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Congenital Positional Renal Anomalies - CT Based Pictorial Essay

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ABSTRACT

Background:
Congenital anomalies of kidney rotation, position, and fusion are very common and are frequently identified by chance. They can be considered as a "normal variant," as long as they do not cause complications. However the may be associated with urinary drainage impairment, stone formation, dysplasia, vesicoureteric reflux, infection, hypertension, other associated malformations and renal failure in childhood or as adults. Renal fusion anomalies often coincide with genital tract malformations due to shared embryological origins and can affect other organ systems like the skeletal, cardiovascular, gastrointestinal, and central nervous systems.

Objective:
This educational article aims to provide a comprehensive overview of the aetiology, classification, diagnosis, and clinical implications of these anomalies, enhancing the understanding of renal positional anomalies among healthcare professionals.

Conclusion:
Imaging plays a pivotal role in aiding in early detection, follow-up, surgical planning, and identification of complications and associated malformations. Early diagnosis and management, whether medical or surgical, are imperative to mitigate renal damage and delay or prevent progression to end-stage renal disease.

KEYWORDS: Congenital anomalies of the kidney and urinary tract, CAKUT, Horseshoe kidney, malrotation, renal ectopia, crossed renal ectopia, crossed fused renal ectopia.

INTRODUCTION

The definitive kidneys arise from the metanephros in the sacral region and ascend cranially to their final location in the lumbar Region due to differential growth of the anterior abdominal wall. Congenital anomalies of kidney rotation, position, and fusion are very common and are frequently identified by chance. They can be considered as a "normal variant," as long as they do not cause complications. However the may be associated with urinary drainage impairment, stone formation, dysplasia, vesicoureteric reflux, infection, hypertension, other associated malformations and renal failure in childhood or as adults Renal fusion anomalies often coincide with genital tract malformations due to shared embryological origins and can affect other organ systems like the skeletal, cardiovascular, gastrointestinal, and central nervous systems. Early diagnosis and management, whether medical or surgical, are imperative to mitigate renal damage and delay or prevent progression to end-stage renal disease. Imaging plays a pivotal role in achieving these objectives, aiding in early detection, follow-up, surgical planning, and identification of complications and associated malformations.[1,2]

RELEVANT EMBRYOLOGY:

Renal development begins at week 4 with the urogenital ridge splitting into the nephrogenic cord and the

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gonadal ridge. Three temporary forms emerge: the non-functional pronephros at week 4, followed by the mesonephros between weeks 6-10. Around week 5, the permanent metanephros forms, establishing their architecture with collecting tubule formation by week 6. The kidneys initially lie close together in the sacral region but gradually ascend to the lumbar region by the 9th week, while the renal arterial supply receives irrigation from progressively higher branches as they ascend.[1,3]

CLASSIFICATION:

