

Incidentally detected pancake kidney: case report of an extremely rare congenital anomaly.

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Case Report

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Abstract

Background:

Congenital anomalies of urinary system are very common and have extremely varied presentation. Among them most rarely found structural anomaly is the pancake kidney. When both kidneys are fused along their medial surfaces to form a round shaped single renal mass is termed as Pancake kidney. In this case report, a pancake kidney was incidentally detected in a girl. The majority of subjects who have pancake kidney are usually symptom less but surgeons should be aware of coexisting malformation of other organs and its potential risk of developing malignancy.

Case presentation:

A 12-year-old young lady attended out-patient department with mild dull aching lower abdominal pain and dysuria. She had no history of fever, haematuria, menstrual abnormality, pelvic inflammatory disease or trauma. Urine examination showed traces of albumin and 10-12 pus cells/ HPF. She had normal kidney function test and digital X-ray of KUB region. A USG of the whole abdomen showed normal intra-abdominal organs except empty bilateral renal fossa. The Multi-Detector Computed Tomography(MDCT) scan of the whole abdomen revealed one round shaped mass measuring approximately 9 cm(vertical) X 10 cm(horizontal), in the pelvic cavity. That mass was finally identified as a pancake kidney. She was prescribed antibiotics based on urine culture, and sensitivity test that cured her symptoms. She was advised to follow-up regularly in Out-Patient Department to evaluate her kidney function and to rule out any neoplastic change.

Conclusions:

This condition can be managed conservatively, if the subject remains symptom less, by regular monitoring of renal function. Surgeon should remain alert for the development of infections, any obstructive manifestations leading to calculus formation and any malignant changes. The person should be careful in avoiding trauma to low-lying pelvic kidney. Extensive surgeries should be avoided and only selective procedures should be done so that the patient may lead a normal lifestyle.

Background

Congenital malformative uropathies, rank third among the most common congenital anomalies after cardiac and skeletal defects. Congenital anomalies of urinary system have extremely varied presentation and may involve the kidney, ureter, bladder or urethra. There may be different developmental renal anomalies, like renal agenesis, ectopic kidney and different fusion anomalies (most common being horse-shoe type kidney). Most rarely found structural anomaly (less than 10%) [1] is Pancake kidney. When kidneys are fused along with the medial surfaces of each pole forming a ring-like or doughnut shaped structure, it is called Doughnut Kidney. When both kidneys are extensively fused along the whole medial surface, a disc-shaped or shield shaped single renal mass is formed, which lacks any intervening

septum, be termed as Lump or Pancake kidney. Respective half of the kidney is drained by own collecting system which lacks connection with the opposite side. The pelvis of this renal mass is placed anteriorly and two ureters usually (in rare cases only single ureter) remain uncrossed and enter the urinary bladder following a normal but shorter path.

In this case report, a pancake kidney was incidentally detected in an otherwise healthy young lady who was initially found having bilaterally absent kidneys following ultrasonographic study during treatment of her urinary tract infection. The majority of subjects with Pancake kidney are asymptomatic but there may be associated malformation of other systems or organs. Although in present case no other abnormalities could be detected, and she was managed conservatively.

The aim of this case report is to get acquainted with the potential traumatic, iatrogenic, and possible neoplastic complications of apparently asymptomatic pancake kidney disease and literature was reviewed to explore possible etiologies and therapeutic strategies.

Case Presentation

A 12-year-old young lady with no significant previous medical or surgical history, presented in out-patient department with chief complaint of mild dull aching lower abdominal pain for last two months and dysuria for last seven days. She denied any history of fever, haematuria, menstrual abnormality, pelvic inflammatory disease or trauma. Physical examination revealed normotensive girl with Body Mass Index (BMI) of 19.5, without any significant finding. Her respiratory system and abdominal examination was unremarkable. Abdomen was scaphoid with umbilicus in the normal position. No intra-abdominal mass was noticed. There was normal bowel sound with no abdominal or renal angle tenderness. There was neither shifting dullness nor any guarding of abdominal muscles.

