

Renal anomalies of position, shape and fusion: radiographic analysis

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ABSTRACT

Among the anomalies of the upper urinary tract are the anomalies of ascent, which includes simple renal ectopia, and the anomalies of form and fusion, which includes crossed renal ectopia with and without fusion and horseshoe kidney. Since these pathologies are frequently asymptomatic, the diagnosis is most commonly made after a routine imaging studies due to other causes or when recurrent UTI or urinary obstruction are associated.

The radiologic findings of the cases registered in two of the largest and of major national reference are described.

Key words: renal ectopia, horseshoe kidney, anomalies of the upper urinary tract, renal anomalies, imaging studies.

RESUMEN

Entre las anomalías del tracto urinario superior se encuentran las anomalías de la migración, que incluye a la ectopia renal simple, y las anomalías de la forma y fusión, que incluyen a la ectopia renal cruzada con y sin fusión y el riñón en herradura. Habitualmente estas patologías son de curso asintomático, por lo que su hallazgo suele ser casual durante exploraciones radiológicas rutinarias por otras causas ó en medio del estudio de los infrecuentes casos sintomáticos caracterizados por la aparición de infecciones a repetición o de clínica de uropatía obstructiva.

Se describen los hallazgos radiológicos de los casos registrados en dos de los centros de Radiodiagnóstico más grandes y de mayor referencia nacional.

Palabras clave: ectopia renal, riñón en herradura, anomalías del tracto urinario superior, anomalías renales, estudios de imagen.

• DEFINITIONS

- simple renal ectopia (ERS) or ectopic kidney (RE) ipsilateral: a kidney that is on the same side of the body that bore its corresponding ureter, but in an abnormal position (outside flank, L1 - L3) . It may be unilateral or bilateral.
- crossed renal ectopia (ERC): one or both kidneys cross the midline, reaching opposite the corresponding ureteral orifice. It may be unilateral or bilateral and can also occur with or without fusion contralateral kidney.
- Horseshoe kidney (RH): consists of two distinct renal masses lying vertically on either side of the midline and fused in their lower poles by an isthmus parenchymal

or fibrous tissue that crosses the median plane of the body.

Embryology and Pathophysiology

Congenital renal anomalies in the position and in the renal fusion are the result of impaired cephalic migration from the pelvis to the flank of the ureteric bud and metanephric blastema, a process that begins in the fifth week of gestation and ends in the ninth week.^{1, 2}

SIMPLE RENAL ECTOPIA: Clinical Features And Image

- • The incidence is 1:900, no differences regarding sex. It shows a slight left dominance. Bilateral involvement is rare (10% of cases) 1.2 (Figure 1 and Figure 2)

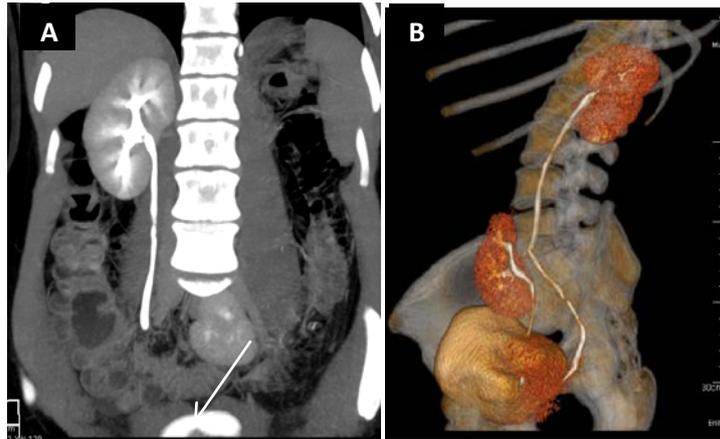


FIGURE 1. ERS unilateral in a woman of 20 years old. (A) MPR coronal excretory phase. Note the empty left flank and iliac position left kidney (white arrow). Orthotopic right kidney. (B) Oblique sagittal reconstruction CT volume. RE Note the lower ureter insertion length and normal bladder.

