Anatomy & Physiology: Current Research

ISSN: 2161-0940

Babu, Anat Physiol 2015, 5:S5
DOI: 10.4172/2161-0940.S5-001

Review Article Open Access

Renal Fusion Anomalies: A Review of Surgical Anatomy

CSRamesh Babu*, Vinay Sharma and Om Prakash Gupta

Department of Anatomy, Muzaffarnagar Medical College, Muzaffarnagar and Dr.O.P.Gupta Imaging Centre, Meerut, Uttar Pradesh, India

***Corresponding author:** Babu CSR, Department of Anatomy, Muzaffarnagar Medical College, Muzaffarnagar, Uttar Pradesh, India; Tel: +91 9897249202; E-mail: csrameshb@gmail.com

Rec date: Jun 22, 2015; **Acc date:** Jul 24, 2015; **Pub date:** July 27, 2015

Abstract

Congenital renal fusion anomalies, characterized by either partial or complete fusion of the two kidneys, are represented by horseshoe kidney, crossed renal ectopia with fusion and fused pelvic cake kidney. Though these anomalies may remain asymptomatic, in certain cases they may be associated with pathological conditions like nephrolithiasis, hydronephrosis, recurrent urinary tract infections, vesicoureteral reflux and renal neoplasms. Concomitant congenital anomalies involving other organ systems observed with some renal fusion anomalies make them important clinical entities. Thorough knowledge of these renal fusion anomalies, associated renovascular abnormalities, pathologies and anomalies of other organ systems are most essential for appropriate patient management by urologists, surgeons, vascular surgeons and radiologists alike. This review focuses on the surgical anatomy of various renal fusion anomalies, their incidence, anatomical, radiological and pathological features and associated anomalies in the light of available literature.

Keywords: Horseshoe kidney; Crossed fused renal ectopia; Cake kidney; Lump kidney; Renal vascular anomalies

Introduction

Congenital renal and urinary tract anomalies are not infrequent. Renal fusion anomalies are defined as the congenital fusion of the kidneys in early embryonic period either partially or completely. Partial fusion anomalies include horseshoe kidney (HSK) and crossed fused renal ectopia (CFRE) and complete fusion represented by 'cake' kidney or fused pelvic kidney [1]. These renal fusion anomalies exhibit abnormalities of position (ectopia), migration, rotation and vascular supply. They occur more frequently in males [2]. Many fusion anomalies remain asymptomatic and incidentally detected at autopsy, surgery or radiological investigations. Less frequently they may be associated with anomalies of skeletal, cardiovascular, genitourinary and gastrointestinal systems [3]. Presence of such renal fusion anomalies poses difficulties and complications during abdominal aortic aneurysm (AAA) surgery, retroperitoneal and pelvic surgeries, renal transplantation and interventional procedures. Thorough understanding of their anatomical and radiological features will greatly aid in their surgical management and avoid complications. This review aims to describe the developmental basis, classification, incidence, anatomical and radiological features of renal fusion anomalies such as HSK, CFRE and cake kidney.

Development

The development of the kidney begins in the 4th week of gestation by inductive interaction between the ureteric bud and the metanephric blastema. Ureteric bud arising from the mesonephric duct gives rise to the collecting tubules and the pelvicalyceal system and the metanephros develop into excretory part formed by nephrons. Initially the developing kidney lies in the sacral region and acquires blood supply from the neighbouring vessels. During 6th to 9th weeks of development, the lobulated kidney undergoes a complex process of ascent, probably due to differential growth of abdominal and pelvic regions, to reach its adult position in the upper part of posterior abdominal wall. During its ascent, the kidney successively receives arterial supply from median sacral, internal iliac, common iliac and dorsal aorta. These arteries represent lateral splanchnic branches and as the migrating kidney acquires new arteries cranially, the caudally placed arteries undergo degeneration. When the kidney reaches the renal fossa it undergoes 90° axial rotation such that the hilum which was initially anteriorly placed becomes medial. Congenital renal anomalies can occur due to abnormalities of development, migration and rotation.

Mechanism of Fuison Anomalies

The precise mechanism of development of renal fusion anomalies is not fully understood and several theories have been put forward to explain the anomaly. The Mechanical Theory proposes that during cephalad migration, the kidneys pass through the fork between the two umbilical arteries and any positional change in these arteries squeeze the kidneys close together allowing their fusion (result in HSK). Fusion of both nephrogenic blastemas with early arrested migration result in completely fused pelvic kidney. Abnormal position of an umbilical artery can result in abnormal migration of a renal unit to the contralateral side following the path of least resistance (crossed renal ectopia). The Theory of Abnormal Caudal Rotation proposes that fusion occurs due to lateral flexion and rotation of the caudal end of the embryo disturbing the relative position of the nephrogenic blastema and ureteric bud [4]. The distal curled end of the vertebral column permit one ureter to cross the midline and enter the opposite nephrogenic blastema or transplant the kidney and ureter to the opposite side during ascent. Association of scoliosis with crossed renal ectopia supports this theory. The Ureteral Theory states that cross over is strictly a ureteral phenomenon with the developing ureteral bud wandering to the opposite side and inducing the differentiation of the contralateral metanephric blastema and it is assumed that the metanephric tissue that does not receive a ureteric bud regresses.

According to the Teratogenic Theory, HSK results from abnormal migration of posterior nephrogenic cells due to teratogenic insult forming a parenchymal isthmus [5,6]. The increased incidence of malignancies and other organ system anomalies associated with HSK possibly supports this theory [7]. Finally Genetic Theory suggests that genetic influence may play a role because some renal fusion anomalies have been reported to occur in identical twins and siblings within the same family. It is suggested that the sonic hedgehog gene signal is critical for kidney positioning along the mediolateral axis and its disruption will result in renal fusion [8]. Mc Pherson suggested that HSK may occur as a previously undescribed autosomal dominant condition [9]. Analysis of patients with Turner syndrome revealed that 33% patients presented some renal malformations with HSK occurring in 7.1% of these patients, which renders support to the genetic theory [10]. Intense research is going on to unravel the genetic mechanisms underlying the development of congenital kidney and urinary tract anomalies [11].

Classification

A number of classification systems exist in the available literature classifying the various types of renal fusion anomalies. In 1927 Papin and Eisendrath proposed a classification system for renal and ureteral abnormalities [12]. They described three subtypes of anomalies of location as simple unilateral renal ectopia, simple bilateral ectopia and crossed ectopia with or without fusion. They also described median fusion type with four subtypes as horseshoe kidney, L-shaped kidney, cake kidney and sigmoid kidney. Mc Donald and Mc Clellan classified the crossed renal ectopia into four types as crossed renal ectopia with fusion, crossed renal ectopia without fusion, solitary crossed renal ectopia, bilaterally crossed renal ectopia out of which crossed fused renal ectopia (CFRE) represents the fusion anomaly (Figure 1) [13]. They described six forms of crossed fused renal ectopia as unilateral fused kidney inferior ectopia type, sigmoid or S-shaped kidney, lump kidney, L-shaped kidney, disc kidney and unilateral fused kidney superior ectopia type (Figure 2).

