in horseshoe kidney with staghorn calculus.

3. Infection:

30% of patients with horse shoe kidney present with signs and symptoms of urinary tract infections. The most common route of infection is ascending infection secondary to vesico-ureteric reflux (VUR). VUR is seen in 50% of Horse shoe kidney patients; it is diagnosed with the help of Micturating urethrogram (MCU). Renal infection in diabetic patients can lead to emphysematous pyelonephritis.

4. Renal Neoplasms:

HSK has increased risk for Wilm's tumor, renal carcinoids and transitional cell carcinoma. Isthmus is more commonly affected. RCC is the most commoly seen malignant tumor in horseshoe kidney (45% of all tumors). However there is no increased risk of RCC in patients with HSK than in general population. But there is a 3-4-fold increase in the incidence of TCC in horseshoe kidney, which accounts for approximately 28% of all tumors. Though carcinoid tumors are rare in horseshoe kidney, there is 62-fold higher risk than that in the general population. Rare malignant tumours like Squamous cell carcinoma and benign tumors such as oncocytoma and angiomyolipoma also have an increased risk in horseshoe kidney [18].

1. Crossed Fused Ectopic kidney

It is the second most common fusion anomaly of kidneys. The estimated incidence of crossed fused renal ectopia is approximately 1:1300–1:7500 [19]. It is fusion as well as ectopic anomaly occurring in about 0.08% - 0.01% cases. The crossed ectopic kidney crosses the mid line and fuses with its ipsilateral mate in about 90 % cases.

Its prevalence is estimated to be 1 in 1000 live births [4]. Halaseh M reported only 7 cases (1.75%) of CRE in a review of 400 children evaluated by DMSA renal scan [20]. In a retrospective study done by Glodny B et al, they reported the incidence of CRE as 1 out of 3078 CT scans [21]. Although the true incidence of this anomaly is not known as a large number of asymptomatic patients with CRE go undetected.

The male-to-female incidence ratio estimated is 2:1 and it is three times more common on the left side [22,23].

Description:

The crossed ectopic kidney crosses the mid line and fuses with its ipsilateral mate in about 90 % cases. The ureter from normal kidney follows normal course and enters bladder via normal opening, whereas that of ectopic kidney crosses the midline and enters the bladder via its normal opening on the contralateral side. Various types of CRE are identified according to the type of fusion. Mc Donald and Mc Clellan identified four types of CRE: crossed renal ectopia with fusion (85%), crossed renal ectopia without fusion (10%), solitary crossed renal ectopia, and bilaterally crossed renal ectopia[24]. Six variations of CRE have been described: inferior crossed fused ectopia; sigmoid or S-shaped kidney; unilateral lump kidney; L-shaped or tandem kidney; unilateral disc kidney; superior crossed fused ectopia [2] [Figure 3].

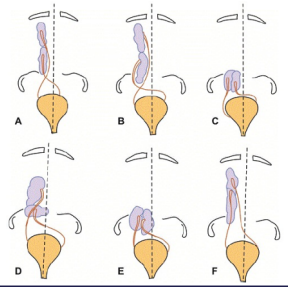


Figure 3 : Six subtypes of crossed fused renal ectopia [9].
A- Inferior ectopia type
B- Sigmoid or S-shaped kidney
C- Lump kidney with fusion of two kidneys
D- L-shaped or Tandem kidney
E- Disc kidney with extensive fusion of two kidneys
F- Superior ectopia type

Inferior crossed fused ectopia: The upper pole of the crossed kidney is attached to the inferior pole of the normal kidney. Both the renal pelvis may face anteriorly.

Sigmoid or S-shaped kidney: The crossed kidney lies inferiorly with its pelvis facing laterally and the normal kidney lies superiorly with its pelvis facing medially thus each pelvis is oriented correctly.

Unilateral Lump kidney: Here, the fusion occurs in a wide renal region hence the total kidney mass is irregular and lobulated and the ascent is prevented beyond the sacral promontory. Both renal pelvis draining separate areas of renal parenchyma face anteriorly.

L-shaped or tandem kidney: Crossed kidney lies in a transverse and inferior plane fusing with the lower pole of normal kidney and assuming the shape of "L" in front of L4 vertebra to lie in the midline or contra lateral para median space.

Unilateral disc kidney: In this type, kidneys are fused at medial aspect along their entire length. The pelvis faces anteriorly and ureters coarse along their normal pathway to drain in to bladder via their normal openings. Each half of the kidney has its own collecting system drains and does not communicate with the opposite half.

Superior crossed fused kidney: it is least common variant. Here the lower pole of ectopic kidney fuses with the upper pole of normally positioned kidney and the ureter crosses the midline to enter in bladder via its normal opening.

