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Anatomical and Clinical Aspects of Horseshoe Kidney: A Review of the Current Literature

Aspectos Anatómicos y Clínicos del Riñón en Herradura: Una Revisión de la Literatura Actual

Krzysztof Balawender[1,2]; Agnieszka Cisek[1]; Ewelina Cisek[1] & Stanislaw Orkisz[1]

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SUMMARY: Horseshoe kidney (HSK) is the most common kidney malformation. This review analyses the literature surrounding the etiology, morphology and clinical anatomy of these kidney fusion anomalies. A systematic literature search was carried out using the Science Direct and Scopus applications. HSK is characterized by three anatomic abnormalities: ectopia, malrotation and vascular changes. The study put emphasis especially on blood supply and upper urinary tract changes in HSK's patients. Arterial blood supply was analysed not only basing on Graves pattern, but also a new model of supply created on horseshoe kidneys computed tomography was taken into account. In addition, study presents clinical aspects of especially significant pathology of HSK, like hydronephrosis, stones and urinary tract infection. This study focuses also on congenital anomalies associated with horseshoe kidney.

KEY WORDS: Horseshoe kidney; Renal fusion; Kidney vasculature.

INTRODUCTION

Horseshoe kidney (HSK) is the most common congenital renal fusion anomaly and is characterised by three morphological anomalies: ectopia, malrotation and changes in vascular supply. Jacopo Berengario da Carpi was the first person to describe this abnormality during autopsies in 1522 (Oktem *et al*., 2008). HSK usually consists of two renal masses fused at their lower poles by a parenchymal or fibrous isthmus (Natsis *et al*., 2014). The frequency of appearance is 1 per 400-600 births and occurs more often in men (2:1 ratio).The reported incidence of this abnormality in anatomical dissections varies in the literature from 0.15 % to 0.48 %. There is no proven genetic determination of HSK, although it has been reported in identical twins and siblings within the same family (Yoshinaga *et al*., 2002). From the clinical point of view, HSK in adults usually produces no symptoms, but it has important implications with regards to secondary renal pathology such as urinary tract infections, hydronephrosis or stone formation. The incidence of fusion anomalies appears in three age groups: in small children HSKs are diagnosed as part of a combination of malformations, in young adults during diagnosis of delayed menarche as a part of Turner syndrome (Glodny *et al*., 2009), while detection of HSK in adults is incidental during routine radiological procedures (transabdominal ultrasounds, computed tomography or intravenous pyelography) performed on them for other reasons.

Search of the literature. A systematic search of the literature was carried out using the Science Direct and Scopus applications. The search terms included: "horseshoe kidney", "kidney fusion", "kidney vascular" and "surgery". In total, 41 articles in journals were refreshed. The search was limited to articles in English with no date limit. Finally, 25 references were included in the manuscript.

Morphology and topography of horseshoe kidney. Morphological classification of HSK is based on the renal shape based on the appearance of two fused renal masses (Pawar *et al*., 2018). The U-shaped HSK is formed by medial fusion. In this configuration, kidneys may be placed in a symmetrical position, U-shaped kidneys are observed in 42 % of incidences of HSK. Another type, which is a result of lateral fusion relative to the vertebral column, is called L-shaped HSK (Fig 1). This type occurs in 58 % of incidences of HSK (Oktem *et al*.). The asymmetry is more commonly left dominant (70 %) (Glodny *et al*.). In over 90 % of cases, fusion between the kidneys occurs at the lower pole; however, we can also observe upper pole fusion resulting in "inverted HSK" (5-10 %). Kidneys ascend from the pelvis to the mesogastrium during the seventh week of fetal development and reach their final position by the end of the eighth or ninth week (Taghavi *et al*., 2016). HSK is found more commonly in

[1] Morphological Sciences Department of Human Anatomy, Medical Faculty University of Rzeszów Leszka Czarnego 4 35-615 Rzeszów, Poland.
[2] Department of Urology, Pope John Paul II Hospital in Zamosc, Poland Al. Jana Pawla II 21 22-400 Zamosc, Poland.