Microscopic examination of urine showed traces of albumin and 10-12 pus cells / HPF (High Power Field) without any RBC. She had normal kidney function test (urea -15.52 mg/dl, creatinine – 0.85 mg/dl) but confusion started when digital X-ray of Kidney, Ureter, and Bladder (KUB) region failed to demonstrate bilateral renal tissue shadows. It showed no KUB region calculus also. An ultrasonographic scan of the whole abdomen also added to the preexisting confusion by revealing no abnormality of intra-abdominal organs except empty bilateral renal fossa(Figure 1). Ultrasonography(USG) also failed to trace any one of the un-ascended kidneys in bilateral para-vertebral regions in that otherwise healthy girl. Finally, Abdomino-Pelvic Multi-Detector Computed Tomography (MDCT) scan [Figure 2,3] showed one round shaped mass measuring approximately 9 cm(vertical) X 10 cm(horizontal), situated in front of the sacral promontory in the pelvic cavity. That mass was finally identified as two ectopic kidneys malrotated postero-laterally and fused together in the medial aspect in mid line anterior to the third, fourth, and fifth lumbar vertebra (L3-L5,below the bifurcation of abdominal aorta) giving rise to a Pancake Kidney. The cortico-medullary differentiation was maintained. Parenchymal enhancement pattern was normal with excretion of contrast material was seen bilaterally. There was no evidence of calculus, obstruction or hydronephrosis. Short, uncrossed, non-dilated ureters were seen, anterior to renal mass, draining

separately into the urinary bladder. MDCT clearly showed both the vascular supply and urinary tract anatomy.

She was prescribed antibiotics based on urine culture, and sensitivity test that cured her symptoms (fever and dysuria). She was advised to follow-up regularly in urology Out-Patient Department to evaluate her kidney function and to rule out any neoplastic change.

Discussion And Conclusions

In 1938, Wilmer first described and classified renal fusion anomalies, which was refined and expanded by McDonald and McClellan in 1957[2]. According to them renal ectopia is of two types: Crossed type and simple/uncrossed type. Crossed Renal Ectopia may be of four types:

- i. Bilateral Crossed Ectopia of Unfused Kidneys.
- ii. Unilateral Crossed Ectopia of Unfused Kidneys.
- iii. Bilateral crossed ectopia of Fused Kidneys: commonest (90%).
- iv. Crossed Unfused renal ectopia.

Other rare fusion anomalies are sigmoid kidney, L-shaped kidney but rarely there is incidence of familial crossed ectopia. 3.3 – 11.1% of the population have congenital anomalies of the urinary system, which accounts for nearly 50% of all congenital abnormalities. The overall occurrence of ectopic kidney is 1 out of 400 autopsy cases and among them 85% had fused kidney.[3] In a study incidence of prenatal ultrasonography detected crossed renal ectopia was 0.003% in India. The commonest (incidence of 1 in every 700 autopsies[4] and 0.25% in the general population) renal ectopia is horseshoe kidney.

Pancake kidney is the rarest type of ectopic fused kidney disease with unknown occurrence. But Miclaus et al had calculated that 1 out of 65,000–375,000 population may get affected[5]. Pancake kidney has male preponderance (Male: Female ratio being 2.5:1). The condition may be detected at any age, although most common is 30–60 years[6]. Crossed Ectopic Kidney (second most common fusion abnormality) has varied presentation. In this situation, both the kidneys may be situated on the ipsilateral side in a fused manner (85%), in unfused manner (<10%), or in extreme rarity it may be bilateral or solitary. The left kidney more commonly (3 times commoner than the right kidney) migrates to the opposite side[7]. Commonly upper pole of the crossed ectopic kidney fuses with the lower pole of uncrossed kidney.

Mc Donald, Mc Clellan et al had classified Crossed Fused Renal Ectopia (in decreasing order of frequency) into six categories[8] like **[Figure 4]**:

- i. Type A: Inferior crossed fusion .Here the superior part of the ectopic crossed kidney fuses with the inferior part of the ipsilateral uncrossed kidney. Pelvis of both the kidneys may be located anteriorly.
- ii. Type B: S-shaped or Sigmoid kidney. Here the ectopic crossed kidney is placed inferiorly with pelvis directed laterally and the ipsilateral uncrossed kidney is placed superiorly with pelvis directed

medially. In this situation, pelvis of both kidneys is placed correctly as two kidneys fuses after completion of rotation on the vertical axis.