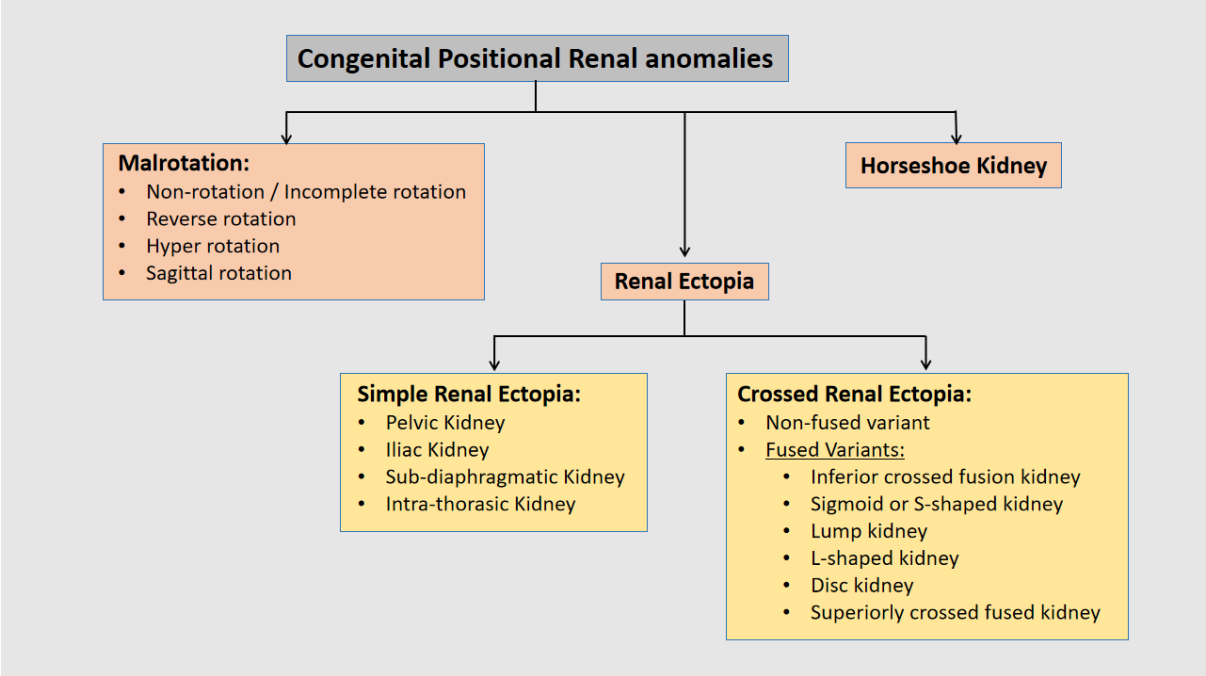

Fig. 1: Classification of congenital positional anomalies of the kidney

IMAGING MODALITIES:

Ultrasonography is typically the first imaging modality used due to its noninvasive nature, wide availability, low cost, and lack of ionizing radiation. However, its effectiveness can be limited by operator and patient factors, as well as incomplete visualization of the entire collecting system.

The use of intravenous urography has significantly declined in recent decades due to advancements in other imaging techniques.

CT and MRI are excellent for identifying anomalies in the upper urinary tract, providing detailed information on renal vascularity, adjacent structures, and complications such as renal stones, infections, and malignancies. CT and MR urography are increasingly preferred for their detailed visualization of the entire collecting system. MRI is particularly advantageous for paediatric patients, those requiring frequent imaging, and individuals allergic to iodinated contrast agents, as it avoids ionizing radiation and provides perfusion and excretion function information.[1]

Anomalies of Renal Position:

Anomalies of renal position stem from abnormalities in the migration of the kidneys from the pelvis to the lumbar region, a process starting around the 4th week and concluding by the 9th week of gestation. These anomalies comprise renal malrotation, simple renal ectopia, and crossed renal ectopia.[1]

Renal Malrotation:

Renal malrotation refers to an abnormal positioning of the kidneys concerning the hilum, which can manifest unilaterally or bilaterally and is more frequently observed in males. This condition typically arises when the

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ureteric bud inserts into an anomalous region of the metanephric blastema. Most patients with renal malrotation are asymptomatic, and are incidentally diagnosed during the assessment of abdominal pain or urinary issues.

The spectrum of renal malrotation includes:

- Nonrotation and Incomplete rotation (most common): The hilum positioned anteriorly or in an intermediate position between anterior and the normal medial position, with the ureter situated laterally.
- Reverse rotation: There is lateral rotation of the kidney such that the renal vessels cross the kidney anteriorly to reach the laterally positioned hilum.
- Hyper rotation: There is hyper rotation of the kidneys medially surpassing 180° but not reaching 360° resulting in a laterally located renal hilum. The renal vessels cross the kidney posteriorly to reach the hilum.
- Sagittal rotation: Rotation of the kidney around its hilum in the sagittal plane, aligning its long axis horizontally.[1,4]

