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# Abnormal vascular supply of the horseshoe kidney: case report and review of the literature

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## Abstract

The horseshoe kidney is a congenital fusion anomaly of the kidneys with an incidence of 0.25% in the general population. We present an anatomical and histological study of a horseshoe kidney with abnormal vascular supply found during a routine dissection at a formalin-fixed male human body at the School of Medicine of the University of Caxias do Sul. This horseshoe kidney was joined at the lower poles by an isthmus of parenchymatous tissue. The vascular and excretory system is also discussed. We also present an embryologic review and the clinical and surgical importance of this anomaly.

**Key words:** anatomy; horseshoe kidney; kidney anomalies; surgical anatomy

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## Introduction

The horseshoe kidney (HSK) is a fusion anomaly of the kidneys with an incidence of 0.25% in general population, being more prevalent in men than women in a 2:1 relation.<sup>[1]</sup> This abnormality has been discovered clinically in all age groups ranging from fetal life to 80 years, but in autopsy series it is more prevalent in children.<sup>[2]</sup> This early age prevalence has been explained by high incidence of multiple congenital anomalies associated with the HSK, some of which are incompatible with long-term survival.<sup>[3]</sup>

There is a wide variation in HSK vascular supply and excretory system. Renal arteries can originate from the abdominal aorta (Ao), the iliac arteries, and the inferior mesenteric arteries. Therefore it is important to know the clinical and surgical anatomy of the HSK. Renal transplants with HSK have been performed and for the proper management of this clinical condition it is necessary to understand the surgical difficulties and the most frequent clinical complications such as urolithiasis.

In view of this important kidney anomaly, we report a case with a unusual arterial supply.

## Case Report

During a routine dissection of the retroperitoneal cavity in a 63-year-old formalin-fixed male cadaver of the Department of Human Anatomy of the School of Medicine of University of Caxias do Sul, was found a HSK joined by an isthmus in the lower poles.

## Size and position

The isthmus was located anteriorly of the Ao and the inferior vena cava (IVC) at the level of the fourth lumbar vertebra. The longitudinal axis measured 10 cm in the right kidney, 9.75 cm in the left kidney and 5cm in the isthmus. In thickness the isthmus measured 2.8 cm, the left kidney 3.55 cm and the right kidney 3.6 cm. The distance from the isthmus to the Ao bifurcation on common iliac arteries was 2.35 cm and to the aortic diaphragmatic hiatus was 13.7 cm.