Brief definition of various forms of fusion anomalies is warranted because some of the terms such as 'cake kidney', pancake kidney', 'lump kidney' and 'disc kidney' are used interchangeably by some authors to describe complete renal fusion type of cake kidney as well as crossed fused renal ectopia type of lump kidney. Horseshoe kidney is defined as a congenital median fusion anomaly in which either the lower poles (most common) or the upper poles (rare) of the two kidneys are connected to each other by an isthmus (made up of either parenchymal or fibrous tissue) placed across the midline resulting in the presentation of an U- shaped or inverted U-shaped renal mass and also exhibiting positional (ectopia) and rotational anomaly. Crossed renal ectopia occurs when a kidney is located on the side opposite from which its ureter enter into the urinary bladder [2]. Ninety percent of crossed ectopic kidneys are fused to their ipsilateral mate and the ureter draining the ectopic kidney characteristically cross the midline. In all the six subtypes of CFRE, the ureter from the ectopic kidney crosses the midline. Cake kidney or fused pelvic kidney represent complete type of renal fusion anomaly and is defined as " an anomaly in which the entire renal substance is fused into one mass, lying in the pelvis and giving rise two separate ureters which enter the bladder in normal relationship" [14]. This anomaly, also known as 'pancake kidney', 'lump kidney' and 'disc kidney' should not be confused with CFRE types of 'lump' kidney and 'disc' kidney in which the ureter of the ectopic kidney crosses the midline. Moreover CFRE type 'lump' kidney is generally located at a higher level lying on one side of the midline, whereas, the cake kidney representing the complete renal fusion is located in the midline in presacral region.

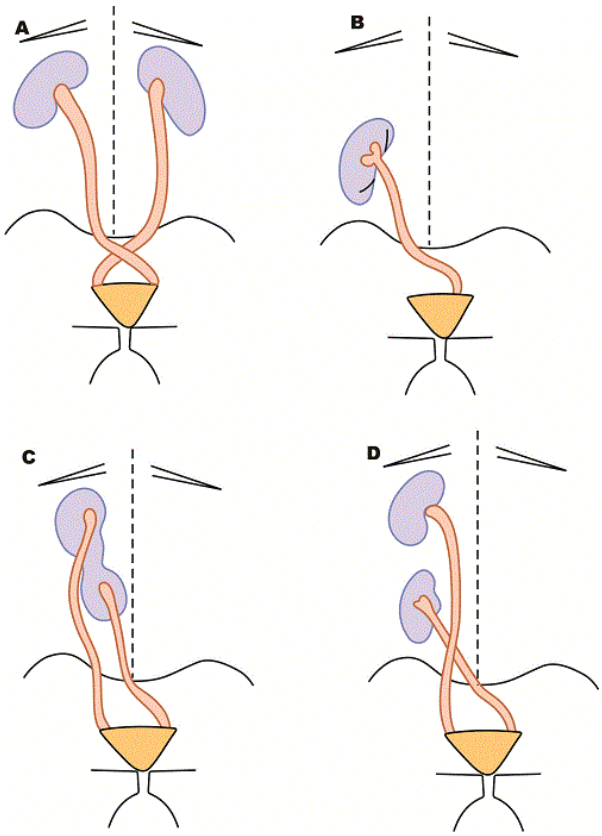

Figure 1: Mc Donald and Mc Clellan's classification of crossed renal ectopias. A bilateral crossed renal ectopia, B- Unilateral crossed renal ectopia; C- Crossed fused renal ectopia; D- Crossed unfused renal ectopia. Note that in all these types the ureter is crossing the midline. Only the crossed fused renal ectopia (C) is the fusion anomaly constituting 90% of all crossed ectopias.

It is preferable to use the terms 'cake kidney' or 'fused pelvic kidney' to denote the complete renal fusion anomaly and restrict the use of 'lump' kidney and 'disc' kidney to CFRE types of fusion anomaly.

It is pertinent to note that 'lump' and 'disc' kidneys are formed by extensive fusion of the two renal masses over a wide margin, but the ureter draining the ectopic renal mass always cross the midline before opening into urinary bladder and to include them as a complete renal fusion anomaly is debatable.

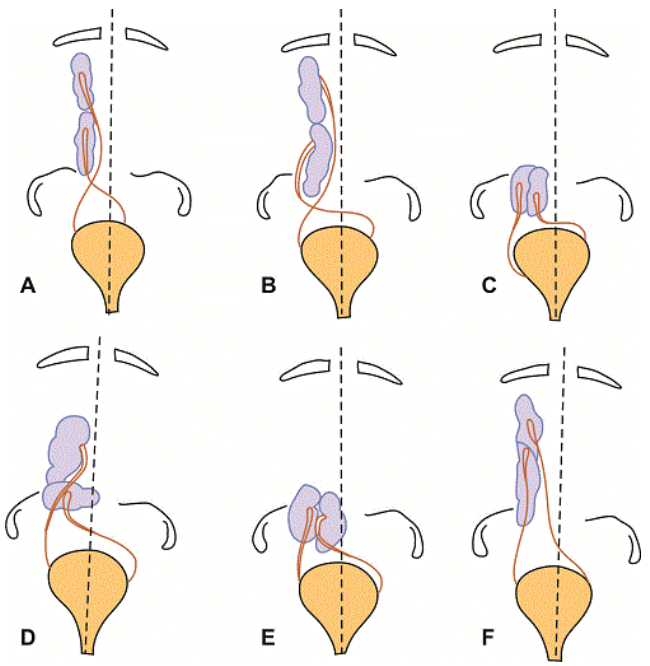

Figure 2: Six subtypes of crossed fused renal ectopia. A- Inferior ectopia type with upper pole of ectopic kidney fusing with lower pole of normal kidney. B- Sigmoid or S-shaped kidney where hilum of ectopic kidney faces laterally and that of normal kidney medially and with fusion form S-shaped mass. C- Lump kidney with fusion of two kidneys over a wide margin with ureter from ectopic kidney crossing the midline. D- L-shaped or Tandem kidney in which the ectopic kidney is placed horizontally fusing with lower pole of normal kidney. E- Disc kidney with extensive fusion of two kidneys forming a disc shaped mass. F- Superior ectopia type with ectopic kidney placed above the normal kidney and fusing with its upper pole.

