

Renal Malformations: Horseshoe Kidney

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Abstract

Horseshoe kidney is one of the most common renal fusion anomalies, occurring in about .25% of the population. This condition exists when the lower poles of the kidney are fused together during development as a fetus. Although many times this malformation can be asymptomatic, there are various illnesses and problems that may accompany the horseshoe kidney. Medical Imaging of the condition is the first step to establish the severity of the problem and what may be done to remedy the symptoms.

Renal Malformations: Horseshoe Kidney

Renal disease and malformations affect the lives of millions of people every year. In some way or another, it will touch the lives of every person at some point in time. It has been estimated that 10% of the population has some sort of urinary tract abnormality (Bonsib, 2010). For hundreds of years doctors and scientists have been trying to discover why things like abnormalities and disease happen like they do in the body. It is now known that urinary tract abnormalities can occur in a number of different ways. Some are believed to be genetic such as ectopic kidney and horseshoe kidney. While others happen over time because of interactions happening in the body such as kidney stones or cancer. These abnormalities differ in severity, ranging from almost harmless, to fatal. Depending on the abnormality, some may live their entire lives and never know that they have had a kidney abnormality. In many cases when a

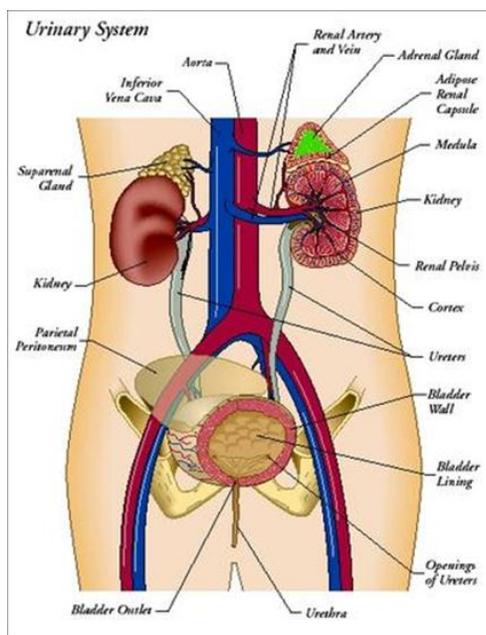


Fig. 1 Urinary system

Note. Kidney and Urinary Tract Stones? Web site. Retrieved November 12, 2011, from <http://healthbasictips.com/kidney-diseases/kidney-and-urinary-tract-stones/>

person is diagnosed with an abnormality, it is a secondary find in a study being performed for a different problem. One of the more common renal malformations is horseshoe kidney.

The Urinary System

In basic terms, the urinary system is comprised of two kidneys, two ureters, a bladder and a urethra. (See Fig. 1) The kidneys are full of little units called nephrons, which act as tiny filters. These nephrons filter the blood as it passes through the kidney. Imperfection, contaminants, and excess chemicals are pulled from the blood with these nephrons. The kidneys will filter

around 190 liters of water every day from the blood. Most of the water from the blood that is filtered is reabsorbed into the body. But a certain amount of the water is excreted as waste. This water travels down the ureters to the bladder. The bladder acts as a storage area for the urine. When the bladder reaches a certain volume, nerves in the walls of the bladder are stimulated letting the body know that it is time to void. Through a complex pattern of bladder contractions and relaxation of muscles and sphincters, the urine is expelled through the urethra where it leaves the body. The system should expel around two liters of urine a day. This system is of vital importance for cleaning of the body. Along with the need to filter the blood and remove imperfections from the body, the kidneys also play an important role in red blood cell production. Erythropoietin, a chemical that is produced in the kidney stimulates the production of the red blood cells. Therefore, severe anemia is often associated with renal failure because of the lack of chemicals produced (Eisenberg & Johnson, 2007).