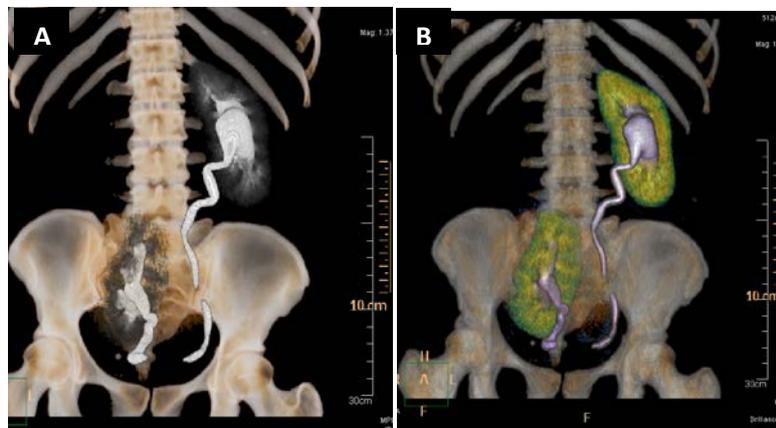


FIGURE 2. Bilateral ERS a woman of 46 years. (A) and (B) coronal CT volumetric reconstruction, excretory phase. Empty right renal fossa, ectopic right kidney iliac or lumbar and abdominal ectopic left kidney. Note the right renal axis vertical orientation and shorter right ureter and altering the rotation of both kidneys with prior arrangement your pelvis.

- The location, in order of frequency, may be pelvic, iliac, abdominal or chest.
- The RE usually smaller and with presence of fetal lobulations.
- The kidney is slightly medial axis or vertical, can even be horizontal.
- The pelvis is usually prior to the parenchyma (instead of medial).
- The ureter has a length according to the location of the kidney.
- vascularization comes from adjacent vessels and multiple arteries typically found. (Figure 3)
- colon malposition is observed in cases of lumbar and pelvic RE.
- The RE classification is based on the kidney in the retroperitoneal position:
 - pelvic or sacral sac opposite, below the aortic bifurcation and ileopectinea line.
 - lumbar or iliac: located in the iliac fossa near the sacral promontory above the iliac vessels.
 - Abdominal: occupies a position superior to the iliac crest, adjacent or below L2.
 - cephalic, cranial or higher: located above the normal position with the hilum subdiaphragmatic level T10.
 - Chest: kidney herniated into the chest through the triangle hole lumbocostal or Bochdalek.

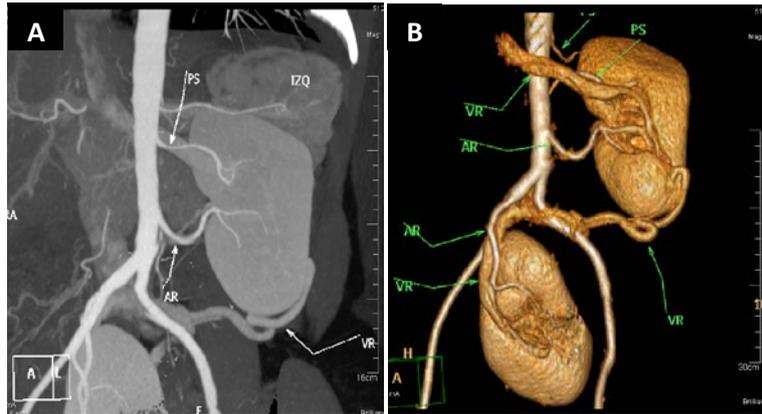


FIGURE 3. Bilateral ERS a woman of 46 years. (A) CT angiography of renal vessels. (B) Volumetric reconstruction. (AR: renal artery VR renal vein PS: polar superior artery.) Note the origin of the right AR near the aortic bifurcation; also left PS and variations in the number and arrangement of the venous tree.

