

Peer Reviewed **Article of Interest****A PICTORIAL PRESENTATION OF THREE KIDNEY ANOMALIES IN COMPUTED TOMOGRAPHY**

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Abstract

This pictorial presentation is a range of computed tomography (CT) images of fusion and developmental abnormalities of the kidney. A brief discussion of each abnormality is presented.

Keywords. congenital abnormalities, ectopic kidney, horseshow kidney, isthmus, polycystic

INTRODUCTION

The kidneys begin their development in the sacral region. As the abdomen and pelvis grow, the kidneys gradually ascend to the lumbar region and separate.^[1] By nine weeks gestation they are at their normal anatomical level adjacent to the adrenal gland. Any interruption of this normal migration will cause positional and fusion abnormalities.

Kidney abnormalities and diseases may commence before a foetus is born. Multiple conditions may present throughout the lifetime of the individual, which may vary from congenital anomalies to reflux, infection, calculi, tumour and chronic renal failure. It is important to know the normal anatomy of the kidneys to identify fusion and developmental abnormalities.

The kidneys normally lie on either side of the vertebrae in

the abdomen. They are paired retroperitoneal bean-shaped organs. There are three distinct spaces in the retroperitoneum: the anterior and posterior para-renal spaces and the perinephric space. The latter has an inverted cone shape, which contains the kidneys, proximal ureters, adrenal glands, vessels and lymphatics.^[2] As shown in Figure 1a (i, ii), the upper poles of the kidneys are deviated medially and the lower ones laterally.^[3] The right kidney is below the liver and lower than the left kidney. It is important to also check which kidney is visualised on sagittal images. On a right sagittal image, the right kidney is below the liver, as shown in Figure 1b (i). The left kidney is below the spleen in a left sagittal image, as shown in Figure 1b (ii). The renal artery, vein, ureter and nerves enter the kidney via the hilum at the level of the first lumbar vertebra.^[1] Figure 1c shows the renal arteries below the renal vein.^[4]

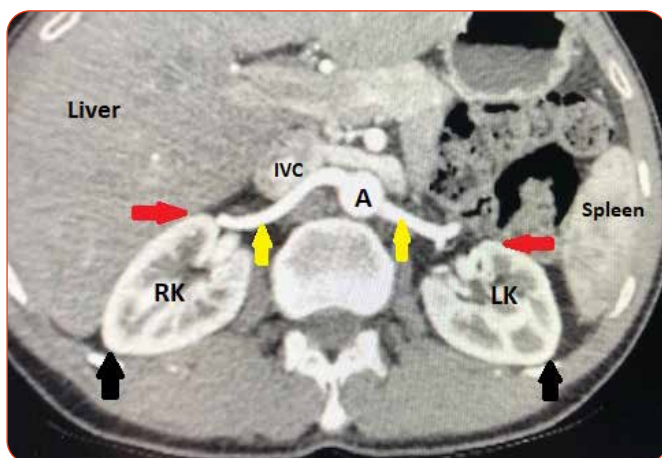


Figure 1a (i). Enhanced axial CT abdomen scan showing the right kidney (RK) and the left kidney (LK). The renal arteries (yellow arrows) arise from the aorta (A). The upper poles of the kidneys are deviated medially (red arrows) and the lower ones laterally (black arrows). Inferior vena cava (IVC).



Figure 1a (ii). Unenhanced axial CT abdomen scan showing right kidney (RK), left kidney (LK) and aorta (A).



Figure 1b (i). Right sagittal view showing the right kidney (RK) below and behind the liver.



Figure 1b (ii). Left sagittal view showing the left kidney (LK) below the spleen.



Figure 1c. Axial CT scan showing right kidney (RK). Red arrow indicates the renal vein of the kidney (LK), and the yellow arrow indicates its renal artery.

ECTOPIC KIDNEY

An ectopic kidney is in an abnormal anatomical position. Figure 2a shows the right kidney in its normal anatomical position; the left kidney is visualised in the pelvic region in Figures 2b and c. A pelvic kidney is a normal kidney, which is located in the pelvis instead of its normal position in the abdomen (see Figure 1a (i)). This occurs when a kidney fails to ascend from the pelvis into the abdomen during normal foetal development.^[5] As a result a kidney in the pelvis has a congenitally short ureter. Its blood supply may arise from the aorta or iliac arteries. Its renal vein enters the IVC at a different level.^[5] A person with a pelvic kidney may not know they have an ectopic kidney.