Development

The development of the kidneys happens in three stages: pronephros, mesonephros and metanephros. The last of these three phases will occur around the fifth week of gestation (Ubetegoyena, Areses, & Arruebarrena, 2011). During this important stage of development, the formation of normal kidneys depends on the union of ureteric buds with the nephrogenic chords (O'Brien et al, 2008). The kidneys migrate from the pelvis where they are formed and ascend to the retroperitoneal space in the upper right and left quadrants. This ascension normally occurs in fourth to ninth week of gestation. (See Fig. 2) It is during this critical time in the early stages of formation and ascension that most of the malformations occur. These renal anomalies are a result of the interruption of the normal migration of the kidney. That is what is thought to happen with the horseshoe kidney. O'Brien et al (2008) states:

At this stage, the renal capsule has not matured and the kidneys still lie within the pelvis. It is suggested that abnormal flexion or growth of the developing spine and pelvic organs brings the immature kidneys together for a longer period than usual, leading to fusion of the two renal elements and hence forming the so-called horseshoe kidney. As this abnormal fusion occurs in the pelvis, the subsequent kidney cannot undergo normal migration and rotation. In the normal kidneys, the lower poles of the kidneys rotate laterally. However, with a horseshoe kidney, these poles remain medially positioned (p. 217).

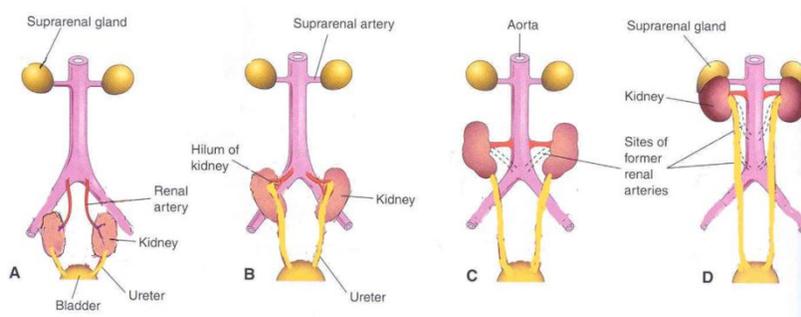


Fig. 2 A. Kidneys are in the first stages of development located in the pelvis. B. The kidneys have started their migration. C. Kidneys are half way. D. Kidneys have reached their spots just below the suprarenal glands.

Note: Retrieved November 12, 2011, from http://academic.amc.edu/martino/grossanatomy/site/Medical/CASES/R&R/pop%20ups/hydronephrosis%20anspop_up4.htm

The horseshoe kidney cannot migrate to the usual position because the fusion will not allow passage by the inferior mesenteric artery.

Anatomy of the Horseshoe Kidney

Horseshoed kidney is the most common type of fusion anomaly. For the most

part, the horseshoe kidney functions as a normal kidney. Many times, kidney malformations are accompanied by lower urinary tract anomalies as well. This is understandable because the kidney and the ureter arise from the same single embryonic structure (Adalat, Bockenbauer, Ledermann, Hennekam, & Woolf, 2010). With horseshoe kidney, the kidneys can be located anywhere along the normal embryologic ascent of the kidneys. Normally they are located lower

in the pelvic region of the body (De la Garza, Uresti, de la Vega, Elizondo-Omaña, & Guzmán-López, 2009).

In 90% of cases, the fusion of the kidneys occurs in the lower poles. (See Fig. 3) In this condition, both kidneys are malrotated and their lower poles are joined. With the fusion, there is an isthmus that crosses the midline of the body to connect the two kidneys. That is what gives this abnormality its name, with the two kidneys facing upwards and the isthmus joining them in the middle on the bottom they tend to appear as a horseshoe. This isthmus is composed of renal