CROSS RENAL ECTOPIA: Clinical Features And Image

The incidence of CKD is 1:2000 and the merged range is 1:1000 live births.

More common in men (2:1) and left-right ectopia is three times more common than right-left ectopia. (Figure 4)

With melt (85 - 90%), without melting (10-15%). (Figure 5 and Figure 6)

The arterial supply usually crossed kidney is abnormal, variable and unpredictable. This finding is also common in the normal kidney. (Figure 7)

- Classification of McDonald and McClellan:

a. ERC Fusion

- Lower: Lower crossed the orthotopic kidney, upper pole fused with normal lower pole. The most common form. (Figure 6)
- Sigmoid (S-shaped) cross the kidney is inferior, fused by its adjacent poles. The lower convex edge of a kidney is directly opposite to the outer edge of its counterpart. The normal kidney ureter courses down before the outer edge of the lower kidney and the ureter

crosses the midline RE before entering the bladder.

- On "cake": kidneys fused together along the inner side edges. Both predate pelvis and parenchyma drain different areas. There ureteral junction.
- L-shaped: cross the kidney makes a transverse orientation orthotopic kidney, which merges at its lower pole.
- discoid or donates: both kidneys completely fused along the medial concave edge of each pole resulting in a renal unit or disc-shaped ring (depending on the degree of fusion), the pelvis and there are prior ureteral junction. Each collecting system drains corresponding parenchyma. (Figure 8)
- Higher: kidney crosses the midline in is above the orthotopic kidney and fused to the upper pole of the latter through the lower pole. The pelvis predate. Rare.

a. ERC without fusion (Figure 4 and Figure 5)

b. ERC lonely

c. ERC bilateral

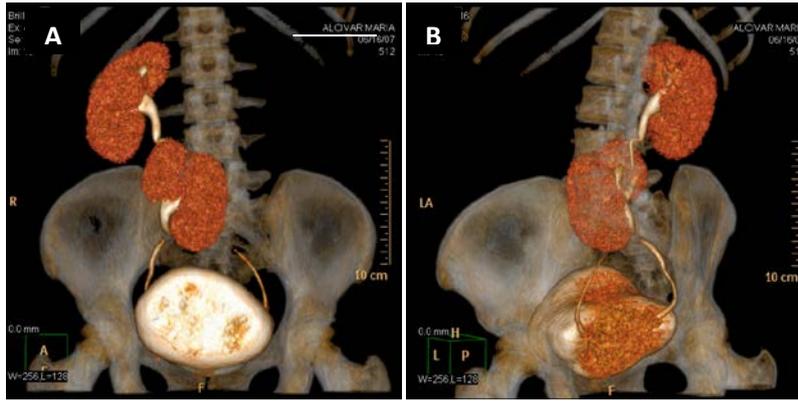


FIGURE 4. ERC without fusion in a woman of 57 years old. Volumetric reconstruction in CT. (A) Coronal. Empty left flank and left RE contralateral lumbar position, which does not contact the right renal unit. (B) Sagittal oblique. Note ureteral junction thereof with preserved bladder insertion.