Pelvic kidneys may be bilateral; a unilateral pelvic kidney is more common. The incidence of pelvic kidneys is approxi-

mately 1 in 2,500.^[6] A pelvic kidney may increase the risk of developing a urinary tract infection or renal calculi. Hydro-nephrosis may also develop. Diagnosis of an ectopic kidney is usually made during a gestational 18-20 week ultrasound scan. There is no specific treatment for a pelvic kidney. Most people with pelvic kidneys live normal healthy lives.

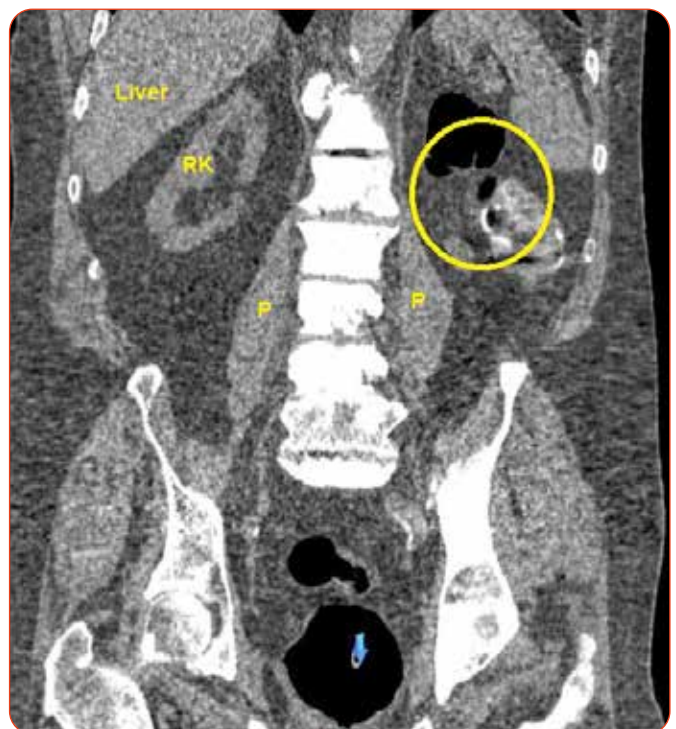


Figure 2a. Coronal view showing right kidney (RK) below the liver. Absent left kidney (yellow circle). Psoas muscle (P).



Figure 2b. Coronal view showing left kidney (LK) in the pelvis. Green arrow = left iliac bone. Caecum (C) and ascending colon (AC).

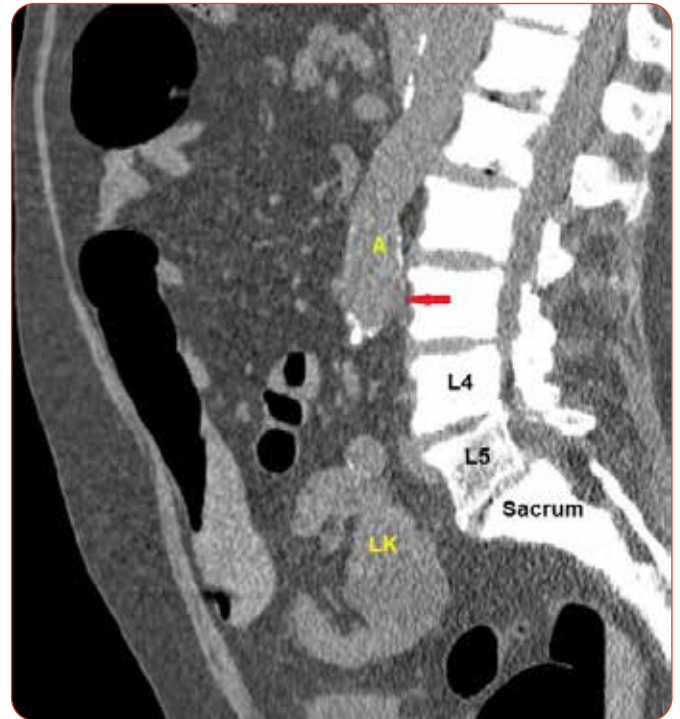


Figure 2c. Left sagittal view showing left kidney (LK) in the pelvis below lumbar vertebra 5 (L5). Aorta (A). Small aortic aneurysm (red arrow).

