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Surgical Problems and Results in Horseshoe Kidney

Atnalı Böbrekte Cerrahi Sorunlar ve Sonuçlar

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Abstract

Objective: We aimed to evaluate the clinical features, accompanying surgical problems, and renal development outcomes during nephrological follow-up in patients with horseshoe kidney (HSK).

Method: We retrospectively reviewed the medical records of 24 patients with HSK who underwent surgery in our pediatric surgery clinic between 2015 and 2023.

Results: Sixteen of the patients were boys and eight were girls. The mean age was 77.3 (1.5-192) months. The mean follow-up period was 48 (12-120) months. HSK was found incidentally in 10 patients and diagnosed prenatally in seven patients. Eleven children had bladder dysfunction and six patients were diagnosed with spina bifida. Thirteen of the patients were found to have frequent urinary tract infections. Anderson-Hynes pyeloplasty for ureteropelvic junction stenosis, ureteroneocystostomy for vesicourethral reflux (VUR) and ureterovesical stricture, vesicourethral injection sting for VUR, upper pole heminephrectomy for nonfunctioning dual system, Holmium laser-guided lithotripsy and unilateral nephrectomy, isthmusectomy and contralateral kidney nephron-sparing surgery were required due to bilateral Wilms tumor. During the postoperative follow-up, three patients continued to have urinary tract infections, five developed renal scarring, three developed proteinuria and four developed hypertension. A total of three patients, including two patients operated for Wilms tumor, had elevated cystatin-C levels and developed chronic kidney disease (CKD).

Conclusion: Patients with HSK should be followed up for urologic abnormalities that may require surgery and postoperative urinary tract infection and scar formation in the kidneys. In our study, it was demonstrated that surgical intervention alone cannot prevent CKD.

Keywords: Child, horseshoe kidney, retrospective study

Öz

Amaç: Atnalı böbrek (ANB) tanılı hastaların klinik özelliklerini, eşlik eden cerrahi sorunlarını ve nefrolojik takipte böbrek gelişim sonuçlarını değerlendirmeyi amaçladık.

Yöntem: 2015-2023 yılları arasında pediyatrik cerrahi kliniğimizde, ANB tanısı olup cerrahi endikasyon konularak ameliyat edilen 24 hastanın tıbbi kayıtları geriye dönük olarak incelendi.

Bulgular: Hastaların 16'sı erkek, sekizi kızdı. Yaş ortalamaları 77,3 (1,5-192) ay olarak hesaplandı. Ortalama takip süresi 48 (12-120) aydı. ANB 10 hastada tesadüfen bulunurken, yedi hastada prenatal olarak teşhis edildi. 11 çocukta mesane disfonksiyonu ve altı hastada spina bifida tanısı vardı. Hastaların 13'ünün sık üriner sistem enfeksiyonu geçirdiği tespit edildi. Hastalara üreteropelvik bileşke darlığı tanısıyla Anderson-Hynes piyeloplastisi, vezikoüretral reflü (VUR) ve üreterovezikal darlık tanılarıyla üreteroneosistostomi, VUR nedeniyle vezikoüretral enjeksiyon sting, nonfonksiyone çift sistem tanısıyla üst kutup heminefrektomi, taş saptanması üzerine holmium lazer eşliğinde litotripsi ve bilateral Wilms tümörü saptanarak tek taraflı nefrektomi, isthmusektomi ve karşı taraf böbrek nefron koruyucu cerrahi şeklinde cerrahi müdahale gerekti. Ameliyat sonrası takip sürecinde, üç hastanın idrar yolu enfeksiyonu geçirmeye devam ettiği, beşinde böbrekte skar oluştuğu, üçünde proteinüri ve dördünde hipertansiyon geliştiği görüldü. Wilms tümörü nedeniyle ameliyat edilen iki hasta ile beraber toplamda üç hastada sistatin-C değerlerinin yükseldiği ve kronik böbrek hastalığı (KBH) geliştiği saptandı.

Sonuç: ANB tanılı hastalar tetkik edilirken cerrahi gerektirebilecek ürolojik anormallikler ve ameliyat sonrasında özellikle üriner sistem enfeksiyonu geçirme ve böbreklerde skar oluşumu açısından takip edilmelidir. Çalışmamızda, cerrahi olarak müdahale edilse de tek başına cerrahi işlemin KBH'sini engelleyemeyeceği ortaya konuldu.