- iii. Type C: Lump kidney. Here,unilaterally, fusion occurs over a wide surface and pelvis of both kidneys are placed anteriorly .The ureter from the ectopically positioned kidney crosses the midline and it is placed more inferiorly than the ipsilateral one.
- iv. Type D: Tandem or L-Shaped kidney. Here the crossed ectopic kidney lies transversely and fuses partly with the inferior part of the ipsilateral uncrossed kidney.
- v. Type E: Disc kidney (Unilateral in position). Here both the kidneys are fused along the whole medial surface.
- vi. Type F: Superiorly fused and crossed kidney (least common type):Here the lower part of the crossed ectopically placed kidney fuses with the superior part of the ipsilateral uncrossed kidney . Pelvis of both the kidnes is anteriorly placed.

The ascent of primitive renal tissue (metanephric blastema) and ureteric bud starts in the 5thweek of IUL which completes at 9th week of gestational age. Congenital anomalies of renal fusion and ectopia occur if there is impaired lateral shift, deviation and internal rotation during the process of cephalic migration from the mid-pelvis to the abdomen due to

- i. Faulty ureteral bud development.
- ii. Under influences of teratogenic factors.
- iii. Aberrant renovascular phenotypes where abnormally positioned umbilical artery causes opposition of the metanephric blastema resulting in fusion anomaly. The retroperitoneal structures may impair fused renal mass to ascent cranially upto the lumbar position.
- iv. Cook and Stephens had proposed that abnormal flexion or growth of the developing hind-end may cause development of pancake kidney.[9]

So, Pancake kidney malformation is a result of extensive fusion of the medial surfaces of metanephric blastema during early intra-uterine life. The renal mass is commonly situated in the pelvis or at the level where aorta bifurcates[10]. Pancake kidney may get blood supply from branches of the abdominal aorta or from numerous branches of both the internal and external iliac arteries. Most commonly, inferior parts of the ectopic, fused kidneys are more medially rotated than superior parts. The pancake kidney is located commonly at the level of third to fifth lumbar (L3-L5) vertebra and lies in front of the great vessels. The parenchymatous or fibrous isthmus lies where the inferior mesenteric artery arises from the aorta.[11]

Urogenital anomalies are found to be linked with 9p tetrasomy and 9p trisomy, .Pancake kidney is often associated with other anomalies, such as un-descended testis or anomalous vas deferens, vaginal agenesis, cornuate (uni/bi)uterus, Fallot's tetralogy, spina bifida, agenesis of sacrum, caudal regression syndrome and strabismus.

Pancake kidney mostly has deformed and rotated collecting system. The shorter length of ureters cause obstruction and stasis leading to hydronephrosis, nephrolithiasis, and vesicoureteral reflux with recurrent infection of urinary tract. So, otherwise symptomless Pancake kidney, may present with vague lower abdominal pain, features of urinary tract infection like pyrexia, hematuria. Aneurysm of iliac vessel, amenorrhea and failure to conceive may also be encountered as extra renal manifestations. The Pancake kidney is supplied by one or multiple renal arteries (branches from distal aortic or iliac artery) and is drained by renal veins (tributaries of iliac vein or inferior vena cava). In case of single vascular supply gravid uterus, pelvic mass or trauma may lead to renal ischemia. Hypertension[12] may result from stenosis of aberrant renal arteries due to atherosclerosis of the aorta and iliac arteries in the process of ageing.

In spite of scarcity of reported cases of pancake kidneys, subjects with ectopic fused kidney anomalies are more inclined towards development of various primary malignancies, including renal cell carcinoma, Wilms tumor or rarely rhabdomyosarcoma.[13] Person with horseshoe kidney is nearly two times more prone to develop Wilms tumour in comparison to one with normal renal anatomy.