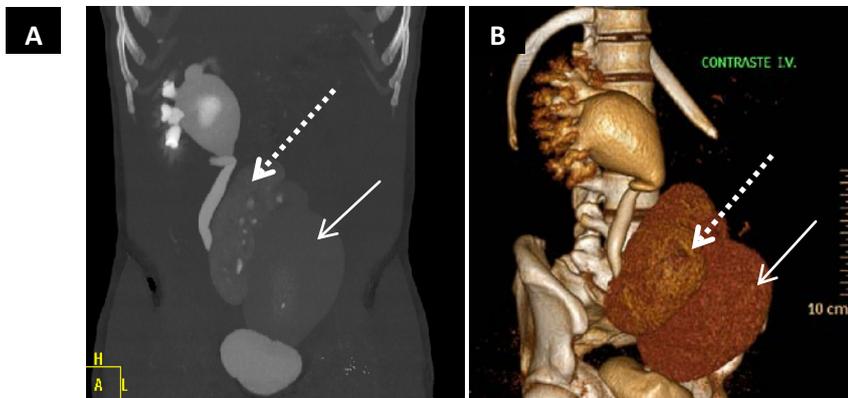
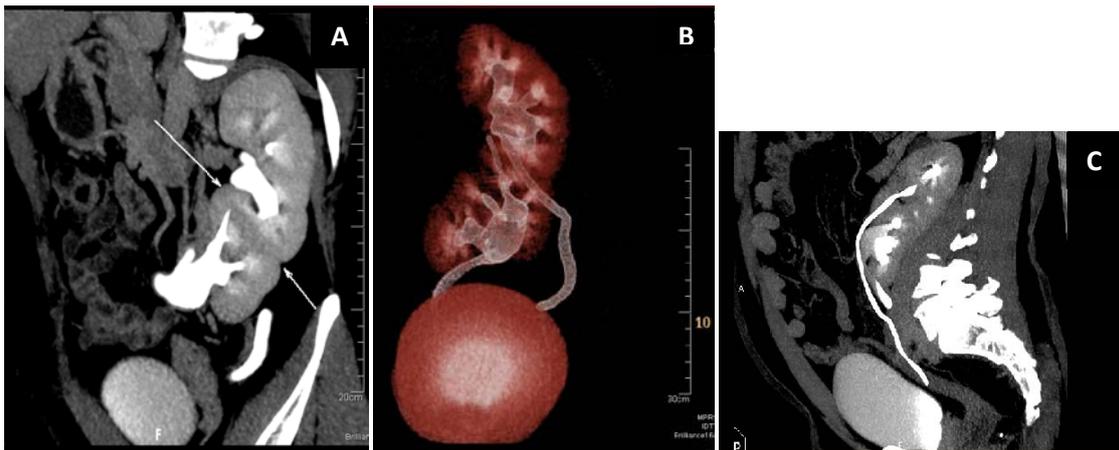


FIGURE 5. Left-right ERC without fusion in a man of 45 years. (A) MPR and coronal (B) CT volume. Note the change in the position and rotation of both kidneys, as well as large dilated ureter RE.