Horseshoe Kidney

Incidence

Horseshoe kidney is the most common renal fusion anomaly which combines three anatomical abnormalities: ectopia, malrotation and vascular changes and is found more commonly in men than in women with a ratio of 2:1 [7]. It accounts for 90% of all fusion anomalies and occurs in about 0.25 % of the population [1,3]. An incidence of 1 in 666 cases was found after analyzing radiological data of 15320 patients at a single institution [15]. Glodny et al. estimated the prevalence rates for HSK in adults examined by sonography as 1:708 and by CT as 1:474 [16]. In a cross sectional study on 12000 patients using sonography and contrast urography, only 4 cases (0.03%) of HSK was found [17]. In contrast, a sonographic study from Nepal has reported an incidence of 1 in 516 (61 HSKs out of 31498 patients screened, 0.2%) with male to female ratio of 1:2 (20 males; 41 females) [18]. We have reported an incidence of 1 in 97 cases (7 out of 682 cases; 1.02%) after retrospective analysis of multidetector CT scans of 682 patients [19]. Natsis et al has reported an incidental finding of HSK in 1 male out of 250 (0.4%) Greek cadavers dissected during a period of 30 years [7]. Though it is suggested that there is no racial determination for HSK, available literature indicates that the incidence of horseshoe kidney varies in different populations and further research is needed to throw some light on this aspect.

Morphology

Horseshoe kidney can be classified into three types of shape according to the morphological appearance of fusion [7]. Midline fusion of the lower poles of two symmetrically placed kidneys on either side of the vertebral column result in the formation of U-shaped HSK, while similar fusion of upper poles result in inverted U-shaped HSK. Lateral fusion of two asymmetrically placed kidneys, one vertical and one horizontal result in L- shaped HSK with laterally placed isthmus. Horseshoe kidneys are also classified as symmetrical (midline fusion) and asymmetrical (lateral fusion) based upon the site of fusion [4]. Glodny et al. have observed midline fusion in 40% cases, left lateral in 38 % and right lateral in 22% cases [16]. We have reported midline fusion in 4 and lateral fusion in 3 cases [19] (Figure 3). Most commonly the isthmus lies opposite to L-3 to L5 vertebral levels, below the origin of inferior mesenteric artery and anterior to abdominal aorta (AA) and inferior vena cava (IVC). Rarely the isthmus lies posterior to great vessels or runs between them.

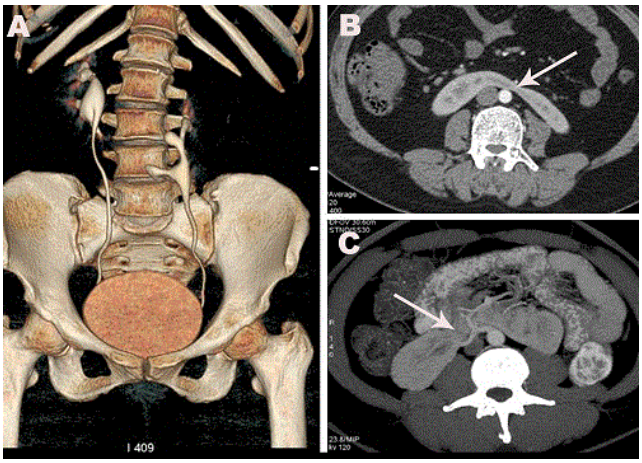

Figure 3: A: Volume rendered MDCT urography of a 50 year old female patient showing pelvicalyceal system and the ureters of HSK. B: Axial image showing left lateral fusion (arrow) in a 33 year old male patient and C: axial image showing right lateral fusion (arrow) of HSK. Note the presence of precaval course of a right renal artery in a 42 year old male patient (unpublished from our archive).

The position of isthmus posterior to IVC but anterior to AA results in a IVC anomaly named as 'preisthmic IVC' which has been sporadically reported in the literature [20-22]. Because of arrested cephalad migration and fusion, HSK exhibits malrotation and axis deflection. The long axis is deflected downwards and medially and the hilum faces anteriorly or anterolaterally. Pelvicalyceal system is arranged atypically with the calyces pointing inwards. The ureters pass downwards crossing the isthmus anteriorly.

Vascular supply

During development when the metanephric blastema is lying in the pelvis, it acquires branches from distal part of AA, common iliac, internal iliac and median sacral arteries. In the process of ascent the arterial supply continuously changes with generation of new arteries cranially and degeneration of old arteries caudally. Since HSK develops due to arrested migration, it is obvious that its arterial supply will show abnormalities in the form of additional renal arteries and ectopic origin [23]. In fact single renal artery for each renal moiety of HSK can be found in less than 30% cases (Figure 4). Papin's autopsy study of 139 horseshoe kidneys served as the basis for a classification system consisting of three groups.

Group 1 kidneys have normal renal arteries and account for 20% of all horseshoe kidneys; Group 2 have three to five renal arteries and account for 66% of cases; Group 3 have more than five renal arteries and account for 14% [24].

A number of other classification systems are available in the literature for renal arterial pattern. Vascular surgeons prefer to use a simple classification system based on the level of origin of renal artery [25]. In type I renal arteries (RA) arise from the normal position. In type II vascularization is by two normal renal arteries and one or more accessory renal arteries of ectopic origin from distal aorta or iliac arteries. In type III all RAs supplying HSK have ectopic origin. Graves described six basic patterns of arteries supplying HSK studied by means of resin cast [26] (Figure 5). Pattern of blood supply may be similar to that of normal kidney with single artery supplying upper, middle and lower segments (Type 1).

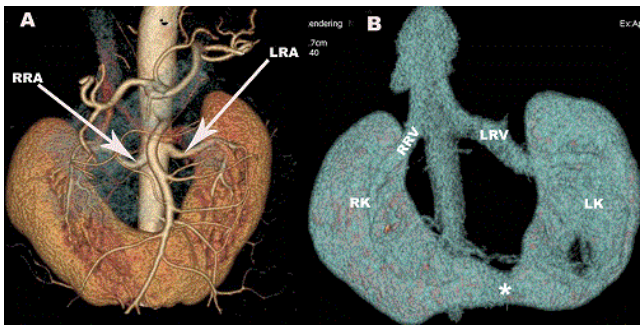

Figure 4: Volume rendered MDCT angiography of a 50 year old male patient with HSK. A: Single renal artery arising from lateral aspect of AA supplies each renal moiety of HSK. RRA- right renal artery; LRA- left renal artery. B: Single renal vein from each renal moiety of HSK drain into IVC. RK- right kidney; LK- left kidney; RRV right renal vein; LRV- left renal vein. In B- isthmus is indicated by * (unpublished from our archive).

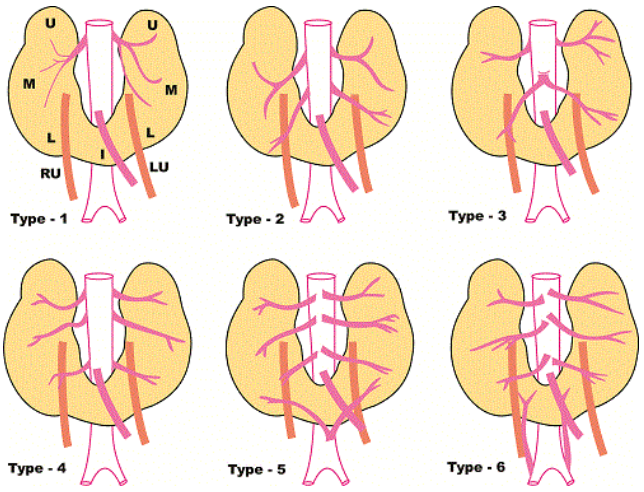

Figure 5: Scheme of six arterial patterns (Type-1 to Type-6) of HSK described by Graves. U,M,L represent upper, middle and lower segments of the kidney. I-isthmus: RU- Right Ureter; LU- Left Ureter. For details see text.