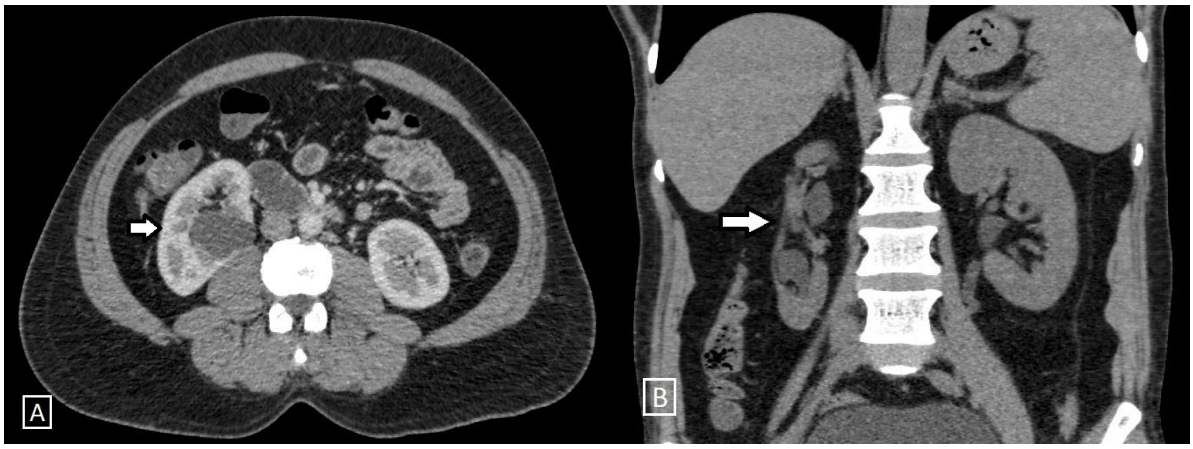

Fig. 2: (A) Post contrast axial section showing sagittal rotation of right kidney, wherein long axis of the kidney is seen in axial sections (white arrow). (B) Coronal reformatted plain CT section of a different patient showing malrotation of the right kidney with anteriorly oriented renal hilum (white arrow).

Renal Ectopia:

Ectopic kidney, or renal ectopia, denotes an abnormal kidney position resulting from its failure to ascend to the retroperitoneal renal fossa during embryonic development. This condition can manifest as simple (on the same side as the ureter) or crossed (on the opposite side of the ureteric orifice) and may occur unilaterally or bilaterally.

Simple Renal Ectopia:

Simple renal ectopia occurs in approximately 1 in 3000 cases and is bilateral in 10% of the cases. It is commonly located in the pelvis, iliac region, abdomen, or chest, often with varying degrees of malrotation and smaller size potentially leading to compensatory hyperfiltration and hypertrophy in the contralateral kidney. Blood supply typically originates from the iliac or infrarenal abdominal aorta, often with multiple arteries.

Types of simple renal ectopia include:

- Pelvic kidney: (most common) Located opposite the sacrum and below the aortic bifurcation and is prone to poor drainage.
- Iliac kidney: located in the iliac fossa near the sacral promontory above the iliac vessels
- Intrathoracic kidney: Herniated into the thoracic cavity through a diaphragmatic defect.
- subdiaphragmatic kidneys: Located at a level higher than the normal position just below the diaphragm.

Intra-thoracic sub-diaphragmatic forms represent less than 5% of cases, showing a male predominance and

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left-sided predominance, likely due to anatomical barriers.

Simple renal ectopia typically presents with normal function and no symptoms, often incidentally discovered during imaging. However, there's a heightened risk of hydronephrosis due to kidney malrotation and anteriorly positioned renal pelvis, potentially leading to impaired urinary drainage, infections, renal stones, and susceptibility to traumatic injuries due to their abnormal retroperitoneal location.[1,2,5]

