

Horseshoe Kidney

Manpreet K. Singh, BS; Richard B. Towbin, MD; Douglas C. Rivard, DO; Carrie M. Schaefer, MD; Alexander J. Towbin, MD

Abstract

Individuals with horseshoe kidneys can be asymptomatic or have associated complications from the anatomical anomaly. Various imaging modalities can diagnose this condition; however, it is up to the provider to order imaging studies based on what complication may be present, such as renal US or non-contrast enhanced CT for nephrolithiasis.

Keywords: renal, congenital anomaly

Case Summary

A term male infant was found to have a sacral dimple on newborn physical exam. A spine US was performed, which showed no spinal cord abnormality. Incidentally, midline fusion of the lower poles of the kidneys was identified by the sonographer.

Imaging Findings

Abdominal US, MRI, and nuclear imaging show the anomalous development of the kidneys (Figures 1–3). The normal renal axis of the kidneys is not present. Instead, the kidneys are low-lying and the renal axis is altered, with the lower poles more medially located than the upper poles. The kidneys are fused by a connecting isthmus at the lower renal poles.

Diagnosis

Horseshoe kidney.

Differential diagnoses include pelvic kidney, renal malrotation, and cross-fused renal ectopia.

Discussion

The horseshoe kidney is one of the most common renal congenital anomalies, with an incidence of about 1 in every 500 individuals.¹ Horseshoe kidneys can be fused at either pole, although fusion is most common at the lower poles of both kidneys. In the fetus, the kidneys originally develop from the ureteric buds that are responsible for the formation of the collecting system (collecting tubes, calyces, renal pelvis, ureter) while the metanephric blastema forms the kidney. During normal development, the collecting system and kidney join in the upper sacral region from reciprocal induction.² Renal fusion anomalies have been noted to occur between 4 and 6 weeks of development but can occur at 9 weeks in individuals who have a fibrous isthmus.²

Some theories regarding the causation of this anomaly are abnormal migration of nephrogenic cells across the primitive streak, teratogenic exposure during pregnancy such as alcohol and thalidomide, and the presence of lumbosacral

anomalies.^{3,4} Horseshoe kidneys have also been associated with chromosomal conditions such as trisomy 13, 18, 21, and Turner syndrome.²

In the fetus, kidneys develop in the lower abdomen, eventually ascending during week 7 of gestation and reaching the retroperitoneal space around week 8 or 9. With the fusion of the inferior poles, the kidneys remain in the pelvic area below the inferior mesenteric artery.² However, patients' kidneys with isthmic fusion were found to be directly inferior to the inferior mesenteric artery in only 40% of cases, while 20% of horseshoe kidneys remained in the pelvis.²

Variations in location, vascular supply, and degree of rotation exist among affected individuals. For example, a study showed that individuals with horseshoe kidneys are 10 times more likely to have variations in their inferior vena cava anatomy, thus affecting venous drainage.² Arterial supply also varies, with about 40% originating from the iliac artery, 3% from the median sacral artery, 3% from the lumbar artery, 2% from the internal iliac artery, and about 1% from the external iliac and phrenic artery.²

Although variations of horseshoe kidneys exist, patients can live normal and asymptomatic lives with this anomaly. About one-third of individuals are asymptomatic. In children, symptoms may arise due to a urinary tract infection or obstruction.⁵ Individuals with horseshoe kidneys are at increased risk

Affiliations: University of California, Riverside School of Medicine, Riverside, California (Singh); Department of Radiology, Phoenix Children's Hospital, Phoenix, Arizona (RB Towbin, Schaefer); Department of Radiology, Children's Mercy Hospital, Kansas City, Missouri (Rivard); Department of Radiology, Cincinnati Children's Hospital, University of Cincinnati College of Medicine, Cincinnati, Ohio (AJ Towbin).

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Figure 1. Transverse abdominal US image showing the isthmus of contiguous renal parenchyma between the lower poles of the right and left kidneys (arrow).