We found about 10 patients (71.43%) with left crossed renal ectopia[Figure 4] and 4 patients (28.57%) with right crossed renal ectopia [Figure 4]. One of the left crossed renal ectopia showed the superior ectopia; which is very rare variant, both of the PC system and ureters were dilated [Figure 5].

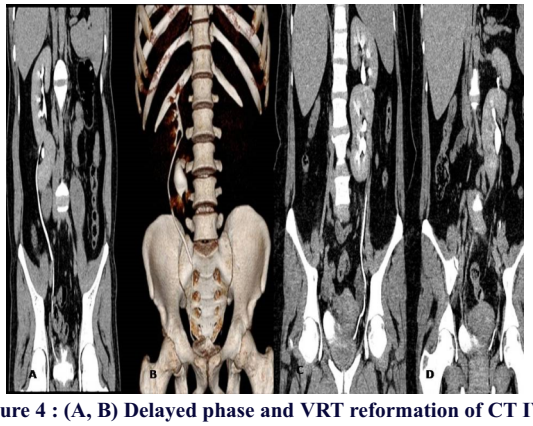


Figure 4 : (A, B) Delayed phase and VRT reformation of CT IVP showing left crossed fused ectopia(S type). (C,D) Delayed phase of CT IVP showing right crossed fused ectopia

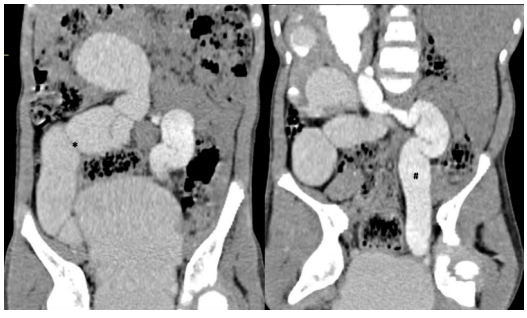


Figure 5: Delayed phase of CT IVP showing left crossed fused ectopia. Right kidney shows hydronephrosis with hydroureter (*) up to lower end and ectopic left kidney shows hydronephrosis with hydroureter(#) which crossed the mid line and drains in to bladder via left VUJ. Suggestive of left crossed ectopia with superior fusion

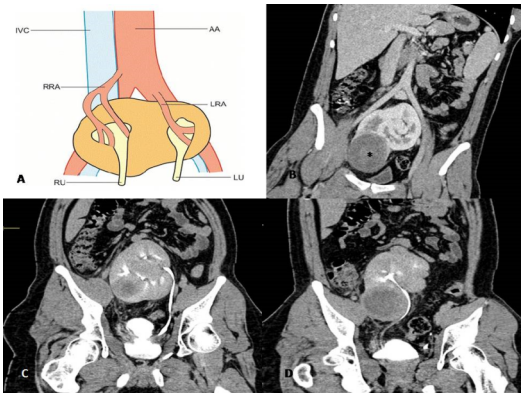


Figure 6 : (A) Schematic diagram of pancake kidney. Note the position of renal pelvis on the anterior aspect and the separate openings of both ureters [9].

(B,C,D)Venous and delayed phase of CT IVP showing minimally enhancing mass lesion(*) in pan cake kidney. Note two separate uncrossed ureters. The right uncrossed ureter is displaced by the mass lesion

Blood supply:
Blood supply to the ectopic kidney arises from the ipsilateral vessels but occasionally arise from contra lateral side. The renal artery arises from upper abdominal aorta

in 25% cases and from lower part of aorta or iliac arteries in 75 % cases. Total 1 to 6 renal arteries may be seen in two kidneys while 2 to 4 major arteries are present at various levels [2, 25].

Associated anomalies:
Frequently associated anomalies are: Imperforate anus (4%), orthopedic anomalies- mostly involving bony pelvis and vertebrae (4%), septal cardiovascular anomalies, Gastrointestinal and other genitourinary abnormality. Abnormality

is commonly seen in the ectopic kidney which includes PUJ Obstruction, cystic dysplasia and carcinoma [2,26]. Vaginal agenesis, VACTERL association, TAR syndrome, renal dysplasia and single ureter are also proposed to be associated to fused CRE. Intestinal mal-rotation association with a lump kidney in a male cadaver was reported by Kulkarni et al [27]. Thyagaraju K et al reported CRE with left sided polydactyly in a 24 week aborted male fetus [28].

Clinical presentation:
Usually the patients are asymptomatic, but when present, the symptoms are abdominal or flank pain, a palpable abdominal mass, hematuria, dysuria and other symptoms due to urinary tract infection. The abnormal position of kidney and aberrant vascular structures leads to poor drainage and stasis which can predispose to hydronephrosis, calculus formation and infection. Other urological complications seen with CRE are PUJ obstruction, reflux, and ectopic ureteroceles [2]. Sporadic cases of mass lesions in CRE have been reported [29].