Upper and middle segments of each kidney may be supplied by a single artery, with a vessel from aorta entering each lower segment (Type 2). Sometime the arteries to lower segment arise from aorta by a common trunk (Type 3). All three segments are supplied by separate arteries arising from aorta (Type 4). The fused segment (isthmus) may also be supplied by arteries which arise above or below the isthmus, these may be unilateral or bilateral and may originate from the aorta independently or by a common trunk (Type 5). Finally the fused lower segment may be supplied on one or both sides by branch originating from the common iliac or rarely from hypogastric (internal iliac) or median sacral artery (Type 6). All the six patterns depict symmetrical supply to both renal moieties of the horseshoe kidney.

Natsis et al in their review analysed about 22 published reports categorizing the arteries supplying HSK according to Graves' classification and in majority of the cases multiple renal arteries (45/71 cases; 63%) supplied HSK [7]. The most common pattern observed was Type-5 (28%) and Type-6 (24%) and least common pattern was Type-4 (4%) and Type-3 (3%). They observed single RA supplying each renal moiety of HSK only in 18% of cases. Symmetrical arterial pattern is not always present and we have noted symmetrical supply only in 3 out of 7 cases of HSK and in the rest asymmetrical supply.

We have reported (Figures 6 and 7) precaval course of main right renal artery in one case and lower right renal arteries in four cases [19] (Figure 8). Analysing the arterial pattern of 90 HSKs, Glodny et al. have observed that the second artery on the right side has a precaval course [16]. Presence of precaval right RA may be mistaken for a gonadal artery especially in imaging studies.

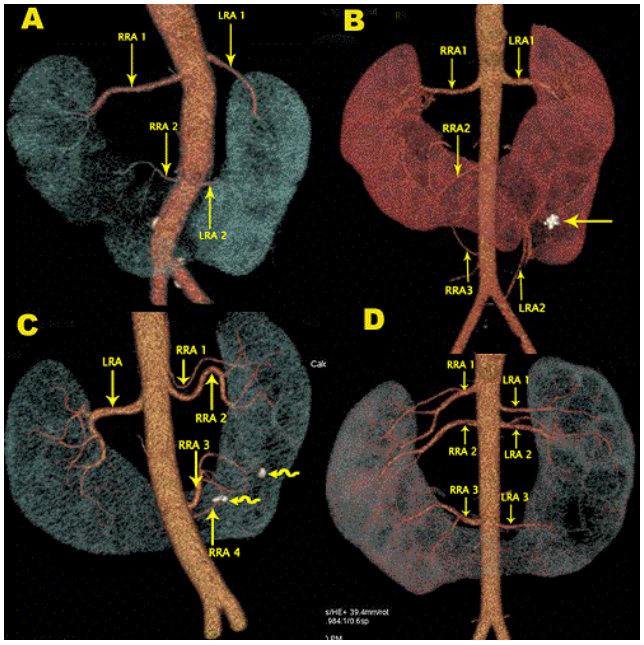

Figure 6: Volume rendered MDCT angiographic images of HSK depicting different arterial patterns. A: Symmetrical supply by two renal arteries arising from AA on each side. B: HSK supplied by 3 RRAs and 2 LRAs with lower LRA (LRA-2) arising from left common iliac. Arrow indicates a calculus in the lower pole of left renal moiety. C: Posterior view showing HSK supplied by a single LRA and 4 RRAs. Calculi present in the right renal moiety indicated by curved arrows. D: Symmetrical supply by 3 renal arteries on each side all arising from AA [19].

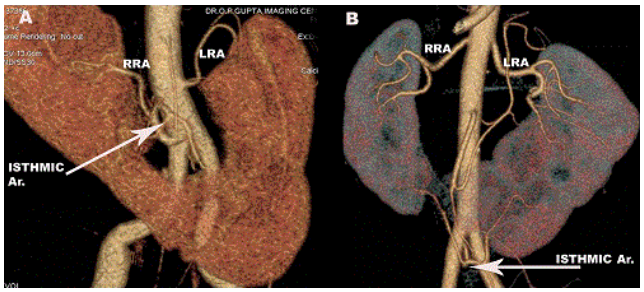

Figure 7: Volume rendered MDCT angiographic images showing arterial supply to isthmus of HSK in a 50 year old female patient (A) and in a 60 year old female (B). A- Single LRA having ectopic origin from AA below the level of origin of inferior mesenteric artery. Single RRA arise ectopically from the anterior aspect just above aortic bifurcation and gives off an isthmic branch. B- Single RRA and LRA having normal origin from AA supply each renal moiety of HSK. Additionally an isthmic branch having ectopic origin from posterior aspect of right common iliac artery, gives two branches to supply lower poles of both renal moieties. (unpublished from our archive).

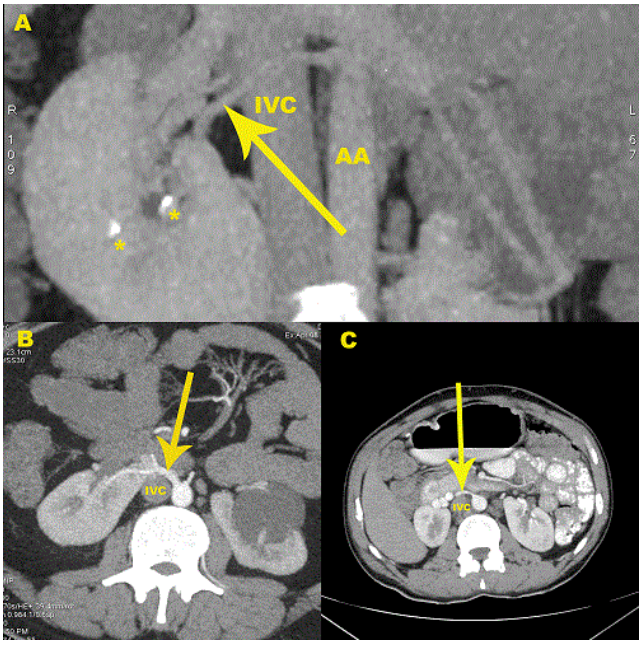

Figure 8: MDCT angiographic images showing precaval right renal artery. ACoronal image, B and C- Axial images showing precaval right renal artery indicated by arrow, passing anterior to IVC [19].