POLYCYSTIC KIDNEYS

Figure 3a to f shows polycystic kidneys. Polycystic kidney disease (PKD) is a rare disease which occurs in approximately 1:1000 cases. It is an inherited autosomal dominant disease which is known as autosomal dominant polycystic kidney disease (ADPKD).^[7] Multiple cysts are formed in the kidneys because a genetic factor interferes with their normal development.^[7-9] The vast majority of cases (75%) are caused by the defective gene PKD1; and 15% of the cases are caused by the PKD2 gene.^[7,8] There is no increased risk of renal cancer in patients with PKD.

In terms of clinical presentation the kidneys are normal at birth. Cysts begin to appear in the kidneys as time goes by.^[7] By the age of 30 years, almost 70% of patients will have multiple cysts visible on an ultrasound examination.^[7,8] With increasing age, there is an increase in the size and number of the cysts, as shown in Figures 3c and f. By the age of 60 years, almost 50% of patients will have developed end-stage renal disease^[7], requiring dialysis or renal transplant. It is important to carefully evaluate the cysts to assess thickness or calcification of the walls, suggesting previous haemorrhage. No normal pelvi-calyceal arrangement is seen. The kidneys are usually larger than normal and may show large differences in the renal size.^[8] In this case the right kidney measured 12 cm and the left one 22 cm. Multicystic renal disease may look similar to polycystic renal disease. Occasionally polycystic renal disease may have small cysts in the pancreas as well as the liver.

The following complications may occur as a result of PKD:

- 8% of patients may suffer from a berry aneurysm; this

may result in an intracranial bleed^[9]

- Hypertension may occur in a large proportion of patients^[7]
- Cysts may grow in size and may become inflamed^[7]
- Nephrolithiasis (renal calculi) may occur^[7]
- ADPKD may also be associated with cysts in the liver, pancreas or spleen.^[7,8] Cysts in these organs are not essential in the diagnosis of PKD.

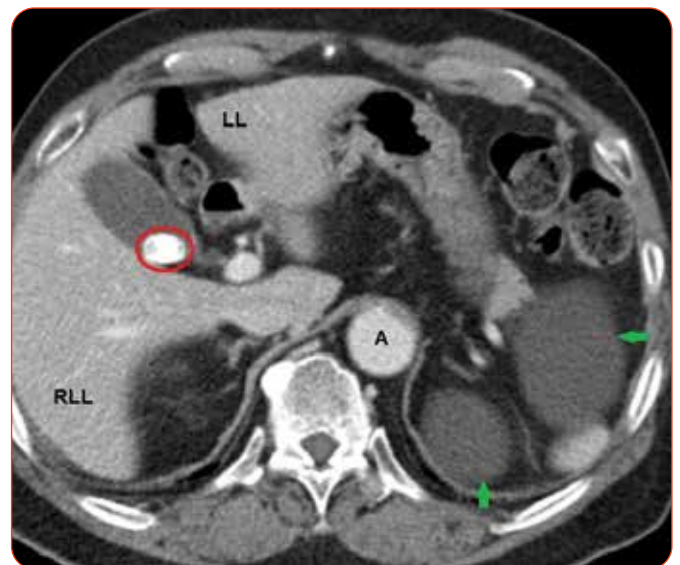


Figure 3a. Axial scan showing: calcified gallstone in a normal appearing gallbladder (red circle); cysts upper pole left kidney (green arrows). RLL = right lobe of liver. LL = left lobe of liver. Aorta (A).

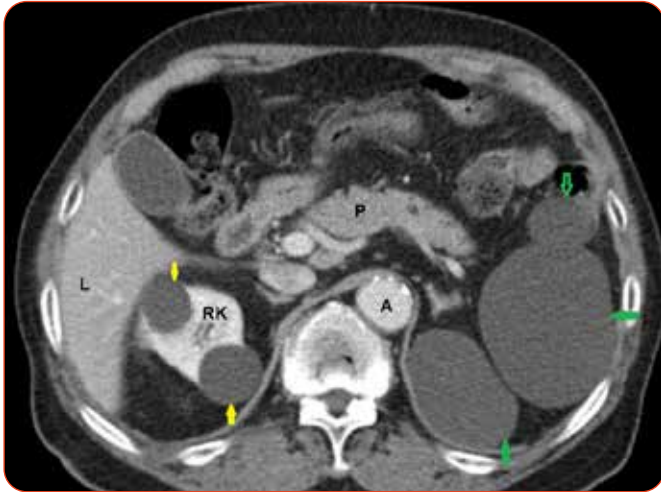


Figure 3b. Axial scan showing pancreas (P). Two small cysts appear in the right kidney (yellow arrows). The cysts in the left kidney appear larger (green arrows); a third cyst is visible on this scan (open green arrow). Aorta (A). Liver (L).