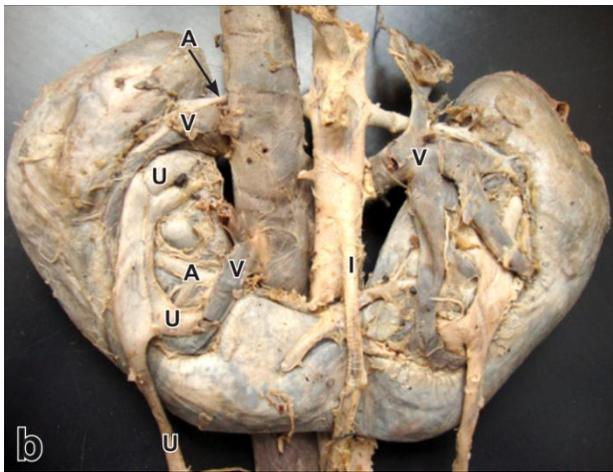


Fig. 3 Horseshoe kidney shown with descending aorta and inferior vena cava.

Note. From “A horseshoe kidney with partial duplex systems,” by K. Ongeti, J. Ongeng, and H. Saidi 2011, *International Journal of Anatomical Variations*, 4, p 56.

parenchyma or fibrous tissue. The ureters usually run anterior to the isthmus. If the fusion of the two kidneys occurs lateral to the midline, then one of the kidneys will be in a vertical position while the other will be mostly horizontal. The collection system of a horseshoe kidney is usually deviated inwards at the lower poles because of the fusion with the isthmus. The ureters arise from the kidneys anterior rather than medially. The

ureter also has a higher insertion point into the renal pelvis than that of a normal kidney (O’Brien et al, 2008).

The blood supply to the horseshoe kidney is also different than most kidneys. (See Fig. 4) There is actually several different ways that it can receive its supply. The blood supply could arise from the aorta, the iliac arteries and the inferior mesenteric artery. And it could occur as one of these, or a combination of all of them. Although in 65% of the cases, the isthmus is

supplied by single vessel from the aorta (O'Brien et al, 2008). As with the artery, the venous system can have its fair share of anomalies as well. It has been estimated that 22-24.8% of patients with horseshoe kidney also have a renal vein anomaly as well. The most common of these is multiple right renal veins (Ichikawa et al. 2011).

Imaging of Horseshoe Kidney

Most of the time, a horseshoe kidney is

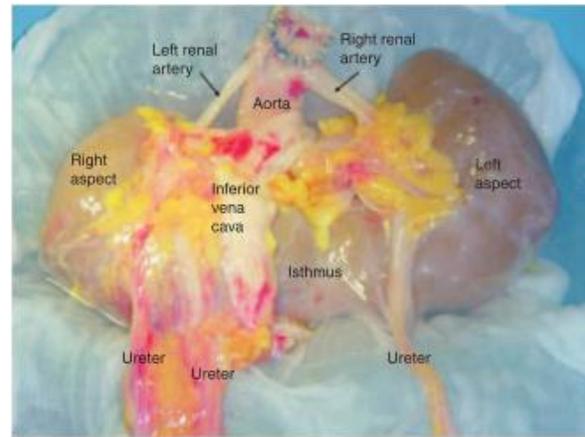


Fig. 4 Horseshoe kidney with labeled vasculature.

Note. From "Back-bench split of a deceased-donor horseshoe kidney for two transplant recipients," by J. Guarrera et al. 2009, *Kidney International*, 76(9), p 1012.



Fig. 5 Plain radiograph demonstrating a horseshoe kidney outlined with arrows.

Note. From "Imaging of horseshoe kidneys and their complications," by J. O'Brien et al. 2008, *Journal of Medical Imaging and Radiation Oncology*, 52(3), p 217.

an incidental find on an exam for some other condition that the patient is having. But once it is discovered, there are many options for imaging the anomaly. The kidneys can be seen on plain radiographs, but the definition is not as clear as in some of the other modalities. They will be discovered on plain radiographs because of their lower location and the location of the lower poles being more medially rotated than would be

expected. Under fluoroscopy or an Intravenous Urogram (IVU) the collecting system will be full of contrast. (See Fig. 6) Thus position and rotation will be more evident, as well as size. During this

exam, it will also be possible to study the ureters as well, to see if there are any abnormalities lower in the system.



