

# Horseshoe kidney: is there still a debate?

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## SUMMARY

Horseshoe kidneys (HSK) represent an interesting surprise during anatomical dissections directed towards teaching of the urinary system. Clinically, the HSK limits access into the retroperitoneal space due to its location, orientation, and positioning of the ureters. In addition, its highly variable arterial and venous patterns provide great difficulties for surgeons during aortic aneurysm correction, and more recently, HSK transplantation.

This case is a morphological study of a noticeably different HSK from the perspective of location, arterial blood supply, and venous drainage, which is further solidified by an embryological review. The debate is opened for further exploration into the theories associated with HSK ascent, its vasculature patterns, and the need for precise diagnostic imaging to serve preoperative planning.

**Key words:** Human gross anatomy – Renal embryology – Horseshoe kidney – Arterial supply – Venous drainage

## INTRODUCTION

A horseshoe kidney (HSK) consists of two well defined kidney masses laying with their longitudinal axes adjacent to the lumbar spine, that are interconnected at their respective poles by a parenchymatous or fibrous tissue isthmus. It is the most commonly seen fusion defect and urogenital deformity observed in the kidneys (Taghavi et al., 2016; Nikumbh et al., 2014). Disregarding its presumably rare occurrence, the embryologic defect of HSK has generated a notable amount of literature. This comprises anatomical dissections and

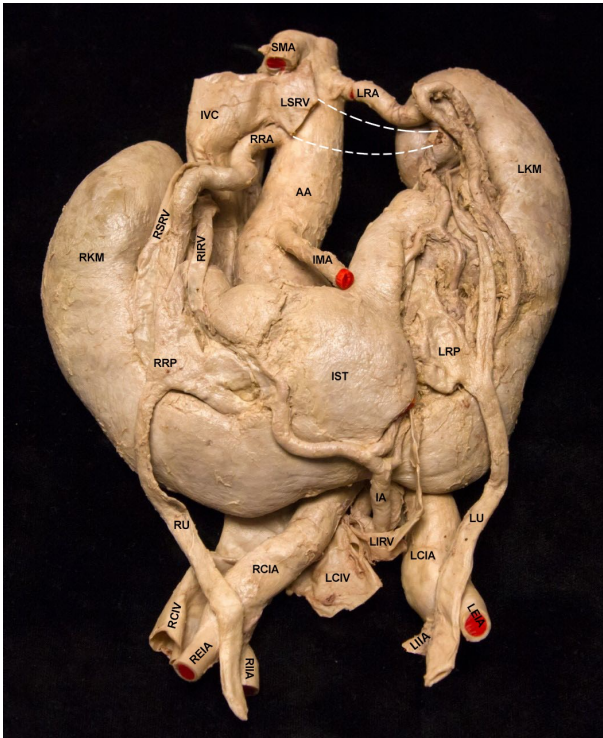
autopsy findings, x-ray and sonography discoveries, surgical corrections directed to the HSK or associated with abdominal aortic aneurysms, which more recently includes transplantation of the HSK. The reported incidence of HSKs vary largely depending on the method of study, with 1 in 400 to 1 in 1600 for autopsy findings (Bergman et al., 2016), 1 in 352 for urography examinations (Dees, 1941), and 1 in 256 for surgical reports (Lowsley, 1952). In addition, group age difference varies between 1 in 600 for new-borns (Sadler, 2005), 1 in 270 for children (Lowsley, 1952), and 1 in 542 for adults (Lowsley, 1952). Lastly, the ratio between males to females is 2.5:1 (Lowsley, 1952; Natsis et al., 2014).

Horseshoe kidneys are usually asymptomatic, therefore their discovery is typically incidental during urological examinations (Natsis et al., 2014; Boatman et al., 1972). However, due to the rise in ultrasonographical and radiographical examinations, such as retrograde pyelography and intravenous urography, HSKs are becoming more detectable (Bergman et al., 2016). There are no known racial or genetic associations. Instead, it has been reported that there are links between HSKs and congenital anomalies such as supernumerary kidneys, Patau syndrome, Edwards syndrome, Turner syndrome, and spina bifida (Natsis et al., 2014). Carlson et al. (2014) further explains that HSKs are more susceptible to infections, ureteric obstructions, hydronephrosis, and it may be associated with anomalies of other internal organs.