Figure 3c. Axial scan showing multiple cysts in the left kidney (green arrows). Note there is also a third cyst in this scan of the right kidney (yellow arrows), with IV contrast now visible. Aorta (A).



Figure 3d. Coronal view showing grossly enlarged left kidney measuring 22 cm (green arrows) with minimal contrast in the kidney. Right kidney with cysts (yellow arrows) showing right renal vein (blue arrow) joining the inferior vena cava (IVC).



Figure 3e. Right sagittal view showing the liver and the right kidney measuring 12 cm with multiple cysts and contrast (yellow arrows).

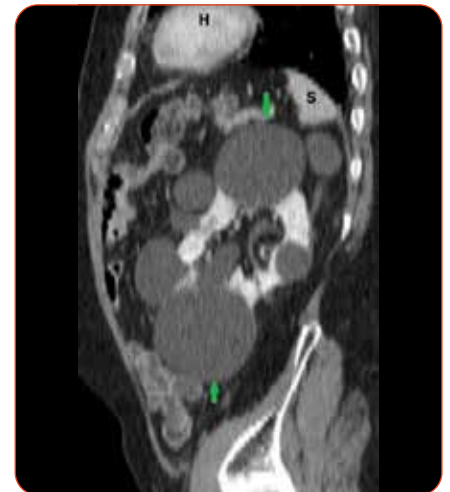


Figure 3f. Left sagittal view showing spleen (S) and a grossly enlarged left kidney measuring 22 cm with multiple cysts (green arrows). Heart (H).

HORSESHOE KIDNEY

Examples of horseshoe kidney (HSK) in three different patients are presented. Figure 4a (i) shows a HSK in a screening CT colonography patient, and Figure 4a (ii) is of another patient. Figures 4b (i) to (ix) are enhanced CT scans of a third patient.

Any interruption of the normal migration of kidneys during gestation will cause positional and fusion abnormalities.^[1] Fusion may be crossed fused ectopia (CFE) or HSK. The latter is the most frequent of fusion abnormalities. Figure 4c is a drawing of a HSK. Fusion of the kidneys across the midline results in a HSK, as shown in Figures 4a (ii) and 4b (ix); fusion occurs in the lower pole in >90% of cases.^[10] The

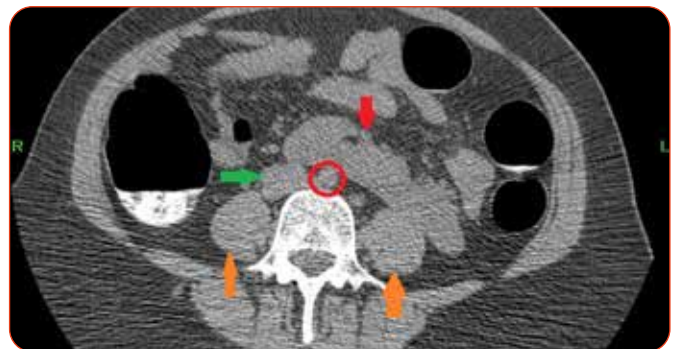


Figure 4a (i). Axial view shows a HSK as the kidneys are not seen in their respective renal fossa. The kidneys have fused anteriorly (red arrow). Green arrow = IVC. Red circle = aorta. Orange arrows = psoas muscle.



Figure 4a (ii). Axial view showing the fusion bar (isthmus), which joins each kidney (red circle) anterior to the aorta (green arrow). Yellow arrow = IVC. Red arrow = L kidney. Purple arrow = R kidney.

isthmus (see Figures 4a (ii), 4b (vi), 4b (viii)) may consist of fibrous or renal tissue. The fusion may be fibrous or consist of functioning renal tissue. Isotopic scan of the kidneys will show whether functioning renal tissue is present; there will be uptake of isotope in that region.^[11] HSK occurs in 0.25% of the population (approximately 1 in 450 persons).^[11] CFE incidence is much lower (1 in 2000 persons).^[10]

A HSK is prevented from reaching its normal position in the lumbar region by the inferior mesenteric artery (see Figure 4c) arising from the aorta. This artery blocks the upward movement of the kidneys during gestation due to the isthmus of the kidneys not being able to ascend any further. The HSK therefore usually lies between lumbar vertebrae 3 to 5. Renal rotation is also affected as shown in Figures 4b (i), (iv) and (vii).