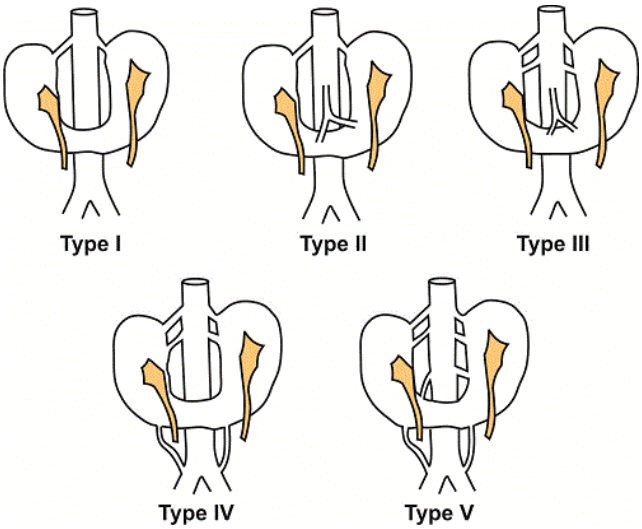

Figure 9: Scheme showing five types (Type I to Type V) of arterial pattern according to the classification by Eisendrath. See text for details.

Recently Ichikawa et al. investigated the RA anomalies associated with HSK using CT angiography and the classification scheme of Eisendrath [27] (Figure 9). According to this classification one RA to each renal moiety is classified as type I. Type II pattern has one RA to each renal moiety and an aortic branch to isthmus, Type III pattern has two RAs to each renal moiety and an aortic branch to isthmus,

Type IV has two RAs to each side of the kidney with one or more arising from the iliac arteries including the isthmus branch and Type V pattern has multiple RAs arising from aorta, mesenteric and iliac arteries. In their evaluation of 39 patients with HSK, Ichikawa et al noted Type V pattern in 22 cases (56.4%) and Type II in 12 cases (30.7%) [27]. The incidence of supernumerary RAs in a normal kidney is generally about 28% to 30% [28]. In contrast, the incidence of supernumerary RAs in patients with HSK was reported as 60% - 81% which is significantly higher than that of general population [2]. Ichikawa et al reported an incidence of 92.3% for supernumerary RAs in patients with HSK and 33% in patients without HSK [27]. Because of wide variations in arterial pattern of HSK reported in the literature, a simple classification system is precluded [29].

Anatomical variations of renal veins (RV) are also observed in patients with HSK. In a study on 105 patients with HSK renal vein anomalies were observed in 24 patients (22.9%) which included 15 double right RVs, 1 triple right RV, 5 double left RVs, 4 circum-aortic and 1 retroaortic left RVs. Two patients had both circum-aortic left RVs and double right RVs [21]. Anomalies of inferior vena cava (3.9% - 5.7%) were found significantly more frequently in patients with HSK than those without it [20-22]. These IVC anomalies include preisthmic IVC, double IVC, left IVC and azygos continuation of IVC. Such renovascular and IVC anomalies associated with HSK pose technical difficulties during abdominal aortic aneurysm surgery [30,31]. Significantly higher incidence of anomalous superior vena cava (SVC) was also reported in patients with HSK (4.2%) than in those without it (0.22%) [32]. Vascular surgeons need to keep the various venous and renovascular anomalies in mind, because they are more frequently associated with a horseshoe kidney. Familiarity with venous and renovascular anomalies is essential for correct interpretation of images to avoid erroneous diagnosis of retroperitoneal masses or lymphadenopathy and to prevent fatal complications [33].

Associated anomalies

Natsis et al. in their review elaborately discussed about the congenital anomalies and pathologic conditions associated with HSK [7]. The most common pathologic conditions include renal calculi, pelviureteric junction (PUJ) obstruction, hydronephrosis and recurrent infections. High insertion of ureter into the renal pelvis and crossing of ureter over the isthmus are implicated as the causative factors for PUJ obstruction and subsequent hydronephrosis. Horseshoe kidney can also be associated with congenital anomalies and the systems most commonly affected are skeletal (hemivertebra, scoliosis, rib defect, club foot, congenital hip dislocation), cardiovascular (Ventricular Septal Defect), GIT (anorectal malformation, malrotation and Meckel's diverticulum), CNS (neural tube defect) and Genito-urinary (vesicoureteral reflux, duplication of ureter, hypospadias, undescended testis, bicornuate or septate uterus) [1,2,7]. Horseshoe kidney is also associated with certain chromosomal anomalies like Turner syndrome, Down syndrome (trisomy 21), Patau syndrome (trisomy 13) and Edwards syndrome (trisomy 18) [7]. Therefore it is necessary to thoroughly investigate the patients with HSK for any associated congenital anomalies and pathologic conditions.

Crossed Fused Renal Ectopia

Incidence

Crossed renal ectopia (CRE) is a rare type of anomaly in which both the kidneys are situated on one side and in about 90% of such cases the crossed ectopic kidney is fused with the orthotopically located kidney. In this condition the ectopic kidney is located contralateral to the side of its ureteric orifice and the ureter of the ectopic kidney cross the midline which distinguishes this condition from horseshoe kidney. Crossed fused renal ectopia (CFRE) is the second most common renal fusion anomaly with an estimated incidence of 1:1300 to 1:7500 [1,2]. It is both a fusion and ectopic anomaly and occurs in about 0.08% - 0.01% cases. The prevalence of the crossed renal ectopia with fusion was estimated to be 1 in 1000 live births [34]. In a review of 400 children evaluated by DMSA renal scan, crossed fused renal ectopia was found in 7 cases (1.75%) [35]. In another retrospective review, the incidence of CRE was reported as 1 out of 3078 CT scans [16]. We have reported 3 cases of CFRE (2 females, 1 male) in a retrospective analysis of MDCT scans of 682 patients with an estimated incidence of 0.43% [36]. The true incidence of this anomaly is not known because a large majority of the patients having this anomaly remain asymptomatic and undetected. The left kidney is most commonly ectopic crossing to the right side of the abdomen and the condition is more common in males [2].

Classification

Mc Donald and Mc Clellan classified CFRE into six types [13] (Figure 2). In decreasing order of frequency they are: (A) Unilateral fused kidney inferior ectopia with the upper pole of the crossed ectopic kidney fusing with the lower pole of the orthotopic ipsilateral mate. Both renal pelves may be anterior. (B) Sigmoid or S-shaped kidney in which the crossed kidney lies inferiorly with the renal pelvis directed laterally and the normally positioned kidney lies superiorly with the pelvis directed medially. Each renal pelvis is oriented correctly in this type because the fusion of the two kidneys occurs after the complete rotation on the vertical axis has taken place. (C) Unilateral Lump kidney with fusion occurring over a wide margin and both renal pelvis directed anteriorly; located more inferiorly. (D) L-Shaped or Tandem kidney in which the crossed kidney lies inferiorly and transversely fusing with the lower pole of the normal kidney. (E) Unilateral disc kidney in which the fusion occurs along the medial borders and (F) Unilateral fused kidney superior ectopia type is the least common type; the ectopic kidney is placed superiorly with its lower pole fusing with the upper pole of the normal kidney. Both renal pelves are anterior.