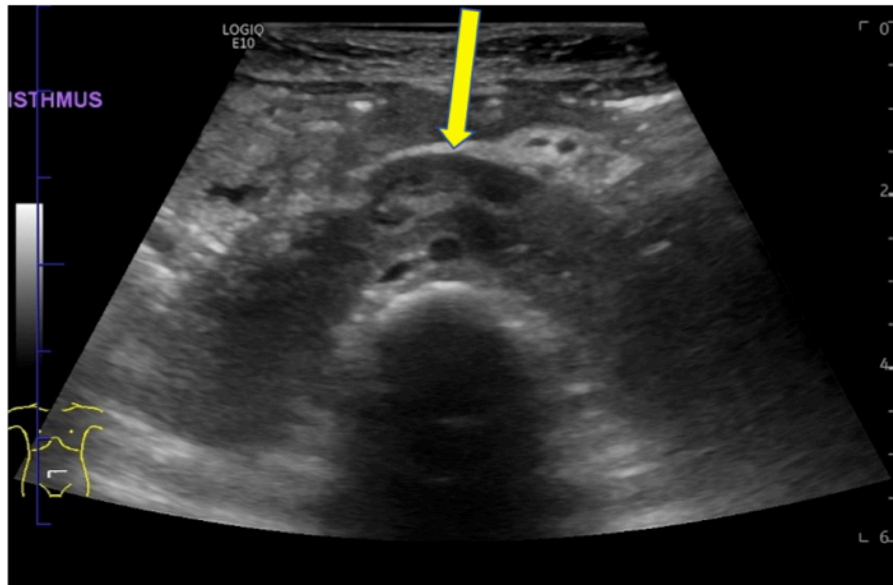
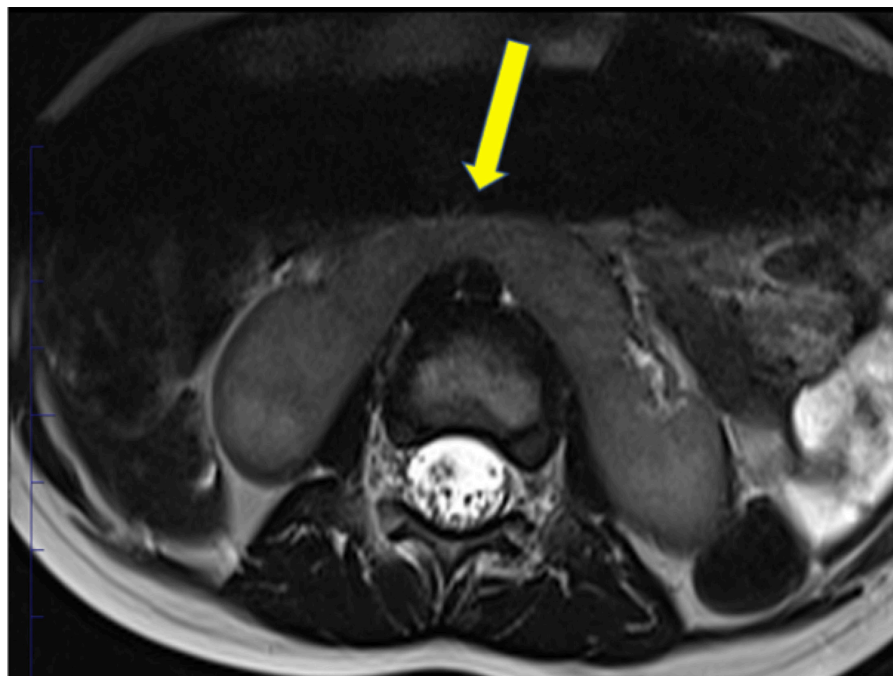


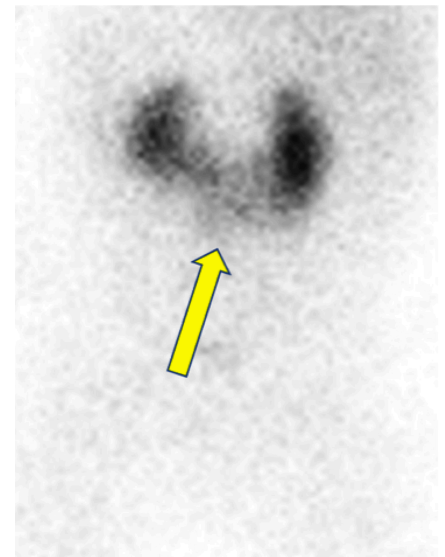
Figure 2. Axial T2-weighted image showing isthmus of renal parenchymal tissue connecting the lower poles of the right and left kidneys (arrow).



of renal trauma, vesicoureteral reflux, hydronephrosis, renovascular hypertension, ureteropelvic junction obstructions, and polycystic kidneys.⁵ About 35% of patients have pelviureteric obstruction,

and 20-60% of patients have obstructing stones.⁵ In addition, polycystic kidneys, although an independent congenital anomaly, can also have atresia of the ureteropelvic junction. If present with

Figure 3. Frontal planar image from mercaptoacetyltriglycine nuclear medicine scan demonstrating isotope uptake in both kidneys with uptake in the isthmus spanning the lower poles of each kidney (arrow).



a horseshoe kidney, treatments require knowledge of arterial supply to avoid damage. One case with concomitant horseshoe kidney and multilocular cystic dysplasia revealed the renal artery in the ventral direction, which is consistent with supplying the isthmus of a horseshoe kidney.⁶

Since many of the symptoms are nonspecific, imaging plays a key role in discovering the underlying etiology. Horseshoe kidneys can be an incidental imaging finding when patients are being worked up for other health concerns. However, if there is concern about abnormal renal function, horseshoe kidneys can be diagnosed via multiple imaging modalities such as intravenous pyelogram, US, CT, radionuclide scan, and MRI.¹ US is the preferred initial imaging modality recommended since it accurately diagnoses the condition at a lower cost without radiation exposure.

The imaging approach taken in children with horseshoe kidneys depends upon what complication is suspected. For example, if one is concerned

about nephrolithiasis, an abdominal radiograph and/or renal US is cost-effective and accurate.⁶ However, a non-contrast-enhanced CT of the abdomen and pelvis is more specific and sensitive and therefore used commonly in the emergency department.⁷ Voiding cystograms can be used to diagnose vesicoureteral reflux.¹ Nuclear medicine scans allow us to differentiate between functional and nonfunctional renal tissue as the isthmus.

Conclusion

Individuals with horseshoe kidneys can be asymptomatic or have associated

complications from the anatomical anomaly. Various imaging modalities can diagnose this condition; however, it is up to the provider to order imaging studies based on what complication may be present, such as renal US or noncontrast enhanced CT for nephrolithiasis.

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