Anahtar kelimeler: Atnalı böbrek, çocuk, retrospektif çalışma

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Introduction

Horseshoe kidney (HSK), the most common congenital fusion anomaly of the urinary system, occurs in approximately one in 400 individuals (1). It is twice as prevalent in males (2). In 20% of cases, HSK is located within the pelvis, while the remainder are at the normal anatomical site (3). In over 90% of cases, the two kidney masses join at the lower poles by a parenchymal or fibrous band. In rare instances, the bridge connects the upper poles, forming an inverse HSK. Some kidneys may develop as nonfunctional or dysplastic (4).

These anatomical abnormalities predispose to drainage issues in the collecting system, urinary stasis, infection, and stone formation. While 30-90% of patients with HSK are asymptomatic, nearly one-third to half have additional urological or systemic abnormalities (5). Accompanying surgical pathologies, primarily causing obstructive hydronephrosis, may include ureteropelvic junction obstruction (UPJO), vesicoureteral reflux (VUR), ureterovesical junction obstruction (UVJO), duplex systems, malignancy, and stones.

Literature data lack long-term outcome information for patients with HSK. This study aims to assess the clinical features, surgical problems leading to obstructive hydronephrosis, and nephrological follow-up outcomes regarding kidney development in patients with HSK.

Materials and Methods

Between 2015 and 2023, at our pediatric surgery clinic, we retrospectively reviewed the medical records of 24 patients with HSK who were examined for this condition and subsequently underwent surgery. Demographic, clinical, laboratory, and radiological data were recorded. We assessed age, gender, clinical presentation, surgical pathology, frequency of urinary tract infections (UTIs), blood pressure (BP), serum creatinine (SCr), cystatin-C, urea, glomerular filtration rate (GFR), and urine protein (UP) levels, as well as the status at the latest follow-up.

The diagnosis of HSK was made using urinary ultrasonography and confirmed with radionuclide scanning scintigraphy. Kidney scars (KS) and renal cortical functions were determined using 99mTc-dimercaptosuccinic acid (DMSA) scintigraphy. KS was defined as a defect in contrast material uptake in the normal renal contour in subsequent DMSA scintigraphy (Figure 1). All patients underwent US and DMSA. A widespread decrease in radionuclide uptake indicated a congenital KS, while focused peripheral defects in DMSA scans were identified as acquired KS. All patients underwent bladder and urethral cystography to detect VUR, UPJO, UVJO, stones, or related urinary system abnormalities. VUR was graded from I to V according to the International Reflux Study Committee and decisions for injection or ureteroneocystostomy were made. Estimated GFR (eGFR) was calculated using the new Schwartz formula [eGFR (mL/min/1.73 m²) = 0.413 x height (cm) / SCr (mg/dL)]. Chronic kidney disease (CKD) was defined as a GFR of <90 mL/min/1.73 m². Patients were followed clinically and radiologically every three to six months with urine analysis, UP, SCr, BP measurements, and serial ultrasonography. Those with frequent UTIs underwent DMSA after six months.

Hypertension was defined as systolic and/or diastolic BP above the 95th percentile for age, gender, and height. BP data were recorded as the average of three consecutive measurements taken with a mercury sphygmomanometer after five minutes of rest. A UP to creatinine ratio >0.2 in the first morning urine sample was considered significant. The presence of significant bacteriuria (>105 cfu/mL) in the urinary culture determined the presence of a UTI.

The study was ethically approved by the relevant Institutional Ethics Committee of University of Health Sciences Turkey, Başakşehir Çam and Sakura City Hospital under the reference number 228-10.01.2021. As our study had a retrospective design, informed consent was not required from the patients.