### Vascular system

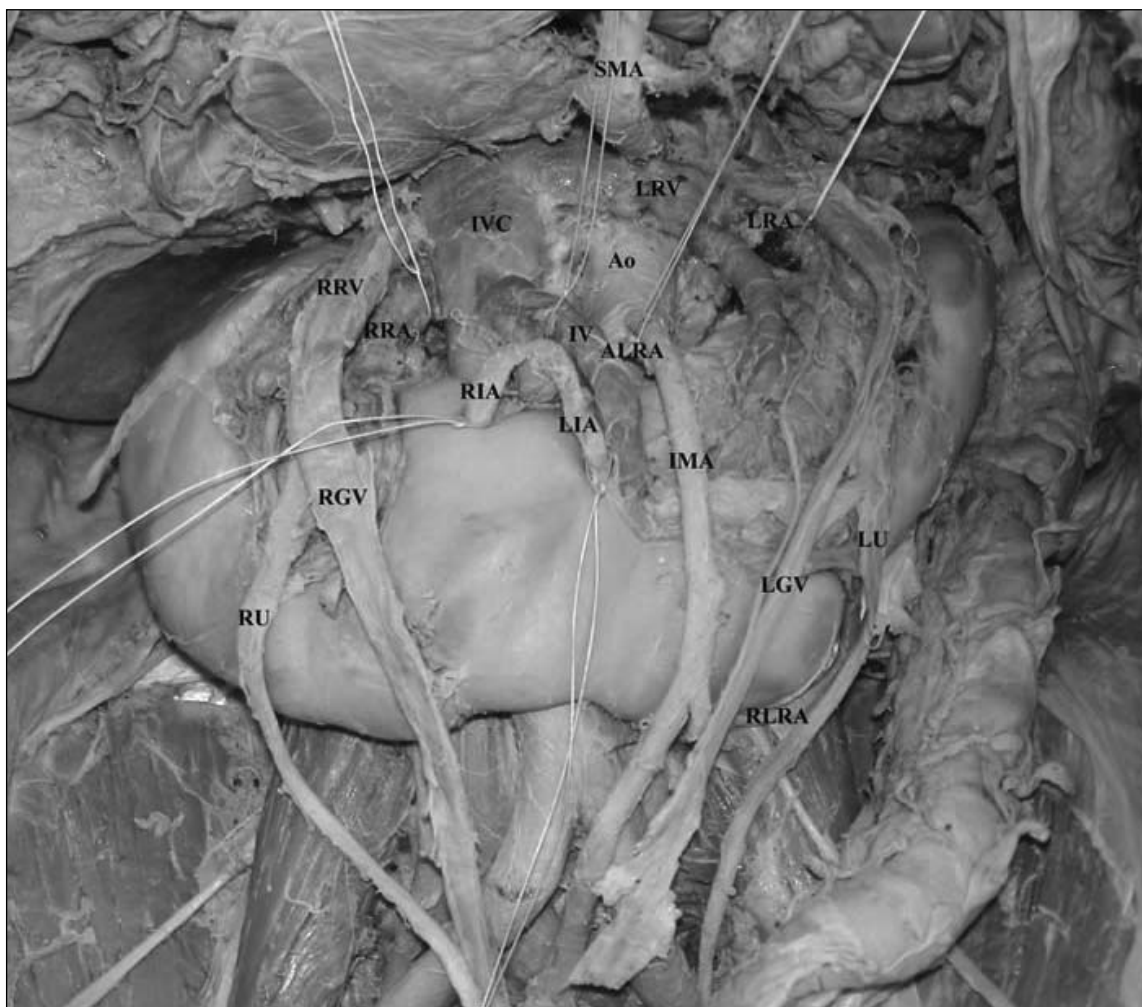
The arterial supply (**Figure 1**) of the right kidney was provided of the right renal artery that was originated in lateral surface of the Ao. The left kidney was supplied by the left renal artery, provided of the lateral surface of the Ao, and by the left accessory renal artery originated from the isthmus arterial trunk. The left kidney even received a recurrent artery, provided from the left surface of the Ao inferiorly of the isthmus, that passed posteriorly of the inferior mesenteric artery. The isthmus was supplied by an arterial trunk, provided from the anterior surface of Ao, that had three branches: right and left isthmus arteries and accessory left renal artery.

Venous drainage system (**Figure 1**) was consisted of three veins that empty into the IVC. Two tributary

veins, superior and inferior veins, arose from the right hilum to form the right renal vein that empty into to the lateral surface of the IVC. The left renal vein, formed by two tributary veins that arose from the left hilum, passed posteriorly of the superior mesenteric artery and emptied into the lateral surface of the IVC. The right and left gonadal veins was tributaries of the respective side renal veins. The isthmus was drained by a single vein that was a tributary of the anterior surface of the IVC. The isthmus vein passed anteriorly of the accessory left renal artery.

### Pelvis and ureter

The right ureter drained exclusively the right kidney. Left ureter drained the left kidney pelvis and the isthmus



**Figure 1.** Horseshoe kidney in the retroperitoneal cavity. Ao: abdominal aorta; RRA: right renal artery; LRA: left renal artery; RIA: right isthmus artery; LIA: left isthmus artery; ALRA: accessory left renal artery; IVC: inferior vena cava; RRV: right renal vein; LRV: left renal vein; IV: isthmus vein; RGV: right gonadal vein; LGV: left gonadal vein; RU: right ureter; LU: left ureter; RLRA: recurrent left renal artery; IMA: inferior mesenteric artery; SMA: superior mesenteric artery.

pelvis. Both ureters crossed anteriorly of the psoas major muscles and the common iliac arteries and inserted independently into the posterolateral angles of the urinary bladder.