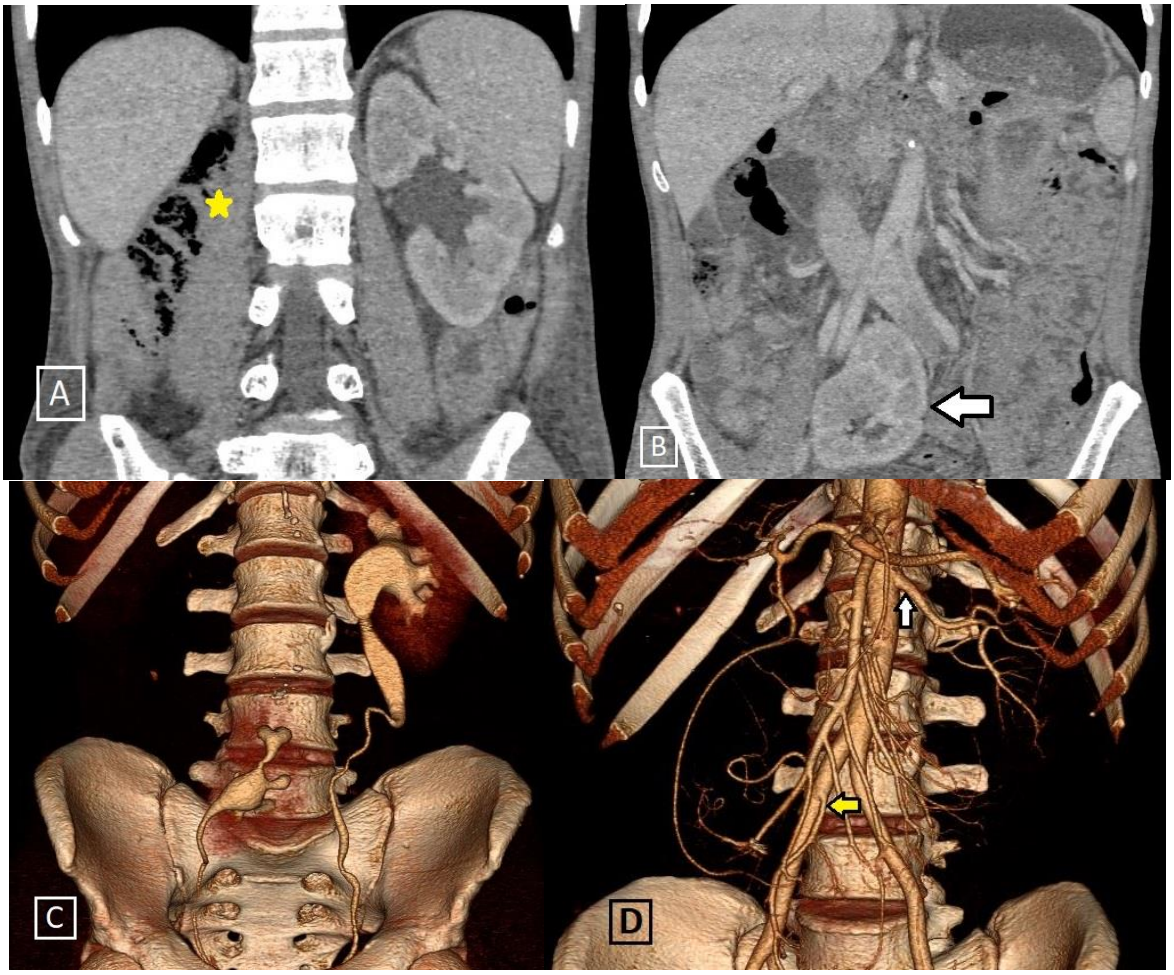

Fig. 3: Post contrast coronal reformatted sections (A, B) showing empty right renal fossa. Right kidney is seen at a lower level in the midline (white arrow). (C) 3D reconstructed images showing a well developed pelvicalyceal system opening into the bladder on the trite side. (D) The ectopic kidney is supplied by an artery arising from the right common iliac artery (yellow arrow). Normal origin of left renal artery seen (white arrow).

Crossed Renal Ectopia:

Crossed renal ectopia (CRE) denotes the displacement of a kidney to the opposite side from its embryological position, while the ureter maintains its normal insertion into the bladder. This relatively rare condition, occurring in approximately 7.5 out of 10,000 newborns, exhibits a male-to-female predominance of three to two. The anomaly arises from the abnormal migration of the ureteric bud and metanephric blastema across the midline during embryonic development. Factors contributing to this phenomenon include the position of the umbilical artery or overbending and rotation of the embryo.