The diagnosis is always incidental. Excretory urography was used previously which is now replaced by ultrasonography, MDCT, CT urography and radio nucleotide scanning for better studies of the urinary system and renal vascular anatomy. Ultrasonography, is the primary modality for pre or post-natal diagnosis of kidney anomalies. CT urography with contrast enhancement (MDCT) is specially useful for studying urinary tract anatomy, which includes kidney parenchyma and collecting systems[14]. MDCT images can also be processed using multiplanar reconstruction methods, maximum intensity projection, volume rendering process which provide a three-dimensional imaging and helps in better diagnosis. Crossed fused ectopic kidney has no specific treatment guidelines[15]. Mere existence of pancake kidney does not herald progressive renal failure. Surgery is only indicated in case of confirmed failure or progressive derangement of kidney function due to urinary outflow obstruction/obstructive uropathy. The treatment is selective towards the associated problems like pyeloplasty for a pelvi-ureteric junction obstruction, bulking agent injection or ureteric reimplantation in case of vesicoureteral reflux. To avoid vascular injury, infarction, necrosis or postoperative renal failure [10], the fused renal mass should not be separated. Conservative management is indicated for symptomless subjects, after exclusion of coexisting anomalies, in the form of regular monitoring of renal function, remaining alert for the development of recurrent infections, any obstructive (viz. ureteropelvic junction) manifestations leading to calculus formation and any malignant changes. The person should be careful in avoiding trauma to low-lying pelvic kidney. In the case of pregnancy in female with pancake kidney, possible compressive effect by gravid uterus and possible trauma during child birth should also be kept in mind. During pelvic surgery, specially abdominal aortic surgery, the pancake kidney is also susceptible to iatrogenic trauma. As the blood supply to pancake kidney comes from aortic bifurcation or iliac vessels, the whole renal mass may suffer ischemia if proximal aortic cross-clamping is done[16].

To conclude, Pancake kidney is the rarest anomaly occurring due to extensive fusion of medial surfaces of both the kidneys which is commonly detected incidentally. There may be increased occurrence of

recurrent urinary tract infections or renal calculus formation, due to the chances of ureteral obstruction by rotated and anomalous collecting systems. The Pancake kidney can be managed conservatively, if the subject remains symptom less, by regular monitoring of renal function, remaining alert for the development of any malignant changes. The person should be careful in avoiding trauma to low-lying pelvic kidney. Extensive surgeries are avoided and only selective procedures should be done so that the patient may lead a normal lifestyle.

Abbreviations

MDCT –Multi-detector Computed Tomography, **CT** – Computed Tomography

Declarations

A. ETHICS APPROVAL AND CONSENT TO PARTICIPATE: Not applicable

B. CONSENT FOR PUBLICATION: Written consent to publish was obtained from the parent of the subject (minor) for the publication of all clinical details and images and the consent form is available for review by the editor of the journal.

C. AVAILABILITY OF DATA AND MATERIAL: Presented within the manuscript, please contact author for additional data requests.

D. COMPETING INTERESTS: The author declares that he has no competing interests.

E. FUNDING: No funding source/grant was available.

F. AUTHOR'S CONTRIBUTIONS: All work regarding this case report was solely done by SB, who is also the corresponding author. The author has read and approved the manuscript.