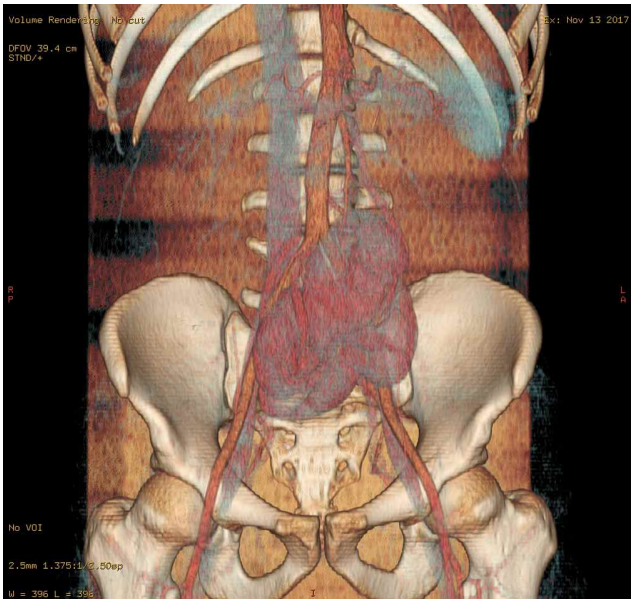

Fig. 1. Coronal reconstruction of axial CT scan of abdomen demonstrating a horseshoe kidney (morphological variant with lateral fusion). Image reproduced with permission from Department of Urology, Pope John Paul II Hospital in Zamosc, Poland.

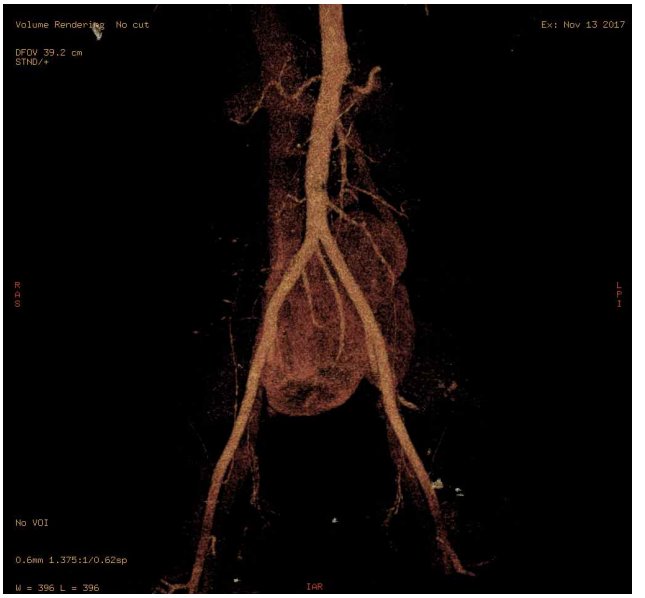

Fig. 2. Arterial supply of male horseshoe kidney. Image reproduced with permission from Department of Urology, Pope John Paul II Hospital in Zamosc, Poland.