Complications:
The complications and the mechanisms behind their occurrence remain more or less same in all fusion anomalies. Obstruction, calculi, malignancy and cystic dysplasia are various complications associated with CRE. Malignancy occurring in CRE is rare. Only 7 cases of malignancy in CRE have been reported. All of them were renal cell carcinoma except for one case of transitional cell carcinoma [11]. We found about 6 patients without complications. About 3 patients (21.43%) had presence of hydronephrosis and 5 of them (35.71%) showed presence of calculi [Table 3].

1. Pan Cake Kidney
Pancake kidney is a complete type of rare renal fusion. Glenn described the pancake or fused pelvic kidney when the entire renal

substance is fused into one mass which lies in the pelvis, and has two separate ureters which do not cross midline and enter in to bladder via their normal openings [Figure 3]. It accounts of only 2% cases of all fused kidneys [30]. The incidence is estimated to be 1/65000 to 1/375000 cases [12].

Only rarely a single ureter drains the pan cake kidney. It may sometimes be called 'lump' or 'disc' kidney; which are also subtypes of CRE. However, in such cases CRE kidneys can be differentiated from pancake kidneys as they show unilateral fused kidneys in lumbar or iliac regions and midline crossing of ureter from the ectopic kidney.

Pancake kidney is more common in males with ratio of about 2-3:1[10]. In pan cake kidney; pelvis faces anteriorly and ureters are short and do not cross the midline to open separately into urinary bladder

Blood supply:
Usually Pancake kidney retains its primitive blood supply and is supplied by *s*ingle renal artery which arises from distal aorta or common iliac artery and is drained by a single renal vein draining in to distal IVC or common iliac veins [9].

Description:
In pancake kidneys, the kidneys are fused at medial borders along its entire length and the lateral surface appears normal in contour. It is mal rotated and the renal pelvis faces anteriorly with uncrossed ureters. Each PC system drains respective half of kidney and never communicate with the contra lateral PC system.

Associated anomalies:
Other associated anomalies with pancake kidney are abnormal testicular descent, tetralogy of Fallot, absent vagina, sacral agenesis, caudal regression syndrome, spina bifida and anal abnormalities.

Clinical presentation:
The cake kidney may remain asymptomatic and may be detected at autopsy. But may get infected and present with changes of urinary tract infection, fever and vague lower abdominal pain. Sometimes it may manifest as extra renal symptoms as amenorrhoea, failure to conceive or iliac vessel aneurysm. It may cause localized pain due to the weight of renal mass causing stretching of renal vessels. We found two pan cake kidneys; both of them showed presence of complications. One of them showed calculus and other Showed presence of mass lesion [Figure 6].

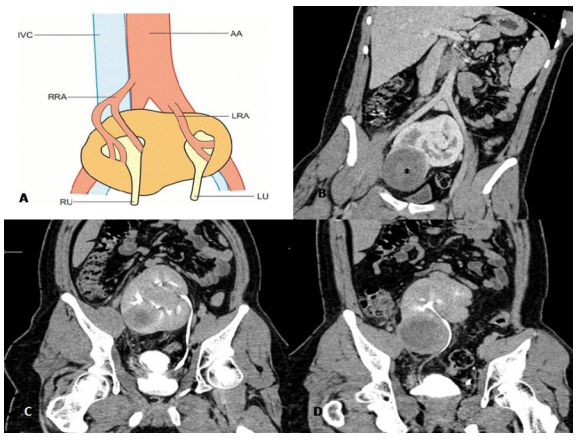


Figure 6 : (A) Schematic diagram of pancake kidney. Note the position of renal pelvis on the anterior aspect and the separate openings of both ureters [9].

(B,C,D)Venous and delayed phase of CT IVP showing minimally enhancing mass lesion(*) in pan cake kidney. Note two separate uncrossed ureters. The right uncrossed ureter is displaced by the mass lesion

Complications:
Usual complications related to stagnant urine, rotational anomaly and short Ureters like calculus and hydronephrosis are noted. The single renal vascular supply of pelvic Kidney is also at higher risk to damage in pelvic trauma, pregnancy or any pelvic lesions [12].

Natsis et al reviewed about 22 reports and proposed categories of the arteries supplying HSK according to Graves' classification and multiple renal arteries were found in 63% cases (45/71 cases) [13]. The most common patterns were Type-5 (28%) and Type-6 (24%) and least common patterns were Type-4 (4%) and Type-3 (3%). Single RA supplying each renal moiety of HSK only in 18% of cases were observed.