CRE is of two types: Fused and Non-Fused variants.
The crossed kidney is fused to the normally positioned kidney in most cases (approximately 90%), with an incidence of one in 2000 cases.

Six forms of fusion described in decreasing order of frequency:

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- Inferior crossed fusion kidney: Fusion occurs between the upper pole of the ectopic kidney and the lower pole of the normal kidney.
- Sigmoid or S-shaped kidney: The ectopic kidney is positioned inferiorly, with its pelvis directed laterally, while the normal kidney has its pelvis turned medially.
- Lump kidney: Both kidneys are completely fused, forming a lump on one side.
- L-shaped kidney: The ectopic kidney lies transversely and inferiorly to the normal kidney.
- Disc kidney: Kidneys are fused along the medial edge of each pole.
- Superiorly crossed fused kidney: The ectopic kidney is positioned superiorly, with its lower pole merging with the upper pole of the normal kidney.

While most cases of CRE are asymptomatic and discovered incidentally, they can potentially lead to complications such as hydronephrosis, infections, vesicoureteral reflux, or nephrolithiasis, often associated with other congenital abnormalities.[1,6,7]

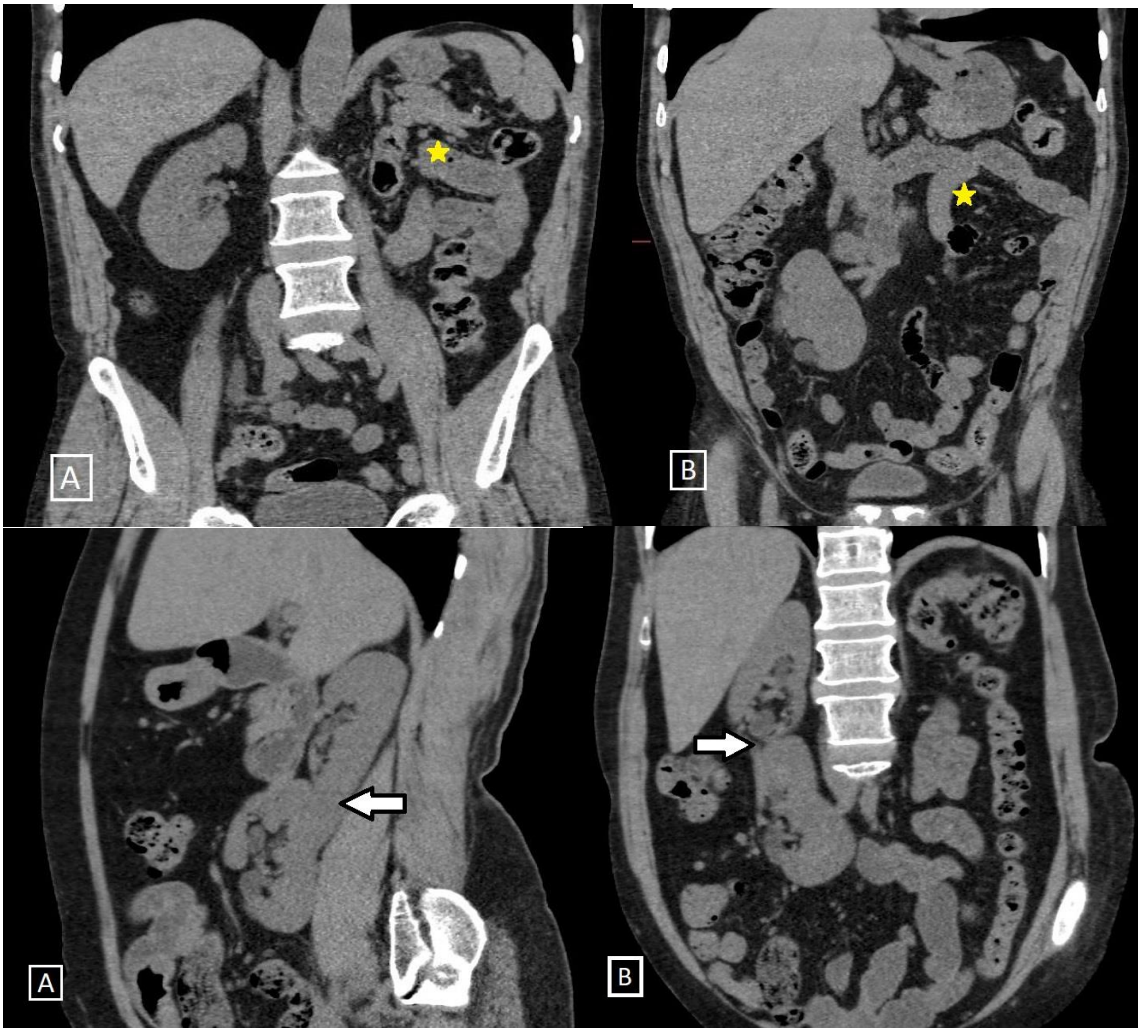

Fig. 4: Plain CT coronal reformatted sections (A, B) showing empty left renal fossa. Two kidneys seen separately on the right side. Sagittal and coronal reformatted sections (C, D) of a different patient shows empty left renal fossa. Two kidneys seen on the right side with fusion of the lower pole of the upper kidney with the upper pole of the lower kidney (white arrow).

Horseshoe Kidney:

Horseshoe kidney, a rare congenital fusion anomaly affecting roughly 1 in 400-600 individuals.There are two main theories that attempt to explain this condition.