As shown in Figure 1a (i) the upper poles of both kidneys are angulated towards the lumbar spine. The upper poles of both kidneys are normally angulated towards the lumbar spine and the lower poles laterally, as shown in Figure 1a (i). However, in HSK cases, the upper poles are displaced laterally and the lower ones medially and join each other via the isthmus.^[12] This rotational change causes the ureters to be anteriorly situated and situated in a higher position (see Figure 4c). This is thought to cause an increased incidence of urinary tract infection (UTI) and renal stone formation because the ureters have to cross anteriorly over the isthmus.

Glodney et al.^[11] found that 30% of their 209 cases had UTI or renal stones. Blood supply to the kidneys is usually by the right and left renal arteries. However, in HSK there is often a variable number of arteries arising from the aorta, iliac arteries, and inferior mesenteric arteries.



Figure 4b (i). Enhanced coronal view showing abnormal rotation of the right kidney (RK). It is horizontal, with the upper pole facing away from the lumbar spine. Contrast is seen in the hepatic veins (red arrow) draining into the right atrium. Bladder (B).



Figure 4b (ii). Compared to Figure 4b (i), this coronal view shows the isthmus (red arrow) of the kidney in the midline. It is the isthmus that is blocked by the inferior mesenteric artery in its upward ascent.

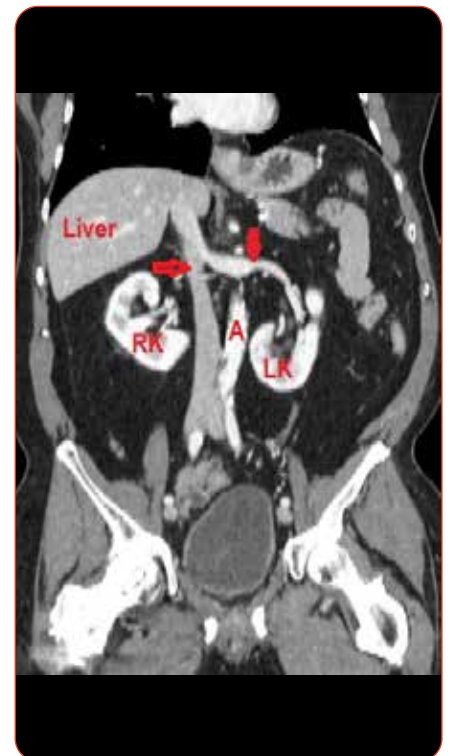


Figure 4b (iii). Enhanced coronal view showing the left renal vein (red arrow) entering the IVC (open red arrow). There is atherosclerotic change in the aorta (A). Right kidney (RK). Left kidney (LK).



Figure 4b (iv). Enhanced axial view showing rotational abnormality of the left kidney (LK). Right kidney (RK). Aorta (A).



Figure 4b (v). Enhanced axial view showing the isthmus (red arrow) of the kidney on the right (RK) advancing anterior to the aorta (A) to join with the inferior pole of the left kidney (LK), which is noted to be lying horizontally.

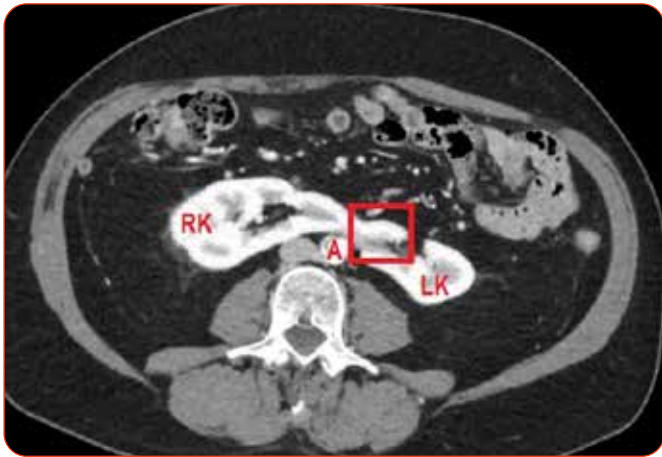


Figure 4b (vi). Enhanced axial view showing the joining of the RK to the LK to form a classical horseshoe kidney. Isthmus (red square) anterior to the aorta (A).



Figure 4b (vii). Enhanced sagittal view shows the right kidney's orientation (RK) beneath and posterior to the liver.



Figure 4b (viii). Enhanced sagittal view showing the isthmus (red rectangle) at L3 level immediately anterior to the aorta (red arrow).