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Figures

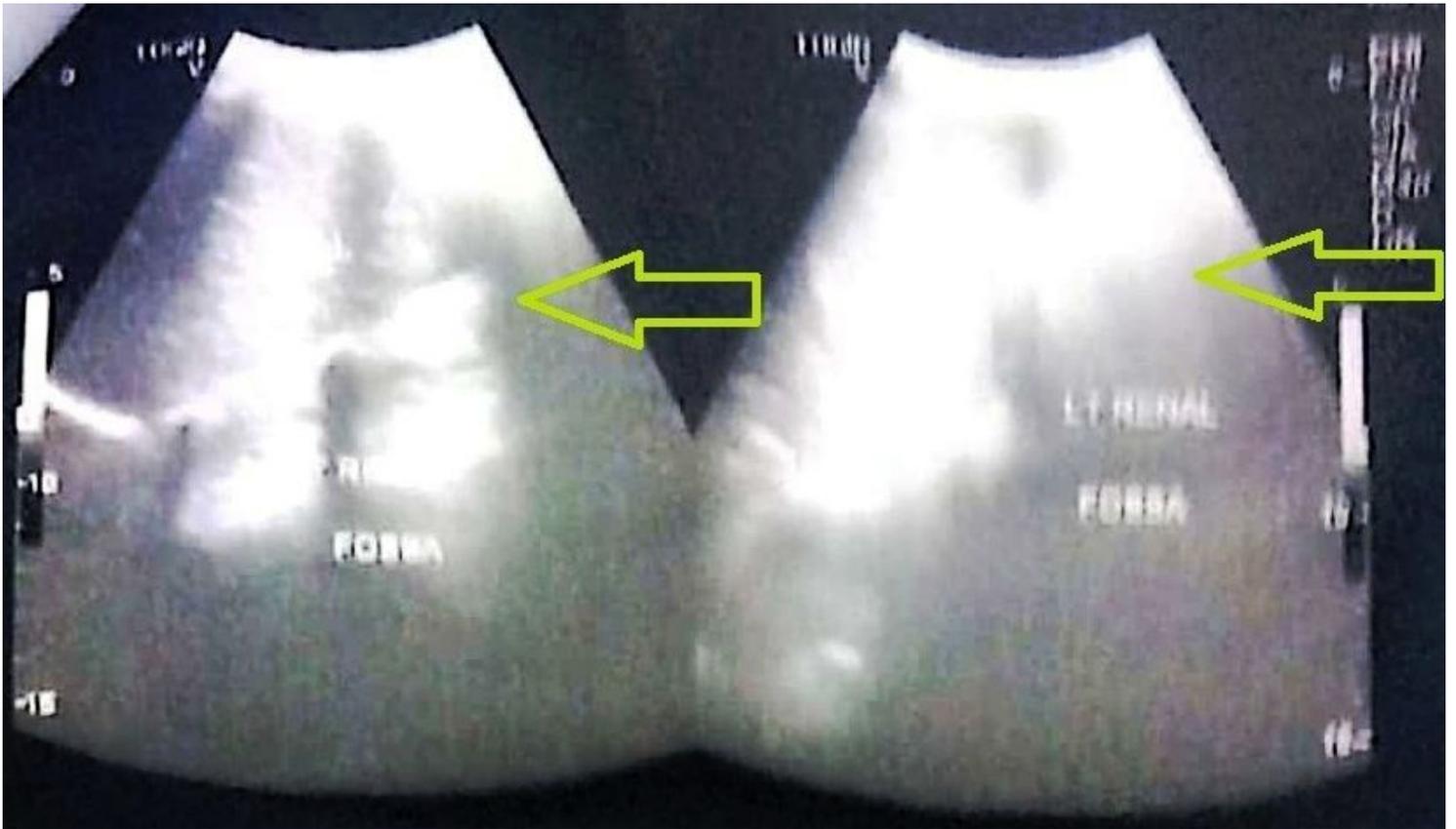


Figure 1

Grey scale Ultrasonography scan of the abdomen showing bilateral empty renal fossae(Green arrows).