Literature review of anatomical features

CFRE is sporadically reported in the literature because this anomaly may remain as a silent clinical entity without producing any signs and symptoms. Turkvatan et al reported four cases, of which two were inferior ectopia type (both females) and two L-shaped tandem kidneys (both males) and hydronephrosis was noted in two cases [37]. Out of 3 cases studied, we have observed two cases of L-shaped kidneys (in females) and a case of inferior ectopia in a male (Figures 10 and 11) [36] .

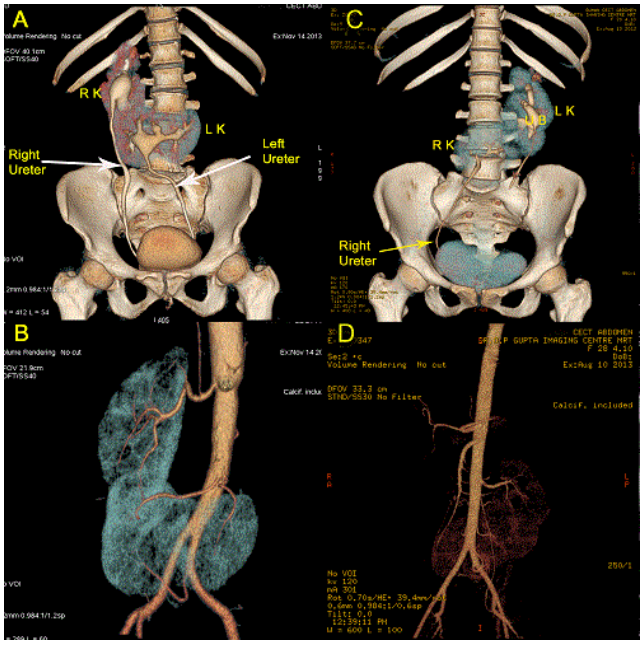

Figure 10: MDCT angiography and urography images showing CFRE and arterial supply. A- Left to right CFRE causing L-shaped tandem kidney with left ureter crossing the midline in front of sacral promontory. B- The normally positioned vertically placed right renal moiety is supplied by a single RRA; the ectopic horizontally placed left renal moiety is supplied by three left renal arteries, two arteries arising just above aortic bifurcation and one from left common iliac artery. C- Right to left renal ecopia with right ureter from the ectopic kidney crossing the midline. D- Single LRA supplies the normally positioned left renal moiety. The ectopic right kidney is supplied by a branch from left common iliac artery having a recurved course [36].

Solanki et al. evaluated 5 boys and 1 girl, all having CFRE (inferior ectopia type) and noted 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 observed as associated anomalies in these children [38]. Sigmoid type of kidney, which is second common type of CFRE, associated with staghorn calculus was also reported [39]. Superior ectopia, the rarest type of CFRE , was reported in a female patient in whom left to right ectopia was seen [40]. Yin et al described a right to left CFRE of superior ectopia type in a male patient with thoracic scoliosis. Most importantly, 'y' type fusion of the two ureters to form a single dilated ureter which crossed the midline posterior to left common iliac artery (retroiliac megaureter) was noted [41]. Right to left crossed fused superior ectopia was incidentally detected in a male patient in the left iliac fossa. Each renal moiety of the fused renal mass was supplied by a single renal artery arising from the common iliac arteries [42]. The so called 'lump kidney' was detected in three cadaveric dissections, all showing left to right ectopia and aberrant vascular supply [43-45]. One case of lump kidney was associated with intestinal malrotation [45]. Kaufman and Findlater reported a 'cake' or 'lump' kidney with left to right ectopia located in the right lumbar region in a 81 year old female cadaver. The fused renal mass was supplied by three renal arteries, two of them having precaval course and three renal veins [3].

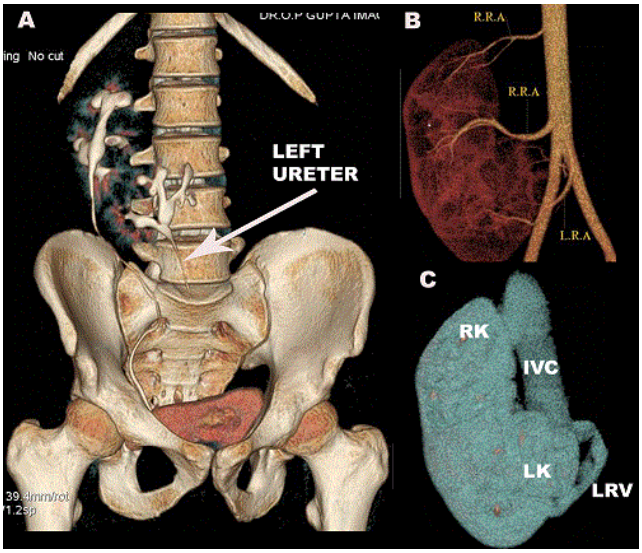

Figure 11: MDCT angiography and urography images showing CFRE inferior ectopia type and its vascular supply in a 32 year old male patient. A- Left to right ectopia with left ureter crossing the midline. B- Two RRAs supply normally positioned right kidney and the ectopc left kidney being supplied by a branch arising just above aortic bifurcation and passing downwards and to the right to reach the lower pole of left kidney. C- Each renal moiety of the fused kidney drained by a single renal vein into IVC.LRV- left renal vein; RK- right kidney; LK- left kidney (unpublished from our archive).

Similar, left to right ectopic fused 'cake' kidney drained by two distinct ureters, was detected in a 12 year old male patient with recurrent urinary tract infections [46]. Though, in both these cases the fused mass was located to the right of midline and the ureter from the ectopic kidney crossed the midline, the authors did not describe this anomaly as crossed fused ectopia of 'lump' type. An extremely rare case of right lump kidney with six renal arteries, two renal veins and two duplicated pelvicalyceal systems was described in a male patient [47]. Pupca et al. reported 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 [48]. 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 [1,36,37].

Cake Kidney Or Fused Pelvic Kidney

Cake kidney or fused pelvic kidney is a very rare congenital anomaly with a few more than 20 cases described in the literature [1,49,50]. The term should be used to describe completely fused renal mass located in the pelvic cavity and drained generally by two ureters which do not cross the midline (Figure 12). Very rarely, a single ureter is found draining the cake kidney. The 'lump' and 'disc' kidneys are terms used alternatively for cake kidney, as well as for the subtypes of CFRE characterized by midline crossing of ureter from the ectopic kidney and unilateral location of the near complete fused renal mass in the lumbar and iliac fossa regions. Cake kidney accounts for only about 2% of all fused kidney types [3]. The estimated incidence is 1/65000 to 1/375000 cases [47].