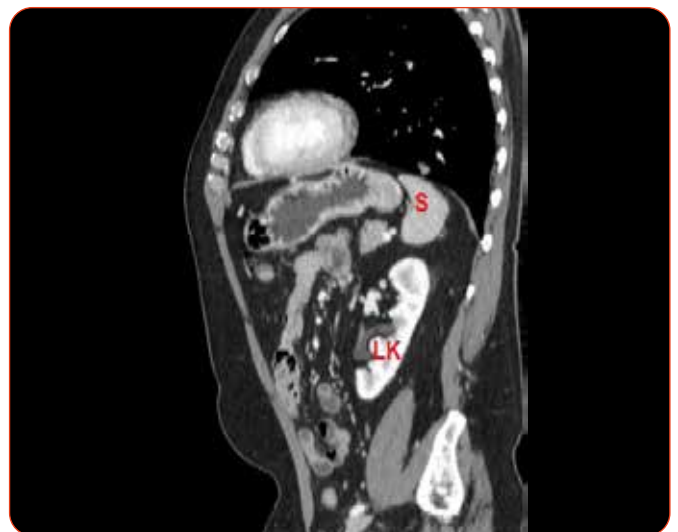


Figure 4b (ix). Enhanced sagittal view showing the left kidney (LK) below the spleen (S) in a posterior position.

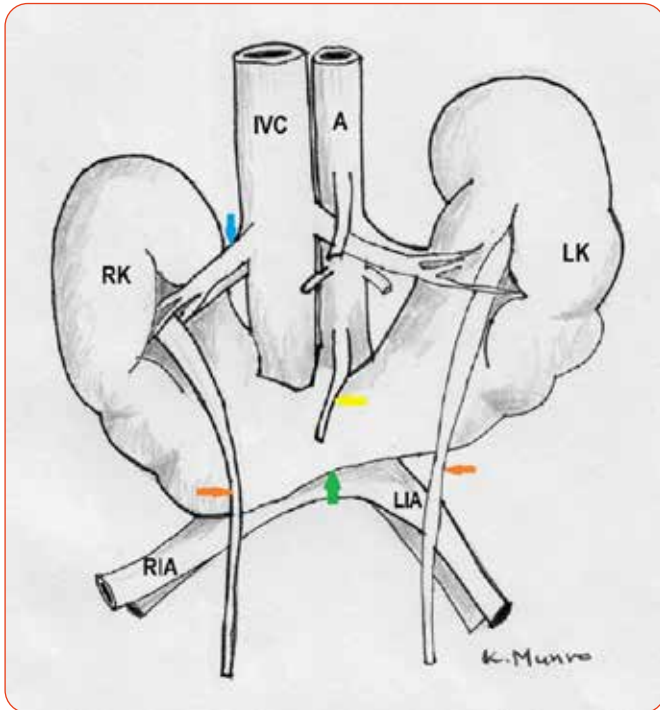


Figure 4c. Drawing of a HSK*. Orange arrows indicate the position of the ureters. Inferior mesenteric artery (yellow arrow). Isthmus (green arrow). Aorta (A). Inferior vena cava (IVC). Right renal vein (blue arrow). Right iliac artery (RIA). Left iliac artery (LIA). (*adapted from^[13]).

KEY MESSAGES

- Knowledge of the anatomy of the kidneys is important
- The upper pole of each kidney is deviated medially and the lower ones laterally
- Check that both kidneys are in their normal position. If not, then check for ectopic kidneys
- Check the outline of each kidney
- Check the orientation of the kidneys
- The right kidney is below the liver in the right sagittal views
- The left kidney is below the spleen in left sagittal views
- The inferior mesenteric artery may prevent the normal ascent of kidneys during gestation, resulting in fusion abnormality
- Ectopic kidneys and fused kidneys are usually incidental findings
- Complications of horseshoe kidney include an increased incidence of renal infections, calculi and hydronephrosis
- Congenital anomalies of the kidneys do not increase the risk of cancer

CONCLUSION

CT with intravenous contrast media is the preferred modality to show developmental abnormalities of horseshoe and pelvic kidneys. The life expectancy of people with these abnormalities is not affected. They may, however, present

with complications such as infection, calculi or hydronephrosis. Polycystic kidney disease (PKD) is an acquired disease, which tends to increase in severity as patients age. It may ultimately lead to dialysis, renal failure and finally kidney transplants. Ultrasound and CT imaging visualise PKD. Polycystic kidneys are usually in their normal position.

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