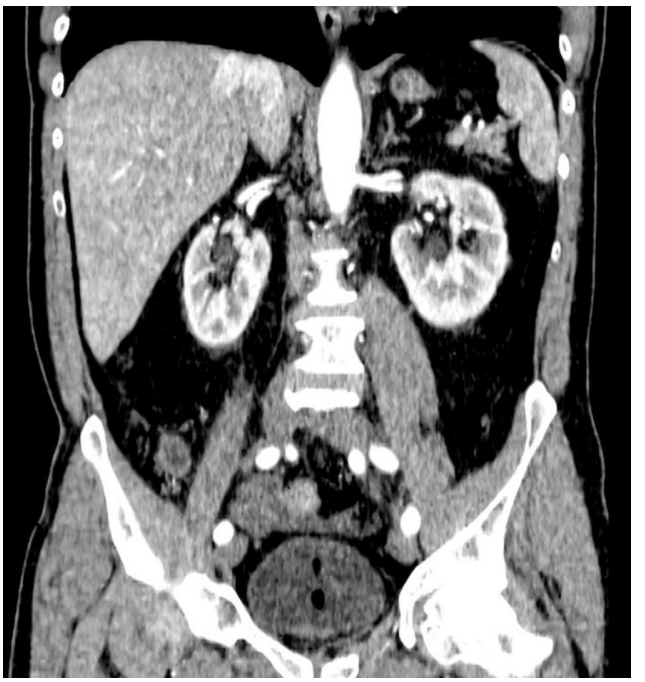

Fig. 3. Coronal reconstruction of axial CT scan shows a horseshoe kidney with a bilateral double renal arteries originate from abdominal aorta. Image reproduced with permission from Department of Urology, Pope John Paul II Hospital in Zamosc, Poland.

a lower position than a normally located kidney because the isthmus is prevented from ascending to its normal position by the inferior mesenteric artery (IMA) (O'Brien *et al*., 2008). The isthmus usually lies under the IMA origin anterior to the large vessels at the level of the third to the fifth lumbar vertebra (Decter *et al*., 1997). Less commonly, the isthmus is situated posterior to those vessels or runs between them. In 80 % of HSK cases, the isthmus contains renal parenchyma, with 20 % fibrous tissue (Taghavi *et al*.). Knowledge about the structure of the isthmus tissue is crucial from clinical point of view in planning HSK surgery, as well as in planning abdominal aorta surgery by laparotomy in vascular surgery.

Vascular system of the horseshoe kidney. Kidney fusion anomalies show great variation in the origin, number and size of renal vessels, depending on where the ascent terminated during development. The vascular pattern of HSK that was proposed by Graves in 1969 nowadays has a rather historical meaning. Graves separated the vascular anatomy of HSK into six groups. In this study, renal arteries can originate from the abdominal aorta, common iliac arteries and the inferior mesenteric artery (FigS. 2,3). The development of imaging techniques allowed a detailed analysis of the vascularisation of HSKs to be performed. Glodny *et al*. in a retrospective cohort study (level of evidence 2b) analysed more than 200 cases of kidney fusion anomalies and only 5 % of patients had a simple situation of only one artery on each side. Based on these studies, due to the wide range of possible arterial origins that include: common iliac artery (40 %), median sacral artery (2.9 %), internal iliac artery (1.94 %), external iliac artery (0.97 %), iliolumbar artery (2.9 %) and phrenic artery (0.97 %). What is more, Glodny *et al*. firstly made allowance for arteries that supplied portions of the contralateral kidney (this

occurs in approximately 25 % of HSK cases). The blood supply of the isthmus also has some variability: it may receive blood from the main renal artery, from the abdominal aorta (originating above or below the isthmus), the common iliac artery or the inferior mesenteric artery (Mano *et al*., 2016). Considering the embryological aspect, the isthmus blood supply reflects vascular changes of the developing kidneys during the course of kidney ascent from the pelvic to the abdominal position (Raman *et al*., 2018).

The incidence of kidney vein anomalies in HSKs is also high (23 %) (Pawar *et al*.). Frequently, HSK entails inferior vena cava (IVC) abnormalities, such as double, left and pre-isthmic IVC. Variations of IVC anatomy are observed ten times more frequently than in the general population (5.7 %) (Ichikawa *et al*., 2012). There is some data from case study reports about double IVC or pre-isthmic IVC (Kehagias *et al*., 1999). Moreover, HSK can be related to double superior vena cava due to the fact that their incidences are associated with cardiovascular malformations (Greenwood *et al*., 1976). Glodny *et al*. made general conclusions about the horseshoe kidney vasculature based on a retrospective cohort study. The cranial kidney vessels on both sides are located typically – the second artery on the left side is precaval, the second vein on the left side is usually retro-aortic. The caudal kidney vessels are located more ventral and often have more variation.