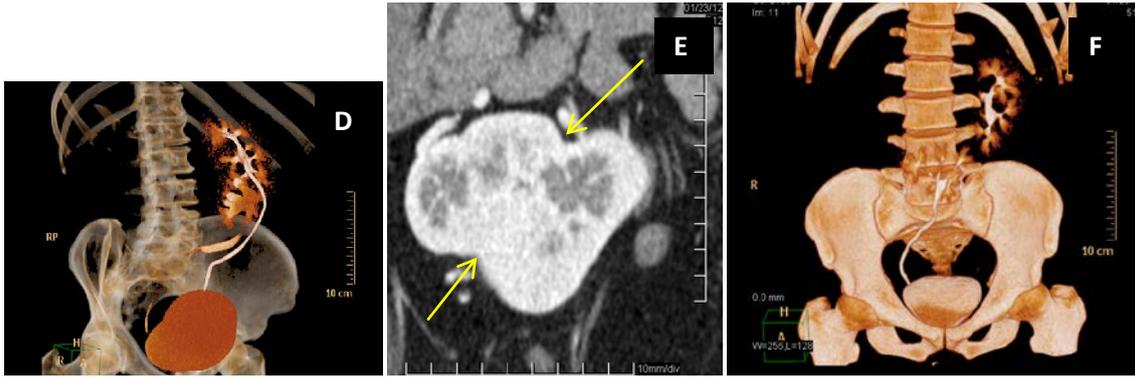


FIGURE 6. Right-left ERC Fusion (RE lower) 30 year old woman (A and B) of a 43 years (C and D) and a man of 34 years (E and F). (A) Oblique coronal MPR, excretory phase. Note the fusion of the upper pole of the RE with orthotopic kidney lower pole (white arrows). (B) Oblique volumetric CT Reconstruction. Note the independence of the two collecting systems. (C) MPR sagittal oblique excretory phase and (D) volumetric oblique coronal CT. Note the independence of collecting systems. (E) Oblique axial MPR, corticomedullary phase. Note the fusion of the renal parenchyma (yellow arrows). (F) Coronal volumetric reconstruction of TC. Shows the full right collecting system and left collecting system, it is observed that the ureter side, but there was a separate drainage.

HORSESHOE KIDNEY: Clinical Features And Image

- The most common congenital anomaly of the urinary tract (1:400 births).
- predominates in males (2:1).
- Fusion may be in the midline (melting symmetrical, 90%) or lateral (asymmetric fusion, 10%).
- In 95% of cases fusion is through the lower poles. (Figure 9)
- They are in a lower than normal situation and incompletely rotated. Renal axis is vertical or oblique medial side.
- The pelvis and ureters are anterior, ventrally crossing the isthmus.

- The calices are normal in number and atypical orientation (rear).
- The isthmus usually lies ahead of L3 or L4, below the emergency inferior mesenteric artery. It is often the ventral aorta and vena cava.
- The vascular supply of these kidneys is highly variable, particularly in the isthmus. Observed with multiple arteries that originate from the aorta or other major vessels. (Figure 10)
- 60% of cases are asymptomatic in the first decade after diagnosis.

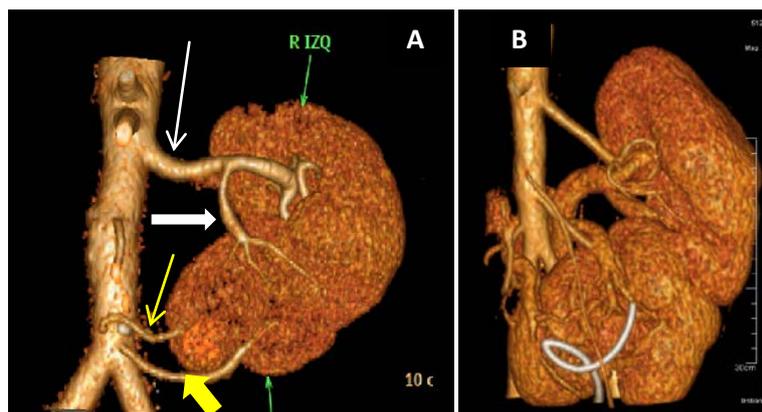


Figure 7. Right-left ERC Fusion (lower RE) in a 36 years. Volumetric reconstruction of TC. (A) single left renal artery (thin white arrow) that emits a branch to the RE (thick white arrow). RE renal artery arises from the anterior aorta just above its bifurcation (gruesaa yellow arrow), there is also a polar aberrant artery (arrow amarille thin). (B) Note the variations in the venous tree. Displays also part of a double J catheter in the lower collecting system was placed to resolve stone disease.