- Mechanical fusion: Abnormal foetal development, potentially involving the spine or pelvic organs around week 4 of embryogenesis, may cause the developing kidney to come into contact and fuse due

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to the lack of a renal capsule at this stage. As these kidneys ascend to their final position, the fused tissue (isthmus) gets trapped under the inferior mesenteric artery, hindering further ascent and rotation.

- Abnormal migration of renal cells: Leading to a parenchymal bridge between the kidneys.

The fusion point can vary, with the lower poles being the most common site (90% of cases). Upper poles or both can also be involved. The ureters and pelvis typically face anteriorly. The isthmus can be composed of functioning kidney tissue or a fibrous band depending on the extent of fusion. Blood supply to the fused kidneys is variable, potentially arising from the aorta, iliac arteries, or less commonly, the middle sacral and hypogastric arteries.

Majority of the individuals experience no symptoms and are diagnosed incidentally. However, potential complications include ureteropelvic junction obstruction (UPJO), lithiasis which occurs in 20-60% of cases, and an increased risk of renal infections. There's also a rare but elevated risk of developing various malignancies like renal cell carcinoma, Wilms tumour, and carcinoid tumours.

Ultrasound is the first-line imaging tool, however, with poor visualisation of the bilateral lower poles prompting further investigation for abnormal tissue (the isthmus) anterior to the aorta. Misinterpretations can occur without proper awareness, leading to underestimation of kidney length or mistaking the isthmus for a mass. CT and MRI provide a more detailed evaluation of the collecting system, blood supply, and the nature of the isthmus (functioning tissue vs. fibrous), aiding in early detection of potential complications.[1,2,8]