Changes in the upper urinary tract of horseshoe kidneys. Upper urinary tracts of HSKs are characterised by the great variation in their number and origin. Typically, calyces are located in the upper two-thirds of each kidney, but an external calyx or an independent ureter may drain the isthmus (Pawar *et al*.) (Fig. 4). The data from case study reports only shows anatomical variation of urinary tracts associated with HSK. Shen *et al*. (2012) described an HSK case with retrocaval ureters. Ongeti *et al*. (2011) presented bilateral ureteral duplication. Ureters usually end in the bladder, but they can also be found in an ectopic position (Cascio *et al*., 2002). Changes in a ureter's position in an HSK are a direct cause of secondary hydronephrosis and uretero-pelvis junction (UPJ) obstruction. UPJ obstruction can be the result of the high insertion of ureters into the renal pelvis, causing delayed pelvic emptying (Mano *et al*.). Rotation abnormalities of kidneys are also a significant cause of ureter obstruction. Normally, during the 6-8th week of gestation, the kidney hilum rotates from an anterior to a medial position. One of the anatomic abnormalities which is observed in HSKs is malrotation. The most common occurrences are incomplete rotations or non-rotations, but we can also observe hyper-rotation or reverse rotation. Nephrolithiasis, occurring in 16 % to 60 % of HSK cases (Glodny *et al*.) and urinary tract infections are clinical consequences of upper urinary tract abnormalities in an HSK.

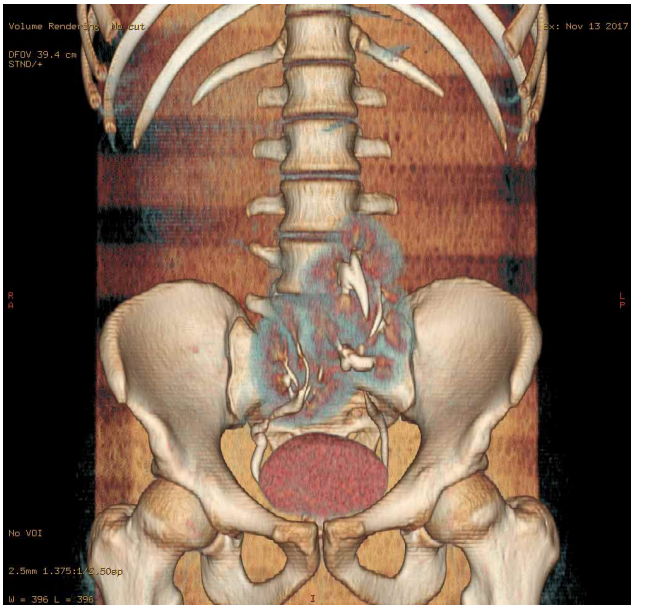

Fig. 4. Coronal reconstruction of axial CT scan of abdomen shows CT urography. Image reproduced with permission from Department of Urology, Pope John Paul II Hospital in ZamoSc, Poland.

Clinical manifestation of horseshoe kidney. HSK is usually asymptomatic and a horseshoe kidney is often discovered as an incidental finding. When symptoms are present, they usually occur because of obstruction, stones or infection. The most common finding in HSK is UPJ obstruction, which occurs in approximately 35 % of HSK patients (O'Brien *et al*.). UPJ obstruction is secondary to a high insertion of ureters into the kidney pelvis, which causes delayed pelvic emptying. Another cause of obstruction can be the crossing of the ureter over the HSK isthmus (Costa *et al*., 2004). A diagnosis of UPJ obstruction is made based on a CT urography or intravenous pyelography with a typical appearance of a large pelvis with a high-riding ureter in the pelvis. There are plenty of treatment options for clinically significant UPJ obstruction in HSK. Surgical technique includes pyeloureteroplasty, which nowadays is performed mostly using laparoscopic techniques. During these procedures, the obstructed UPJ area is excised and a new junction is created. The division of the isthmus has rather historical meaning because it is associated with an increased risk of serious complication, such as haemorrhage or renal infarction.