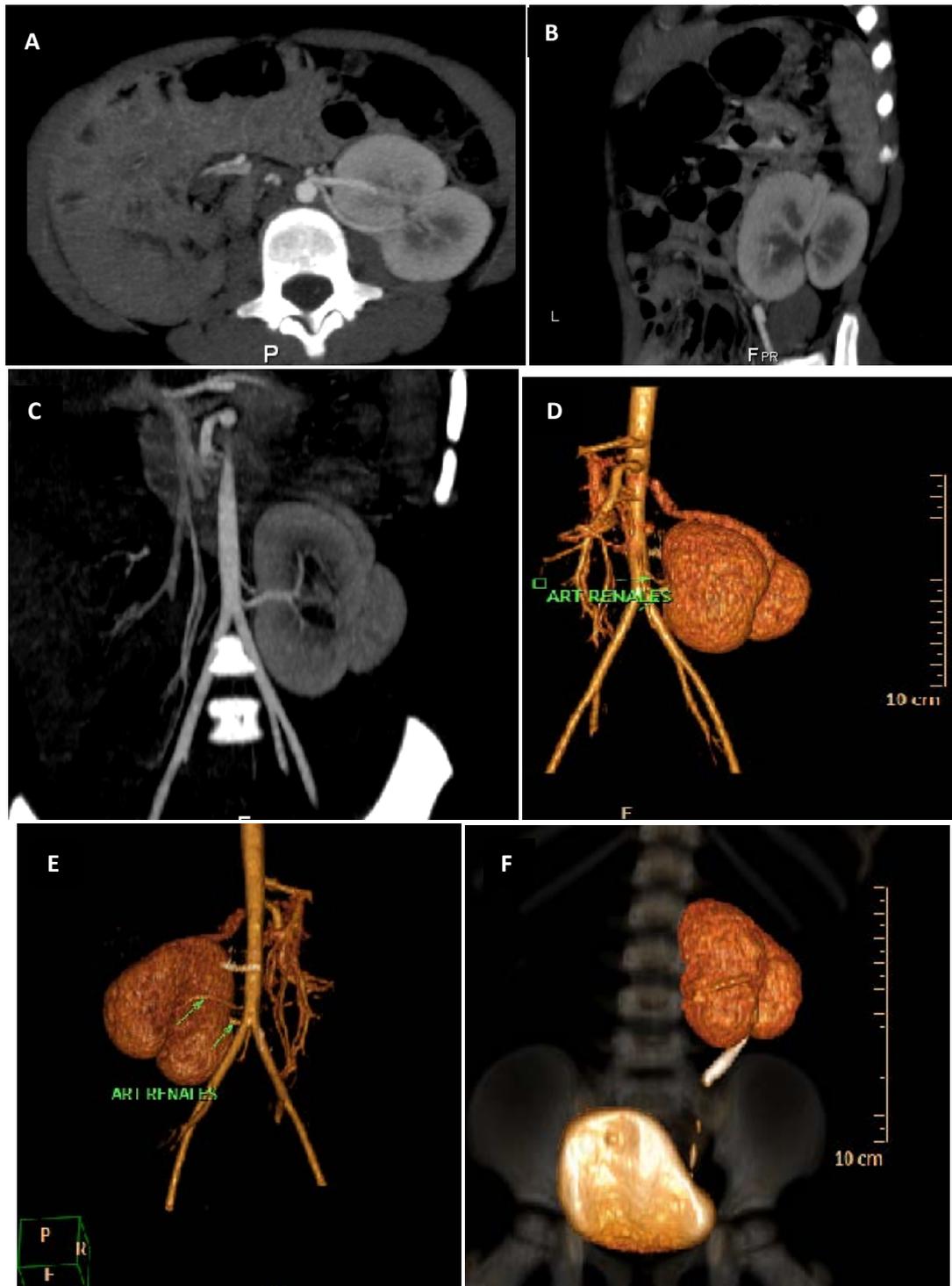


Figure 8. Right-left ERC Fusion (kidney disk) in a 23 years. (A), (B) and (C) MPR axial, sagittal, coronal oblique slant (respectively) corticomedullary phase. Merging observed renal units through their medial edges (hilum) resulting in a disk-shaped mass as well as the aortic origin independent of each renal artery. (D) and (E) Volumetric reconstruction and rear coronal (respectively). Note the arterial and venous tree consists of multiple aberrant vessels. (F) Volumetric reconstruction anterior oblique. Kidney shows that fused drain through a single manifold system (rare).