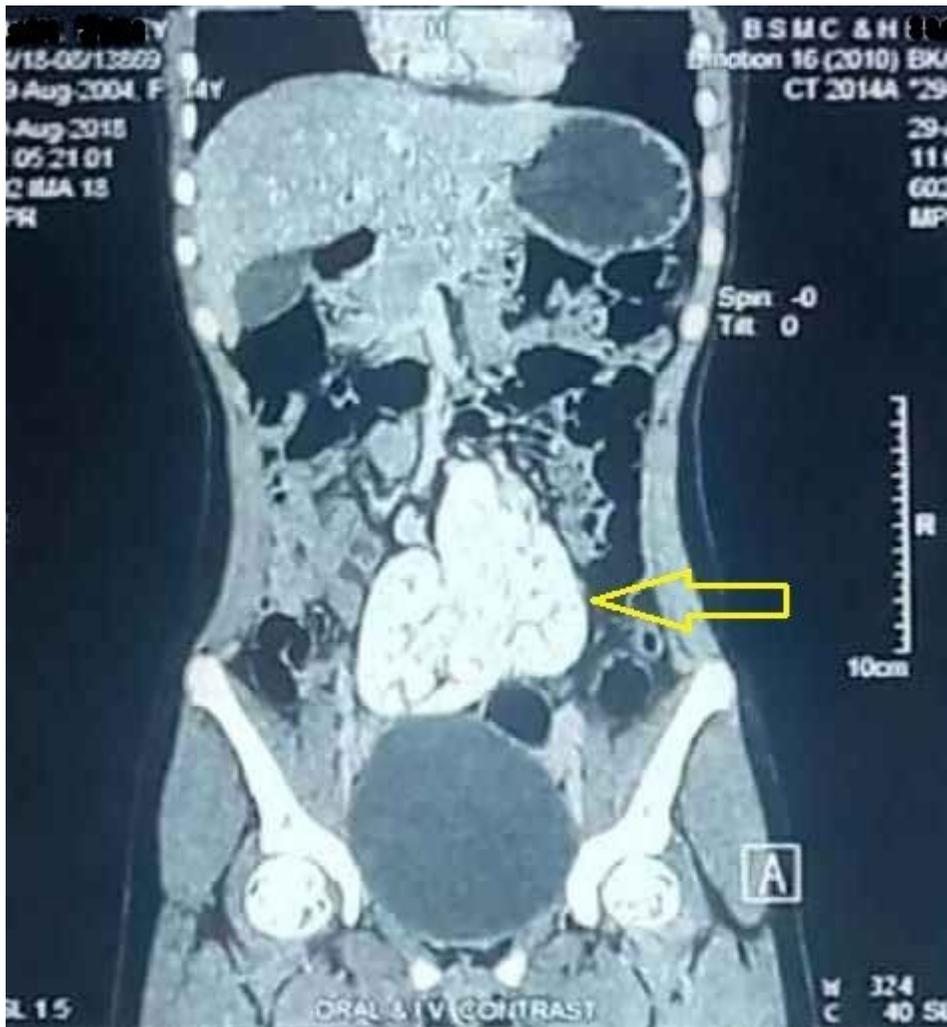


Figure 2

Contrast-enhanced computed tomography (MDCT) scan of abdomen showing: Pancake kidney is placed below aortic bifurcation in the pelvis with short ureters [yellow arrows in coronal section].

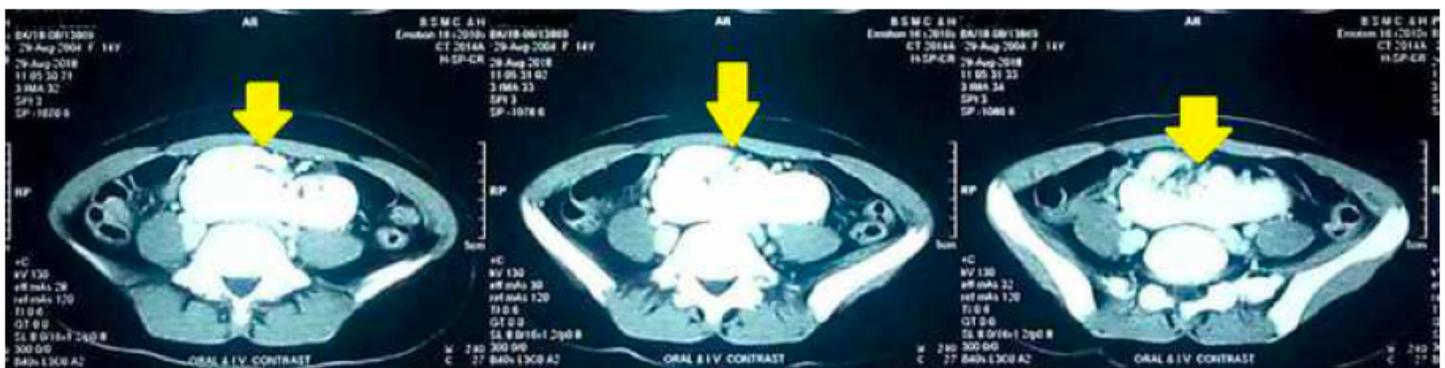


Figure 3

MDCT scan showing the centrally placed pancake kidney [yellow arrows in transverse section].