Over the years, the classical mechanical fusion theory has generated the highest support, as a result of space restrictions within the pelvic cavity during the fourth week of development when the metanephric blastemas firstly appeared. However, a much more recent teratogenic theory points to the atypical migration of the most dorsally placed nephrogenic cells to form a parenchymatous, non-fibrotic, isthmus. The exact fusion mechanism un-

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**Fig 1.** Horseshoe kidney specimen – anterior view. AA – abdominal aorta; IA – isthmic artery; IMA – inferior mesenteric artery; IST – isthmus of HSK; IVC – inferior vena cava; LCIA – left common iliac artery; LCIV – left common iliac vein; LEIA – left external iliac artery; LIIA – left internal iliac artery; LIRV – left inferior renal vein; LKM – left kidney mass; LRA – left renal artery; LRP – left renal pelvis; LSRV – Left superior renal vein; LU – left ureter; RCIA – right common iliac artery; RCIV – right common iliac vein; REIA – right external iliac artery; RIIA – right internal iliac artery; RIRV – right inferior renal vein; RKM – right kidney mass; RRA – right renal artery; RRP – right renal pelvis; RSRV – right superior renal vein; RU – right ureter; SMA – superior mesenteric artery.

derlying the HSK formation still remains unanswered (Bergman et al., 2016; Tijerina et al., 2014; Yoshinaga et al., 2002; Fitzgerald, 1978).

The present study reports on a HSK case with a unique presentation in terms of location and vasculature. Through an embryological review explaining the anomalous positioning and vascular presentation, this case opens the discussion for a new hypothesis linked to an incomplete ascent of the HSK.

## CASE REPORT

In a routine cadaveric dissection of a 73-year-old male in the Department of Anatomy, a retroperitoneal mass was uncovered. After evisceration and removing the posterior parietal peritoneum, a HSK was discovered. Following dissection, the specimen was photographed (Fig. 1) and measured, and the measurements recorded (Table 1).

### General features of the HSK

The specimen displayed a U-shaped orientation,

with a downward facing convexity, located at a relatively lower position than expected. The initial assessment of the HSK showed the isthmic portion laying over the vertebral body L4, partially obscuring the origin of the common iliac arteries, while at the same time its superior border was located 2 cm below the origin of the inferior mesenteric artery.

The ventral surface of the specimen was convex, while the dorsal surface had a flattened appearance at the level of the kidney masses; however, a deep groove was noticed on the dorsal surface of the isthmus that accommodated the abdominal aorta and inferior vena cava.

It appeared that axial rotation of each kidney mass was deficient, therefore the hilum of each mass was positioned anteriorly rather than medially. At the same time, this mal-rotation placed the major calyces and renal pelvis on either side in an extra renal position, and it also altered the relationship between the vascular and urinary components at the level of the renal hilum.

The examination of the ventral surfaces of the right and left kidney masses showed renal hila that were occupied by vascular and urinary structures, both of which completely lacked a renal sinus. The calyceal systems were relatively identical on both sides with the only exception that the left superior major calyx was extremely long and drained directly into the left ureteropelvic junction; therefore, the left renal pelvis looked considerably smaller in size when compared to the right one. The ureteropelvic junction on both sides was related to the anterior border of the renal hilum. The proximal portion of both ureters descended over the anterior surface of their respective kidney mass along the lower portion of the specimen.

### Vasculature of the HSK

The HSK was supplied by three arteries, one for each kidney mass resembling the appearance of a typical renal artery, and an isthmic artery.





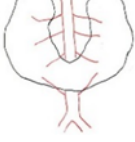
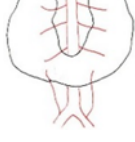
The right kidney mass was supplied by a renal artery which emerged from the abdominal aorta 2.5 cm below the origin of the superior mesenteric artery, in close proximity to the right gonadal artery. The left kidney mass was supplied by one renal artery, which also emerged from the abdominal aorta but 0.5 cm inferior to the origin of the superior mesenteric artery. Both arteries had a diameter of 5 mm and branched into two segmental arteries, distributing to the upper and middle portions of each kidney mass.

The isthmus of the HSK and the lower portions of each kidney mass was supplied by an isthmic artery that measured 5 mm in diameter at the origin. It emerged 1 cm below the bifurcation of the abdominal aorta from the anterior surface of an unusually thick common median sacral arterial trunk. This terminology was used due to the arterial trunk presenting with a diameter of 5 mm that appeared to be continuous with the isthmic artery, while from its posterior surface emerged a median sacral artery proper with a diameter of 0.5 mm that followed

**Table 1.** Measurements of the horseshoe kidney

	Right Kidney Mass	Left Kidney Mass	Isthmus
Length	13 cm	14 cm	5.5 cm
Width	6 cm	6 cm	1 cm
Depth	Superior pole: 5.5 cm Inferior pole: 2 cm	Superior pole: 4.5 cm Inferior pole: 1.5 cm	1 cm