FIGURE 9. Rh Reconstruction MPR coronal corticomedullary phase of a man of 23 years. Merging of the two poles in the midline inferior, leading to functional parenchyma composed isthmus. Note renal axis vertical direction.

DIFFERENTIAL DIAGNOSIS

- renal ptosis or nephroptosis: the kidney initially occupies its usual place, but down in relation to its position in the body, is characterized by a normal vasculature and ureter normal length tortuous. Should be investigated organomegaly or retroperitoneal masses that can cause kidney displacement.
- • Graft renal kidney in the iliac fossa, the renal vessels are anastomosed to the external or internal iliac vessels and the ureter reimplemented into the bladder through a submucosal tunnel, variable axis of the renal pelvis.
- • Autologous renal kidney surgical repositioning of the patient.

- Nephrectomy ipsilateral colon remain in its normal position.
- acquired renal atrophy: kidney smaller proper position.

PROBLEMS ASSOCIATED

Simple renal ectopia:

- It is usually asymptomatic, but in some patients may be associated with hydronephrosis (56%) or vesicoureteral reflux (30%) .1
- It is associated with genital abnormalities by 15% .1
- • They are also associated with malformations of other organs (genitals, skeleton) and part of various multiple malformation syndromes.



FIGURE 10. Horseshoe kidney. Oblique coronal MPR, cortical phase of a man of 23 years. (A) The hilar renal arteries (ARH) arise from the aorta and direct the corresponding kidney. (B) Note the presence of multiple bilateral aberrant inferior polar arteries (PI) leading to the isthmus.

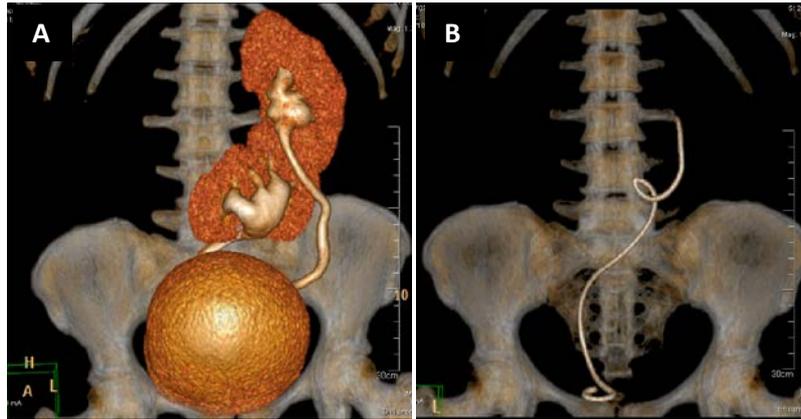


FIGURE 11. Right-left ERC Fusion (lower RE). Man of 36 years with a history of left ureteral lithiasis and endourology resolved by double-J catheter placement (A) Reconstruction of Volumetric CT observed in the fusion of the upper pole of the lower pole RE with orthotopic kidney. (B) Double J catheter in the left collecting system.

Crossed renal ectopia:

- vesicoureteral reflux (VUR) in 20% and 70% for ERC bilateral.
- In the RE is cystic dysplasia, ureteropelvic junction obstruction (29%), reflux (15%) or, in rare cases, carcinoma.
- In cases of ureteropelvic junction obstruction increases the risk of urolithiasis. (Figure 11)
- The ERC solitary genital abnormalities associated with skeletal or by 40 and 50%, respectively, as well as imperforate anus by 20%.

Horseshoe kidney:

Conflict of interest

None reported by the authors

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- ureteropelvic junction obstruction (33%), with consequent hydronephrosis or stones.
- It has reported a slightly increased incidence of Wilms' tumor, hypertension and adenocarcinoma.
- It is observed in 3% of cases of neural tube defects, 20% of trisomy 18 and 60% of Turner syndrome.

RECOMMENDATIONS

The detection of these anomalies requires finding other associated renal and extrarenal malformations. Periodic evaluation will avoid possible complications.

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