Another significant consequence of HSK is nephrolithiasis. The prevalence of stone disease in HSK is in the range 16-60 %. Based on Pawar *et al*. a meta-analysis of the estimated pooled incidence of kidney stones is 36 % (95 % confidence interval [CI]) in adults with HSK. Kidney stones are less common in the paediatric population with HSK with an estimated pooled incidence of 3 % (95 % CI).The incidence of kidney stones in HSK patients is much higher than in the

general adult populations (36 % vs 7 %). The etiology of stone disease in HSK is multicausal. HSK is related to an atypical position of the ureter in the renal pelvis and an usually highly placed UPJ. Another causes are metabolic factors (hypercalciuria, hyperoxaluria, hyperuricosuria and hypocitraturia) which are common in HSK patients and the coexistence of HSK with medullary sponge kidney (a known cause of high-risk stone disease). Pawar *et al.* also assessed the type of kidney stones in HSK stone formers. About 89.2 % of kidney stones were calcium-based stones, 4.2 % were struvite stones, 3.8 % uric acid stones and 2.8 % had other causes. Nephrolithiasis in HSK patients is often multiplied by the risk of large staghorn stones. Treatments are similar to those for a normal kidney. Extracorporeal Shock Wave Lithotripsy (ESWL) or Retrograde Intrarenal Surgery (RIRS) is usually used as a surgical treatment option. Percutaneous Nephrolitothrypsy (PCNL) as a endourological treatment for staghorn stones in HSK patient may be difficult to perform because of the different orientation of kidney calyx. A lower pole approach is taken when possible in a normal kidney because of the lower incidence of complications. In an HSK patient, a percutaneous approach is difficult due to the medial location of the calyx, so a cranial and lateral approach is more often required (Kehagias *et al.*). Eryildirim *et al.* (2017) assessed the efficacy and safety of two different techniques – PCNL versus RIRS – in the management of stones in patients with HSK. Stone-free rates (about 80 % after 1 week period) and the percentage of cases with residual fragments were comparable in both techniques. Complication rates were found to be similar in both groups. Thus PCNL and RIRS could be safe and effective minimally invasive procedures for kidney stone removal in patients with HSK. An alternative technique in the management of kidney stones in HSK can be a laparoscopic approach. In a prospective randomised study, Fawzi *et al.* (2017) compared Transperitoneal Laparoscopic Pyelolithotomy (TPLP) and RIRS for the treatment of kidney stones in HSK patients. Based on the study, TPLP was an effective technique with higher stone-free rates; however, RIRS had merits – less invasive, shorter operative time and lower complication rates (Fawzi *et al.*).

Horseshoe kidney is notably predisposed to urinary tract infections (Natsis *et al*., 2005). Infection occurs as a result of a combination of reflux disease, stasis and stone formation and is an important cause of death. It is found in up to one-third of patients with HSK (Cascio *et al.*). Ascending infection is the most common type, usually caused by vesicoureteral reflux, which is presented in approximately 50 % of HSK patients (O'Brien *et al.*).

A variety of benign and malignant tumours are associated with HSK. The increase in malignancy is thought to be secondary to teratogenic factors present at birth (O'Brien *et al.*). The most common type of tumour is renal cell carcinoma, which constitutes 45 % of all HSK tumours (Fazio *et al*., 2003). Transitional cell carcinoma accounts for 20 % of tumours in patients with HSK (three to fourfold relative increased risk with HSK). This increased risk comes from chronic infection, stones and associated obstruction in the upper urinary tract. The incidence of carcinoid and Wilms tumours is also higher than in the general population (Talpallikar *et al*., 2001) HSK causes a 62-fold increase in the risk of renal carcinoid tumour in comparison with the normal population (Krishnan *et al*., 1997). Wilms tumour accounts for 28 % of malignant tumours (O'Brien *et al.*). Both of these malignancies have a predilection for location in the isthmus, probably related to teratogenesis during embryological development (Krishnan *et al.*).