Fig. 6 Laterally fused horseshoe kidney during IVU. Horizontally rotated right kidney is well visualized.

Note. From "Imaging of horseshoe kidneys and their complications," by J. O'Brien et al. 2008, *Journal of Medical Imaging and Radiation Oncology*, 52(3), p 218.

The modalities of choice for studying a horseshoe kidney are computed tomography (CT) or magnetic resonance imaging (MRI). CT is more commonly used to examine the condition because it allows precise observation of the anatomy as well as

evaluating possible complications. (See Fig.

7) The blood supply and venous returns are also able to



Fig. 7 CT of a 53 year old woman with horseshoe kidney.

be imaged with the help of contrast, which if the patient is going to surgery can be very valuable asset. MRI also provides a detailed study of the anatomy and possible complications. MRI also has a great benefit of no radiation dose to the patient. Additionally, with MRI angiography, vascular anatomy of the kidney will be well demonstrated (O'Brien et al. 2008).

Nuclear medicine and ultrasound can be utilized for finding and diagnosing horseshoe kidney, but they are not as commonly employed. (See Fig. 8) It can be a little more difficult to visualize at times with these two modalities.

Symptoms and Complications

Horseshoe kidney is the most common fusion anomaly in the kidney and occurs in about 1 in 400 people, or about .25%. It is also twice as likely to occur in males as in females (Ongeti, Ogeng, & Saidi, 2011). Although it is not highly common, it isn't uncommon either. Normally one-third of the patients that have horseshoe kidney are asymptomatic, and the condition is noticed incidentally on radiologic examination (Khan, Myatt, Palit, & Biyani, 2011). Although most patients are asymptomatic, there are certain conditions that go with horseshoe kidney quite frequently.

“When symptoms are present, they are usually because of obstruction, stones or infection with urinary tract infection being the most common presenting symptom in children” (O'Brien, 2008, p 219). The most common associated finding in horseshoe kidney is ureteropelvic junction obstruction, which occurs in up to 35% of cases. This obstruction more than likely is a result of the high insertion point of the ureters into the renal pelvis, causing delayed pelvic emptying. Many times this has to be surgically corrected. The next most common occurrence is presence of kidney stones. Kidney stones will develop in anywhere from 20%-60% of patients (Kahn et al. 2011). Kidney stones go hand-in-hand with obstructions; they have a tendency to cause one another. In the case of the horseshoe kidney, the

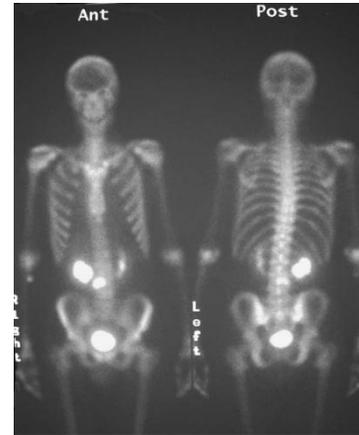


Fig. 8 Horseshoe kidney demonstrated in nuclear medicine.

Note. From “Imaging of horseshoe kidneys and their complications,” by J. O’Brien et al. 2008, *Journal of Medical Imaging and Radiation Oncology*, 52(3), p 219.

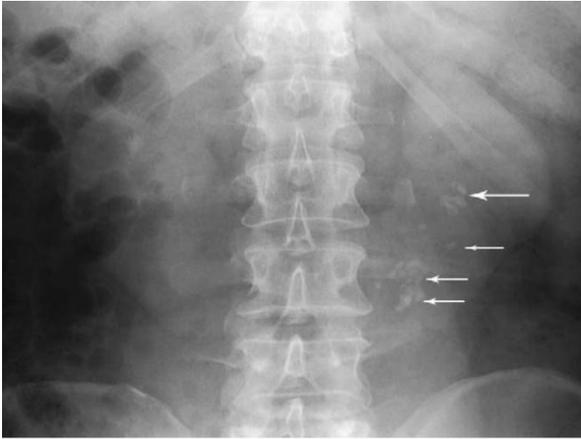


Fig. 9 Appearance of kidney stones in a horseshoe kidney.