As CRE remains asymptomatic; it is only sporadically reported in literature. Turkvatan et al reported four cases, out of which two showed inferior ectopia type (both females) and two L-shaped tandem kidneys (both males) and noted hydronephrosis in two cases [30]. Solanki et al. evaluated 5 boys and 1 girl with CFRE (inferior ectopia type) and reported left to right ectopia in 4 cases and right to left cross over in 2 cases. Urinary tract infection, hydronephrosis, anorectal anomalies and ectopic opening of vas deferens were associated in these children. Sigmoid type of kidney associated with staghorn calculus was also reported. Superior ectopia was reported in a female patient in whom left to right ectopia was seen [31].

Pupca et al. observed the presence of double nutcracker syndrome due to the presence of Two left renal veins crossing anterior and posterior to aorta in a male patient with L-shaped CFRE [32]. Many congenital anomalies are associated with CRE with fusion such as vaginal agenesis, VACTERL association, TAR syndrome, renal dysplasia, intestinal malrotation and a single ureter [3,30,33]. Halaseh M et al found crossed fused renal ectopia in only 7 cases when they evaluated 400 children (1.75%) with DMSA scan [20]. Another retrospective review study by Glodny B stated the incidence of CRE as 1 out of 3078 CT scans and horse-shoe kidney as 1 out of 474 CT scans [21]. Majority of reported cases of pan cake kidneys present concominant anomalies in other organs.

Differential Diagnosis of fusion anomalies:
1. Nephroptosis: here, the kidney initially occupies its usual place, but gets down in relation to its normal position in the body; and is characterized by a normal vasculature and normal length tortuous ureters. It should be investigated for organomegaly or retroperitoneal masses that can cause kidney displacement.

2. Graft renal kidney which is in the iliac fossa, the renal vessels are anastomosed to the external or internal iliac vessels and the ureter is re-implanted into the bladder through a sub mucosal tunnel. It shows variable axis of the renal pelvis.

3. Autologous kidney: where surgical repositioning of the patient's own kidney is done.

4. Acquired renal atrophy: in such cases the kidney becomes smaller and is in proper position.

Imaging Modalities
In a diagnosed or suspected fused kidney, further laboratory and imaging Investigations are required to assess the status of the kidneys and to look for treatable causes of renal pathology. The early diagnosis of complications must be made to prevent permanent renal damage. Usually diagnosis of any fusion anomaly is done by ultrasound, Excretory urography and CT scan. Ultrasound is primary investigation while Excretory urography may not show absent fusion if the two renal masses are not widely Separated. CT scan offers accurate diagnosis; CT urography is also able to show normal urinary tract anatomy along with anomalies and other complications; if present. CT scan also provides useful information about vascular supply of fused kidneys. Main drawback of MDCT is increased risk of radiation exposure and iodinated contrast agents. MRI is good alternative for such patients and in pediatric, pregnant and repeated follow up patients. However it is not sensitive for calculus detection, has less availability and higher cost.

Management
Pelvi-ureteric junction treatment options include primary palliative techniques such as percutaneous nephrostomy or ureteric stenting, until more definitive surgery can be planned. Surgical treatment options include open ureteroplasty or laparoscopic pyeloplasty.

Surgical division of isthmus was also used previously to bring the kidneys and their drainages as close to physiological as possible, but it is not practiced now due to increased risk of complications including bleeding and renal infarction [18].

Extracorporeal shockwave lithotripsy (ESWL) and open surgery are used to treat calculus.

Percutaneous nephrostomy (PCN) can also be used as a palliative measure in hydronephrosis and pyonephrosis to decompress the collecting system and provides symptomatic relief.

In patients with infection, PCN can be used as a temporary measure until the antibiotics take effect. PCN contributes to drainage of renal abscess as well as decompression of the obstructed collecting system. Treatment options for VUR may include injection of bulking agent or re- implantation of ureters.

Interventional radiology plays an important role in the treatment of smaller tumors. The smaller tumors in horseshoe kidney can be treated by radiofrequency ablation or cryoablation to avoid the need for surgery. Vascular embolization also has a role in the treatment of larger tumors by reducing their size and vascularity which leads to symptomatic relief or can be used as a preoperative measure. In our study; according to the complication present, the patients were managed with various options like percutaneous nephrostomy, percutaneous nephrolithotomy, DJ stenting, extracorporeal shockwave lithotripsy and others [Table 4].

Conclusion

The diagnosis of fused kidney is not always associated with a poor prognosis. It requires long-term follow-up, early diagnosis of complications and to look for concomitant congenital anomalies. MDCT urography provides better evaluation of such patients in a single examination. Three-dimensional reformatted images can provide particularly good delineation of congenital fusion anomalies of the kidney along with its vascular supply information which is very important to the surgeon.

Abbreviations:

MDCT- multi detector CT scan
PUJO- pelvi ureteric junction obstruction
HSK- Horse shoe kidney
CRE- Crossed renal ectopia
IVC- Inferior vena cava
SVC- superior vena cava
RCC- renal cell carcinoma
TCC- transitional cell carcinoma
PC system- Pelvi calyceal system

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