Surgical treatment of tumours in HSK, due to the location of the mass, limited mobilisation of the fused kidney and its multiple arterial blood supplies, is difficult and usually needs treatment in an advanced urological centre. Nowadays, minimally invasive surgical techniques including laparoscopic and robot-assisted approaches are recommended (Raman *et al.*). Patients with a kidney cortical tumour may be treated with partial nephrectomy with limited blood loss and preservation of kidney function. However, the overall complication rate is relatively high (43 %) (Mano *et al.*).

Horseshoe kidney, due to the location in the lower part of abdomen and the presence of the isthmus across the midline, is associated with a higher risk of blunt abdominal trauma (Murphy *et al*., 1996).

HSK is not well protected by the ribs and may be injured across the lumbar vertebral column (O'Brien *et al.*). Thus, after lower abdominal trauma in HSK patient, especially with associated haematuria, radiological investigation to look for hematoma and other injuries is necessary.

Congenital anomalies associated with horseshoe kidney. HSK is associated with many congenital anomalies, especially central nervous system, skeletal or chromosomal abnormalities. 60 % of Turner syndrome and 20 % of Down and Edwards syndrome patients have a HSK (Natsis *et al*., 2005). There are case study reports showing spina bifida associated with HSK patients (Taghavi *et al.*). Henley *et al.* described supernumerary kidney with HSK (Natsis *et al*., 2014). Other anomalies may include undescended testis and hypospadias (O'Brien *et al.*).

CONCLUSION. HSK is the most common congenital anomaly of the urinary tract. These kidney fusion anomalies are mostly asymptomatic; however, significant symptoms,

especially hydronephrosis, stones and infections, constitute important causes of death in HSK patients. An abnormal kidney is usually related to a range of anatomical changes. Vessel relations and supply are highly variable. Furthermore, horseshoe kidneys are characterised by various ureteric configurations. All of these abnormalities have important clinical implications.

BALAWENDER, K.; CISEK, A.; CISEK, E. & ORKISZ, S. Aspectos anatómicos y clínicos del riñón en herradura: Una revisión de la literatura actual. *Int. J. Morphol., 36(4)*:12-16, 2018.

RESUMEN: El riñón en herradura (RH) es la malformación renal más común reportada en la literatura. Esta revisión analiza la literatura que rodea la etiología, morfología y anatomía clínica de esta anomalía de fusión renal. Se realizó una búsqueda sistemática de la literatura utilizando las aplicaciones Science Direct y Scopus. El riñón en herradura se caracteriza por tres anomalías anatómicas: ectopía, malrotación y cambios vasculares. El estudio puso énfasis especialmente en el suministro de sangre y los cambios del tracto urinario superior en los pacientes con RH. El suministro de sangre arterial se analizó no solo basándose en el patrón de Graves, sino que también se tuvo en cuenta un nuevo modelo de suministro creado en los riñones en herradura. Además, el estudio presenta aspectos clínicos de patología especialmente significativa de RH, como hidronefrosis, cálculos e infección del tracto urinario. Este estudio se centró también en las anomalías congénitas asociadas con el riñón en herradura.

PALABRAS CLAVE: Riñón en herradura; Fusión renal; Vasculatura renal.

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Corresponding author:
Krzysztof Balawender
Morphological Sciences
Department of Human Anatomy
Medical Faculty University of Rzeszów
Leszka Czarnego 4 35-615
Rzeszów
POLAND

Email: balawender82@gmial.com

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