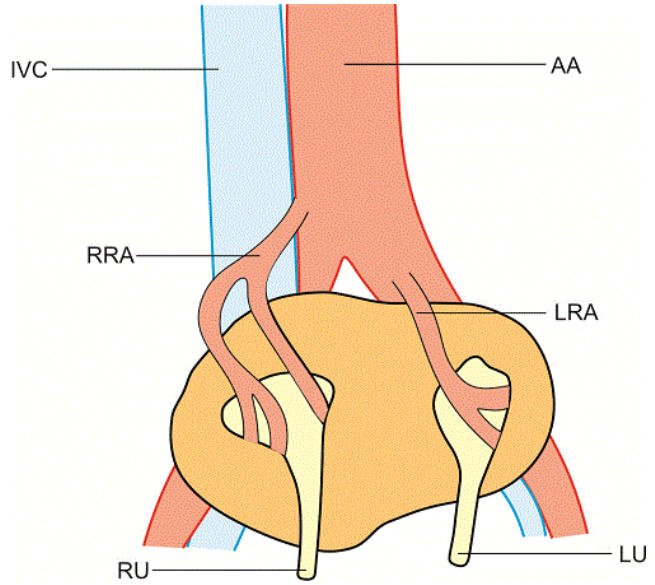

Figure 12: Schematic diagram showing some features of cake kidney. Note the position of renal pelvis on the anterior spect and the emergence of right ureter (RU) and left ureter (LU) separately.

Generally 'Cake kidney' remains asymptomatic and may be detected at any age. This condition may be present associated with other congenital anomalies like anomalies of testicular descent, anomalies of vas deferens, vaginal agenesis, bicornuate or unicornuate uterus, sacral agenesis, caudal regression syndrome, tetralogy of Fallot and spina bifida [51-53]. A triad of complete dorsal pancreatic agenesis, bicornuate uterus and cake kidney was detected in a middle aged woman [54]. Like other renal fusion anomalies it is more common in males with a male to female ratio of about 2-3:1 [3]. Developmentally, when the renal anlagens fail to ascend and remain in the pelvic cavity extensively fusing with each other, a cake kidney is formed retaining the primitive vascular supply. Vascular supply may be derived from a single renal artery (from distal aorta or common iliac) and a single renal vein (drain into IVC or common iliac veins) [55]. The single renal vascular supply to cake kidney is at increased risk of damage by pelvic trauma, pregnancy or space occupying lesions. The renal pelvis is anteriorly placed and uncrossed short ureters open separately into urinary bladder [56].

Conclusion

Renal fusion anomalies, though rare and asymptomatic, are of immense clinical importance because of their concomitant congenital anomalies involving other organ systems. Many of these are detected incidentally at autopsy, surgery and radiological investigations. Various imaging modalities used to investigate renal fusion anomalies include sonography, intravenous pyelography, computed tomography, renal scintigraphy, MDCT angiography and MRI. Available literature suggest that MDCT angiography with delayed phase urography is the modality of choice comprehensively depicting renal fusion anomalies as well as associated anomalies, if any, in a single examination for appropriate patient management.

Acknowledgement

Technical assistance rendered by Mr. Arjun Singh of Dr. O.P. Gupta Imaging Centre , Meerut, Mr. Sushil Kumar and Mr. Titender of Muzaffarnagar Medical College, Muzaffarnagar and Mr. Pran Praksh of J.R.S. Hospital, Meerut in preparation of images and figures is sincerely acknowledged.

Conflict Of Interest

None to declare. No funding received from any source.

References

1. Türkvatan A, Olçer T, Cumhur T (2009) Multidetector CT urography of renal fusion anomalies. Diagn Interv Radiol 15: 127-134.
2. Bauer SB (2002) Anomalies of the upper urinary tract. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ (Eds) Campbell's Urology, Vol.3, 8th. Ed. Philadelphia: WB Saunders.
3. Kaufman MH, Findlater GS (2001) An unusual case of complete renal fusion giving rise to a 'cake' or 'lump' kidney. J Anat 198: 501-504.
4. Cook WA, Stephens FD (1977) Fused kidneys: morphologic study and theory of embryogenesis. Birth Defects Orig Artic Ser 13: 327-340.
5. Doménech-Mateu JM, Gonzalez-Compta X (1988) Horseshoe kidney: a new theory on its embryogenesis based on the study of a 16-mm human embryo. Anat Rec 222: 408-417.
6. Tijerina GO, Uresti J, Urrutia VE, et al. (2009) Anatomical study of the horseshoe kidney. Int J Morphol. 27: 491-494.
7. Natsis K, Piagkou M, Skotsimara A, Protogerou V, Tsitouridis I, et al. (2014) Horseshoe kidney: a review of anatomy and pathology. Surg Radiol Anat 36: 517-526.
8. Shapiro E, Bauer SB, Chow JS (2012) Anomalies of the upper urinary tract. In: Alan J Wein (Ed) Campbell-Walsh Urology. Vol. 4, 10th Edn., Philadelphia, Elsevier Saunders. Chapter 117, pp. 3145.
9. McPherson E (2007) Renal anomalies in families of individuals with congenital solitary kidney. Genet Med 9: 298-302.
10. Lippe B, Geffner ME, Dietrich RB, Boechat MI, Kangarloo H (1988) Renal malformations in patients with Turner syndrome: imaging in 141 patients. Pediatrics 82: 852-856.
11. Yosypiv IV (2012) Congenital anomalies of the kidney and urinary tract: a genetic disorder? Int J Nephrol 2012: 909083.
12. Papin E, Eisendrath DN (1927) Classification of Renal and Ureteral Anomalies. Ann Surg 85: 735-756.
13. MC Donald JH, MC Clellan DS (1957) Crossed renal ectopia. Am J Surg 93: 995-1002.
14. Glenn JF (1958) Fused pelvic kidney. J Urol 80: 7-9.
15. Weizer AZ, Silverstein AD, Auge BK, Delvecchio FC, Raj G, et al. (2003) Determining the incidence of horseshoe kidney from radiographic data at a single institution. J Urol 170: 1722-1726.
16. Glodny B, Petersen J, Hofmann KJ, Schenk C, Herwig R, et al. (2009) Kidney fusion anomalies revisited: clinical and radiological analysis of 209 cases of crossed fused ectopia and horseshoe kidney. BJU Int 103: 224-235.
17. Asghar M, Wazir F (2008) Prevalence of renal ectopia by diagnostic imaging. Gomal J Med Sci. 6: 72-76.
18. Shrestha O, Shrestha GL. Ultrasonographically detected renal fusion anomalies in western region of Nepal. J Chitwan Med Coll. 2014: 4: 25-28.
19. Sharma V, Ramesh Babu CS, Gupta OP. (2015) Horseshoe kidney: A multidetector computed tomography study. Int J Anat Res. 3: 1049-1055.
20. Kehagias DT, Gouliamos AD, Vlahos LJ (1999) Horseshoe kidney associated with anomalous inferior vena cava. Eur Radiol 9: 935-936.