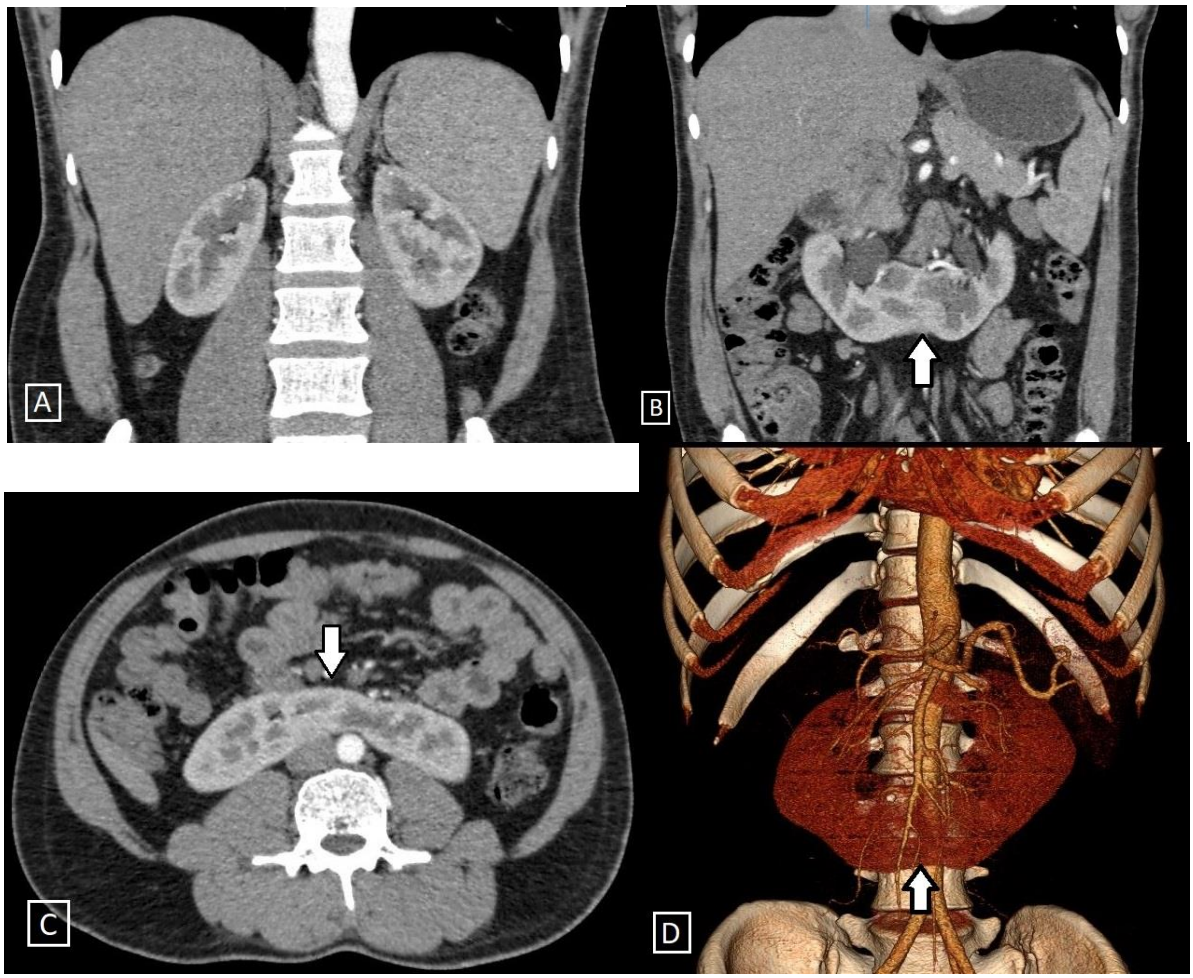

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Fig. 5: Post contrast coronal (A,B) reformatted, axial (C) sections and 3D reconstruction (D) of the same patient showing fusion of lower poles of left and right kidneys in the midline while the upper poles remain separate (white arrow).

CONCLUSION:

Congenital anomalies of kidney rotation, position, and fusion are very common developmental disorders that can lead to significant complications in adulthood, including stone formation, infections, hypertension, and renal failure, and may be associated with genital tract malformations.Specific anomalies such as renal malrotation, simple renal ectopia, crossed renal ectopia, and horseshoe kidney present unique challenges and potential complications, though many are asymptomatic and found incidentally.Imaging is crucial for diagnosis and management, with ultrasonography as the initial noninvasive method. However, advanced techniques like CT and MRI provide detailed visualization of renal anatomy and function, particularly beneficial for pediatric patients and those needing frequent imaging.Early detection and appropriate imaging are essential for effective management, reducing complications, and improving patient outcomes.

AVAILABILITY OF DATA AND MATERIAL:

The data are taken solely from our institution: Meenakshi medical college Hospital and Research Institute, Kanchipuram, Tamil Nadu.

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CONFLICT OF INTEREST:

None declared.

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