Note. From “Imaging of horseshoe kidneys and their complications,” by J. O’Brien et al. 2008, *Journal of Medical Imaging and Radiation Oncology*, 52(3), p 221.

delayed draining of the renal pelvis may cause stones to form more readily. Stones may be a painful experience that will resolve, but it may also require surgical intervention for removal and stent placement. The horse shoe kidney is particularly vulnerable to infection. “This is because of a combination of reflux disease, stasis and stone formation. Infection occurs in up to one-third of patients. Infection is one of the important causes of death in patients with a

horseshoe kidney.” (O’Brien et al., 2008, p. 222). There are also a variety of tumors that have been associated with horseshoe kidney, the most common being renal cell carcinoma. Along with these illnesses and conditions, the horseshoe kidney is also more vulnerable to trauma. Because of where it is sitting low in the pelvis, it isn’t protected by the ribs like it would be in a normal situation. Therefore it is more open and easily injured.

Conclusion

Horseshoe kidney is a malformation that affects .25% of the population. Many of these people will never know they have it because one-third of those that have it are asymptomatic. For those that do know that they have the condition, they must worry about infections, stones, and obstructions. There isn’t any real cure for horseshoe kidney; the doctors will only treat the conditions that are presented by it. Regardless, there are many ways that the disorder might be imaged to help the doctor decide the best course of action to take for the well-being of the patient.

References

- Adalat, S., Bockenhauer, D., Ledermann, S., Hennekam, R., & Woolf, A. (2010). Renal malformations associated with mutations of developmental genes: messages from the clinic. *Pediatric Nephrology*, 25(11), 2247-2255. doi: 10.1007/s00467-010-1578-y
- Bonsib, S. M. (2010). The classification of renal cystic diseases and other congenital malformations of the kidney and urinary tract. *Archives Of Pathology & Laboratory Medicine*, 134(4), 554-568
- De la Garza, O., Uresti, J., de la Vega, E., Elizondo-Omaña, R., & Guzmán-López, S. (2009). Anatomical study of the horseshoe kidney. *International Journal of Morphology*, 27(2), 491-949.
- Eisenberg, R., & Johnson, N. (2007). *Comprehensive Radiographic Pathology*. St. Louis, Mo: Elsevier Mosby.
- Ichikawa, T., Kawada, S., Koizumi, J., Endo, J., Lino, M., ... Imai, Y. (2011). Major venous anomalies are frequently associated with horseshoe kidneys. *Circulation Journal*. doi: 10.1256/circj.CJ-11-0613
- Khan, A., Myatt, A., Palit, V., & Biyani, C. (2011). Laparoscopic heminephrectomy of a horseshoe kidney. *Journal of the Society of Laparoendoscopic Surgeons*, 15(3), 415-420. doi: 10.4293/108680811X13125733356512
- O'Brien, J., Buckley, O., Doody, O., Ward, E., Persaud, T., & Torreggiani, W. (2008). Imaging of horseshoe kidney and their complications. *Journal Of Medical Imaging And Radiation Oncology*, 52(3), 216-226.
- Ongeti, K. W., Ogeng'o, J. & Saidi, H. (2011). A horseshoe kidney with partial duplex systems. *International Journal Of Anatomical Variations*, 4, 55-56.

Ubetegoyena, A. M., Areses, T. R., & Arruebarrena, L. D. (2011). Anomalías renales de posición y de fusion. *Anales de Pediatría*, 75(5), 329-333. doi: 10.1016/j.anpedi.2011.05.011