### Histology of the isthmus

The isthmus was biopsied for histological analysis in order to observe the tissue of the isthmus. That isthmus specimen was paraffin-embedded prepared and colored with hematoxylen and eosin (HE). We conclude that the isthmus consisted of parenchymatous tissue with many renal corpuscles and renal tubules (**Figure 2**).

### Discussion

The anatomical knowledge of the HSK is increasingly important, in view of more than one hundred HSK was transplanted during the last three decades.<sup>[4]</sup>

### Embryology review

This congenital anomaly results from an abnormal medial fusion of the metanephric blastema originally destined to result in anatomically normal right and left kidneys. This abnormal fusion causes failure of ascent and rotation. Ascent is arrested by the inferior mesenteric artery, which arises anteriorly from the Ao just before its bifurcation. The fused kidney therefore lies more inferiorly in position compared with normal kidneys and, because of incomplete rotation, the renal pelvis is anterior to all of the calyces.<sup>[5]</sup>

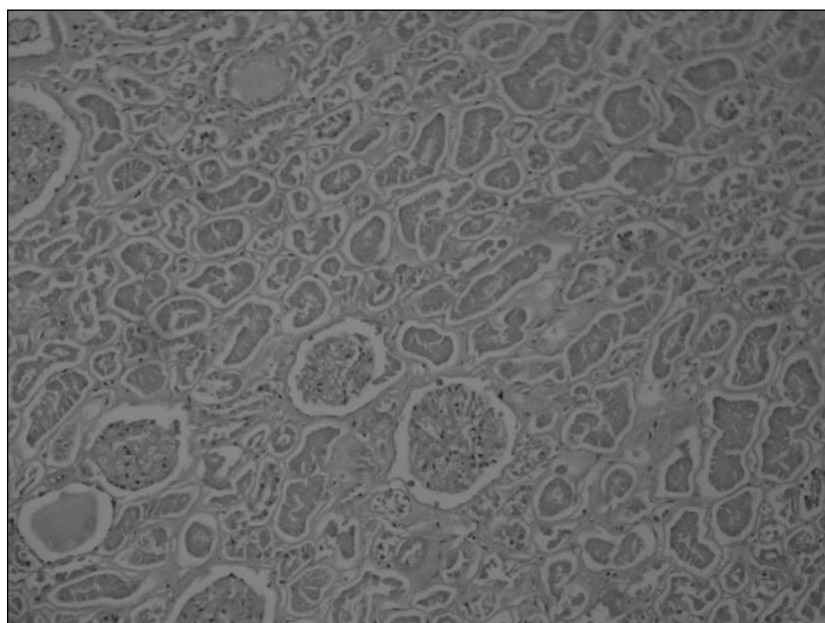
### Relations with chromosomal anomalies

Kidney malformations associated with chromosomal anomalies incidence is present. Thirty-three per cent of patients with Turner syndrome presents some renal malformation and 7.1% of that patients present HSK.<sup>[6]</sup> Despite of most structural malformations do not cause dysfunction, silent hydronephrosis can result from an obstruction of a duplicated collecting system, that is presented in 10% of Turner syndrome patients.<sup>[7]</sup>

### Clinical and surgical significance

Urolithiasis is the most common complication of HSK, with an incidence higher than 20%. The tortuous tract of the ureter, especially as it crosses the isthmus, makes semirigid ureteroscopy for proximal ureteral or renal calculi exceedingly difficult and dangerous.<sup>[8]</sup> Extracorporeal shock wave lithotripsy may be a reasonable choice for patients with a small stone burden in HSK. However, for patients with large stones, percutaneous nephrolithotomy is an effective procedure for the treatment with similar success and complications rates of normal kidneys.<sup>[9]</sup>