21. Ichikawa T, Kawada S, Koizumi J, Endo J, Iino M, et al. (2011) Major venous anomalies are frequently associated with horseshoe kidneys. Circ J 75: 2872-2877.
22. Ichikawa T, Kawada S, Koizumi J, Endo J, Itou C, et al. (2013) Anomalous inferior vena cava associated with horseshoe kidney on multidetector computed tomography. Clin Imaging 37: 889-894.
23. Boatman DL, Cornell SH, Kölln CP (1971) The arterial supply of horseshoe kidneys. Am J Roentgenol Radium Ther Nucl Med 113: 447-451.
24. Papin E (1928) Chirurgie du rein, anomalies du rein. Peris. G.Doin. pp 205-220.
25. Crawford ES, Coselli JS, Safi HJ, Martin TD, Pool JL (1988) The impact of renal fusion and ectopia on aortic surgery. J Vasc Surg 8: 375-383.
26. Graves FT (1969) The arterial anatomy of the congenitally abnormal kidney. Br J Surg 56: 533-541.
27. Ichikawa T, Tanno K, Okochi T, Koizumi J, Mori N, et al. (2015) Evaluation of Renal Artery Anomalies Associated with Horseshoe Kidney Using CT Angiography. Tokai J Exp Clin Med 40: 16-21.
28. Ozkan U, Oğuzkurt L, Tercan F, Kizilkiliç O, Koç Z, Koca N et al. (2006) Renal artery origins and variations: angiographic evaluation of 855 consecutive patients. Diagn Interv Radiol 12: 183-186.
29. Stroosma OB, Scheltinga MR, Stubenitsky BM, Kootstra G (2000) Horseshoe kidney transplantation: an overview. Clin Transplant 14: 515-519.
30. Chaudhuri A (2011) Exclusion of an infrarenal AAA with coincident horseshoe kidney and renovascular anomalies is feasible using a standard stent-graft. Eur J Vasc Endovasc Surg 41: 654-656.
31. Radermecker MA, Van Damme H, Kerzmann A, Creemers E, Limet R (2008) Association of abdominal aortic aneurysm, horseshoe kidneys, and left-sided inferior vena cava: report of two cases. J Vasc Surg 47: 645-648.
32. Ichikawa T, Sekiguchi T, Kawada S, Koizumi J, Endo J, et al. (2012) Study of the association between an anomalous superior vena cava and horseshoe kidney. Circ J 76: 1253-1258.
33. Sato A (2011) Venous anomalies and horseshoe kidney. A minefield in open vascular surgery. Circ J 75: 2759-2760.
34. Abeshouse BS, Bhisitkul I (1959) Crossed renal ectopia with and without fusion. Urol Int 9: 63-91.
35. Halaseh M, Alkhawaldeh K, Al-Ibraheem A, Al-Adwan H, Al-Kaylani H. (2011) Detection of congenital renal anomalies in children being investigated by Tc 99m- DMSA renal scan. J. Royal Med Serv., (JRMS) 18: 36-42.
36. Sharma V, Ramesh babu CS, Gupta OP (2014) Crossed fused renal ectopia multidetector computed tomography study. Int J Anat Res. 2 (2): 305-309.
37. Turkvatan A, Olcer T, Cumhur T, Akdur PO (2008) Multidetector computed tomographic urography for evaluation of crossed fused renal ectopia. Journal of Ankara University Faculty of Medicine 61: 149-154
38. Solanki S, Bhatnagar V, Gupta AK, Kumar R (2013) Crossed fused renal ectopia: Challenges in diagnosis and management. J Indian Assoc Pediatr Surg 18: 7-10.
39. Amin MU, Khan S, Nafees M (2009) Crossed fused renal ectopia with staghorn calculus and gross hydronephrosis. J Coll Physicians Surg Pak 19: 69-70.
40. Patel TV, Singh AK (2008) Crossed fused ectopia of the kidneys. Kidney Int 73: 662.
41. Yin Z, Yang JR, Wei YB, Zhou KQ, Yan B (2014) A new subtype of crossed fused ectopia of the kidneys. Urology 84: e27.
42. Akdogan L, Oguz AK, Ergun T, Ergun I (2015) The Rarest of the Rare: Crossed Fused Renal Ectopia of the Superior Ectopia Type. Case Rep Nephrol 2015: 742419.
43. Potu BK, Subramaniam B, Cheng PS. (2012) Crossed fused renal ectopia: a case report. Eur J Anat. 16: 79-81.
44. Rajaram V,Govindarajan M (2011) Crossed fused renal ectopia- a case report. Int J Anat Sci. 2: 19-21.
45. Kulkarni R, Appaji AC, Kulkarni RN (2013) Crossed renal ectopia associated with malrotation of intestine: A rare case report. Int J Anat Res. 1: 53-56.
46. Gun S, Ciantelli GL, Takahashi MAU, Brabo AA, Morais LA (2012) Complete renal fusion in a child with recurrent urinary tract infection. Radiol Bras. 45: 233-234.
47. Miclaus GD, Pupca G, Gabriel A, Matusz P, Loukas M (2014) Right lump kidney with varied vasculature and urinary system revealed by multidetector computed tomographic (MDCT) angiography. Surg Radiol Anat .
48. Pupca G, Micläuş GD, Bucuraş V, Iacob N, Sas I, et al. (2014) Left crossed fused renal ectopia L-shaped kidney type, with double nutcracker syndrome (anterior and posterior). Rom J Morphol Embryol 55: 1237-1241.
49. Calado AA, Macedo A Jr, Srougi M (2004) Cake kidney drained by single ureter. Int Braz J Urol 30: 321-322.
50. Türkvatan A, Demir D, Olçer T, Cumhur T (2006) Cake kidney: MDCT urography for diagnosis. Clin Imaging 30: 420-422.
51. Goren E, Eidelman A (1987) Pelvic cake kidney drained by single ureter. Urology 30: 492-493.
52. Rosenkrantz AB, Kopec M, Laks S (2010) Pelvic cake kidney drained by a single ureter associated with unicornuate uterus. Urology 76: 53-54.
53. Schwartz MJ, Bartolotta R, Brill PW, Kovanlikaya A, Hanna M (2010) Pelvic cake kidney with a solitary ureter and bilateral congenital absence of the vas deferens. Urology 75: 170-172.
54. Lal A, Pavunesan SK, Mahalingam H, Garg M, Sinha SK, et al. (2015) A triad of complete dorsal pancreatic agenesis, pancake kidney and bicornuate uterus. An association or an incidental finding: first case in literature. JOP 16: 189-191.
55. Bhimarao, Nagaraju RM (2015) Multidetector computed tomography urography in pancake kidney: A rare case. J of Evidence Based Med & Hlthcare 2: 2014-2017.
56. Tiwari AK, Choudhary AK, Khowal H, Chaudhary P, Arora MP (2014) Pancake kidney: A rare developmental anomaly. Can Urol Assoc J 8: E451-452.

This article was originally published in a special issue, entitled: **"Anatomy"**, Edited by Abdul Hafeez Baloch