**Table 2.** Adaptation of Boatman's six basic arterial patterns supplying the HSK

Type	Drawing	Description
Type 1a		Arterial pattern similar to that of normal kidneys; each kidney mass is supplied by a single renal artery that distributes to the upper, middle and lower segments, including the isthmus
Type 1b		The upper and middle segments of each kidney mass are supplied by a single renal artery, while the lower segments and isthmus are each supplied by a branch from the abdominal aorta
Type 1c		Similar to type 1b, however, the lower segments and isthmus are each supplied by a common trunk arising from the abdominal aorta
Type 1d		There are multiple renal arteries on each side that distribute to the upper, middle and lower segments, including the isthmus
Type 1e		Similar as type 1d, however, the lower segments and isthmus are also supplied by arteries that arise from the abdominal aorta below the isthmus, independently or by a common trunk
Type 1f		Similar as type 1e, however, the lower segments and isthmus are also supplied by arteries that arise from the common iliac arteries, or rarely, from the internal iliac arteries, or middle sacral artery

an unremarkable course. The isthmic artery looped around the inferior border of the isthmus, then ascended on its anterior surface prior to branching into the feeding arteries to the isthmus and lower portions of each kidney mass. Although there is apparent symmetrical arterial blood distribution to the HSK through three arteries, their peculiar origins provided an interesting case from an arterial pattern point of view.

The venous blood of the right kidney mass drained through two renal veins, separated 1 cm apart as they opened directly into the inferior vena cava. The venous blood of the left kidney mass

drained through two renal veins. The superior renal vein was a tributary to the inferior vena cava, while the inferior renal vein descended next to the isthmic artery and drained into the left common iliac vein, making it a unique presentation from a venous drainage point of view.

## DISCUSSION

The formation of HSKs must be understood in the context of the development of normal kidneys. Development of the kidneys require a series of time sensitive interactions between: (a) degenerat-

ing mesonephric system and emerging metanephric system; (b) relative ascent of the kidneys, in relation to the embryo; and (c) vascular growth and resorption between aorta and the developing kidney.

Alteration of these processes leads to kidney anomalies, including but not limited to renal fusions. In the event of a HSK formation, the presence of an isthmus impedes the complete ascension of the organ that is blocked by the inferior mesenteric artery, and therefore migration of the organ is arrested in a relatively lower position within the abdominal cavity (Fitzgerald, 1978). Finally, the dynamic of the vascular rearrangements explains the formation of accessory renal arteries that may originate from any of the following sources: common iliac artery (39.8%), median sacral artery (2.9%), internal iliac artery (1.94%), external iliac artery (0.97%), or phrenic artery (0.97%) (Glodny et al., 2009).

While the embryological review explains the formation of accessory renal arteries, in this case an isthmus originating from a median sacral arterial trunk, it does not explain the abnormally inferior positioning of the specimen that: (1) through its isthmus related to the topographic level L4; (2) the inferior border obscured the origin of the common iliac arteries; and (3) the superior border located 2 cm below the origin of the inferior mesenteric artery. The lack of resorption of the isthmus during development appeared to have hindered the HSK from further ascent. A side contribution to this process in the present case seems to come from the lack of resorption of the left inferior renal vein, a tributary to the left common iliac vein.

Another unique attribute identified was the arterial pattern supplying the HSK. Literature searches were unsuccessful in finding another specimen with the same vasculature patterns to the HSK. The six basic pattern arterial system that has initially been proposed by Graves, using resin casts and later solidified by Boatman's angiographic study, is presented in Table 2 (Graves, 1969; Boatman et al., 1971). In general terms, the arterial blood supply of the presented case appeared to be a hybrid between type 1a and type 1f. The type 1a designation is supported by a pair of renal arteries supplying the upper and middle portions of the right and left kidney masses, whereas type 1f by an isthmus artery to the lower portion of the kidney masses and isthmus originating from the middle sacral artery.

On the other hand, the venous drainage of the HSK has been often overlooked, most likely due to the limitations associated to the methods of study. In line with the well-known heterogeneity of venous patterns draining the kidneys, the presented HSK case displayed two right renal veins draining the right kidney mass into the IVC and two left renal veins draining the left kidney mass into the IVC and the left common iliac vein. Therefore, just the venous anomalies alone as they associate to the HSKs may represent a "minefield" in open vascular

surgeries (Sato, 2011).

The coexistence of HSKs with unpredictable arterial and venous vascular patterns stresses the increased need for careful preoperative imaging evaluations in order to minimize the risks associated with surgery. In light of the challenges faced with the HSK surgical correction, transplantation, as well as abdominal aortic aneurysm surgical repair in the presence of a HSK, understanding the vasculature becomes extremely important.

In conclusion, the topic of HSK is not as clear-cut as once understood: the embryological development, vascular patterns, as well as its positioning should perhaps be viewed on an individual detailed anatomical basis in order to provide the most accurate and up to date clinical information.

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