The vascular supply of HSK can be quite variable, in 30% of cases, it consists of one renal artery to each kidney.<sup>[10]</sup> However, it may be atypical with duplicate or even triplicate renal arteries supplying one or both kidneys. The isthmus arterial supply and lower poles is also variable and may receive a branch from each main renal artery or have their own arterial supply from the Ao originating either above or below the level of the isthmus. Less frequently this area is supplied by branches



**Figure 2.** Light microscopic photograph of the isthmus, colored with HE, evidencing the parenchymatous tissue with many renal corpuscles and renal tubules (x100).

from the inferior mesenteric artery and common or external iliac arteries.<sup>[11]</sup>

Vaniya<sup>[12]</sup> reported a case of multiple renal arteries that originated from different vertebral levels of the Ao. The right kidney irrigation provided from four arteries originated from the right surface of Ao at the level of L1, L2 and L3. The left kidney irrigation provided from two arteries originated from left surface of Ao at the level of L1 and L3. The artery originated from left L1 level divided in three terminal branches before it entered into the upper part of left hilum. The isthmus was irrigated by two arteries that originated from the front of the bifurcation of the Ao at L5 level.

Yoshinaga et al.<sup>[13]</sup> reported a case that the kidney was supplied by four arteries arising from the Ao and the isthmus had its own blood supply from two renal arteries arising below the inferior mesenteric artery. In the case reported by Tijerina et al.,<sup>[14]</sup> a renal artery for each kidney that originated from the lateral surfaces of the Ao just below the superior mesenteric artery was found. The isthmus was supplied by two short renal arteries that originated directly from the Ao, below the inferior mesenteric artery. In view of the intense variable arterial supply and other variations of the HSK, the knowledge of the anatomy of HSK is very important for planning surgical or clinical treatment.

The abdominal aortic aneurysm surgery associated with HSK gives rise to technical difficulties on account of the location of the isthmus, anteriorly of the aneurysm, that frequently needs to be divided to expose the Ao.<sup>[15]</sup> The technical difficulties are related to preserve the kidney vascular system. or aorto-iliac anastomosis with preservation of the irrigation of the left kidney would be a challenge, in view of the triple irrigation with different, but closely origin. Therefore, considering this variable arterial anatomy, angiography is very helpful when planning renal surgery on HSK. However, in trauma patients, principally the hemodynamically unstable patients, there is no time for planning. Besides, it is important to remember that patients with HSK do not have the protection of the lower ribs and the kidney may be compressed or lacerated against the lumbar vertebral column by blunt abdominal trauma.<sup>[16]</sup>

## Transplant

The first HSK transplantation reported in the literature was performed by Nelson and Palmer in 1975.<sup>[17]</sup> More than 100 HSK was transplanted in the last three decades. HSK can be transplanted into a single recipient, en bloc, or divided in the renal isthmus and transplanted into 2

patients.<sup>[18]</sup> Careful anatomic inspection of these kidneys and consideration of the donor and recipient parameters are key factors to the success of the transplant.

To preserve multiple or anomalous renal vessels or if the urinary collecting system crosses the midline, the removal of the HSK must be performed en bloc with a large segment of Ao and IVC.<sup>[19]</sup> When distal Ao is too short for perform the anastomosis the distal ends of common iliac arteries can be joined producing a conduit for arterial anastomosis and preserving renal arteries coming from distal Ao, common iliac arteries and bifurcation of Ao.<sup>[4]</sup> In our case, the recurrent left renal artery arises lose to the aortic bifurcation, therefore, if necessary this conduct could be applied.

Previous studies showed that the results of HSK transplantation, either en bloc or split, are equal to the post-transplant results of kidneys with a normal anatomy.<sup>[20]</sup> Finally, we conclude that HSK must be considered for renal transplantation, in view of the large waiting list of patients for this procedure.

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