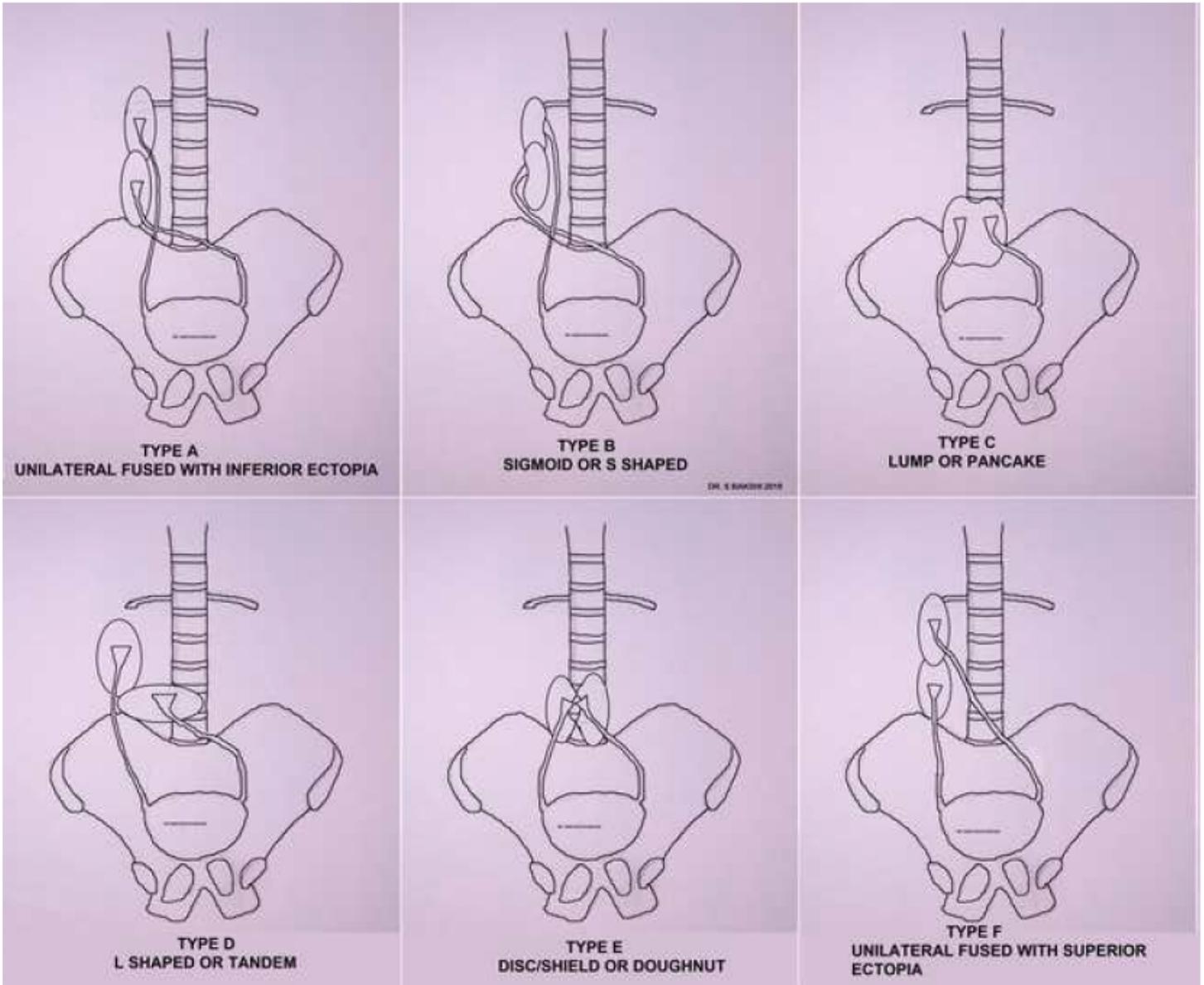


Figure 4

Illustration showing Mc Donald and Mc Clellan's classification of Crossed Fused Renal Ectopia.