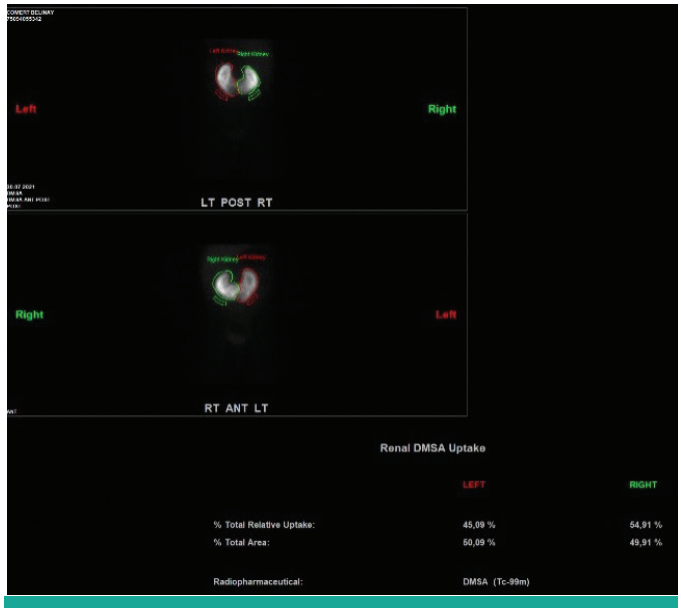

Figure 1. DMSA image
DMSA: Dimercaptosuccinic acid

Statistical Analysis

In this study, we employed a range of statistical techniques to analyze patient data. This involved using descriptive statistics to determine frequencies and characteristics specific to each variable. For continuous variables, we calculated the mean and standard deviation, or median and interquartile range, as appropriate for summarizing the data. We also evaluated the distribution of these continuous variables with the Shapiro-Wilk and Kolmogorov-Smirnov tests to check for normality. In cases where the continuous data did not exhibit a normal distribution, we opted for non-parametric methods over the standard t-test, which is better suited for normally distributed continuous variables. For the analysis of categorical variables, the chi-square test was primarily used, along with Fisher's Exact test in specific situations. All data processing and analysis tasks were carried out using SPSS Statistics software, Version 26.0 (IBM Corp., Armonk, NY, USA), with a p-value of less than 0.05 considered to indicate statistical significance.

Results

Of the patients, 16 were male (66.6%), and eight were female (33.3%). The average age was 77.3 months (ranging from 1.5 to 192 months). The mean follow-up period was 48 months (ranging from 12 to 120 months). HSK was incidentally discovered in 10 patients (41.6%) and prenatally diagnosed in seven patients (29.1%). In patients undergoing surgery for urinary system anomalies, diagnoses included VUR in 7 patients, UPJO in 8, duplex systems in 2, UVJO in 2, urinary stones in 3, and bilateral Wilms tumors in 2. Bladder dysfunction was noted in 11 children (45.8%), and six patients (25%) were diagnosed with spina bifida. It was found that 13 patients (54.1%) had frequent urinary system infections.

Surgical interventions were required for various conditions: Anderson-Hynes pyeloplasty for UPJO (n=8), ureteroneocystostomy for VUR and UVJO (7 patients), vesicoureteral injection-sting for VUR (n=3), upper pole heminephrectomy for nonfunctional duplicated systems (n=2), holmium laser lithotripsy for stones (n=3), and unilateral nephrectomy, isthmusectomy, and contralateral kidney nephron-sparing surgery for bilateral Wilms tumors (n=2) (Table 1).

During the postoperative follow-up of the surgically treated patients, it was observed that three patients (12.5%) continued to experience UTIs, five (20.8%) developed KS, three (12.5%) had proteinuria, and four (16.6%) developed hypertension. Including two patients who underwent surgery for Wilms tumors, a total of three patients (12.5%) developed CKD (Table 2).

The analysis of age at diagnosis revealed an average of 76.6 months (range: 1.5-187 months) for males and 79.2 months (range: 2-192 months) for females, with no significant difference between genders (p>0.05). Regarding the type of diagnosis, incidental findings were observed in 43.8% of males and 37.5% of females, while antenatal diagnoses accounted for 31.2% and 25%, respectively. Other included flank pain (6.25% in males and 12.5% in females), enuresis (12.5% for both genders), and hematuria (6.25% in males and 12.5% in females) (p>0.05). Bladder dysfunction was identified in 43.8% of males and 50% of females, VUR in

Table 1. Demographic and clinical characteristics

Age at diagnosis	**77.3 (1.5-192)**
	n (%)
Gender	
Male	16 (66.6)
Female	8(33.3)
Type of diagnosis	
Incidental	10 (41.6)
Antenatal diagnosis	7 (29.1)
Flank pain	2 (8.3)
Enuresis	3 (12.5)
Hematuria	2 (8.3)
Bladder dysfunction	11 (45.8)
Urinary tract abnormality	
VUR	7 (29.1)
UPJO	8 (33.3)
UVJO	2 (8.3)
Duplex system	2 (8.3)
Stone	3 (12.5)
Wilms tumor	2 (8.3)
Systemic abnormality	
Spinal deformity	6 (25)

VUR: Vesicoureteral reflux, UPJO: Ureteropelvic junction obstruction, UVJO: Uretterovesical junction obstruction

Table 2. Nephrological problems of patients with HSK who were operated on

Outcome	**n (%)**
Surgery	24 (100)
UTI	3 (12.5)
Renal scarring	5 (20.8)
Proteinuria	3 (12.5)
Hypertension	4 (16.6)
CKD	3 (12.5)

UTI: Urinary tract infection, CKD: Chronic kidney disease

Table 3. Comparisons in terms of gender

	Male (n=16)	Female (n=8)	p-value
Age at diagnosis (month)*	76.6 (1.5-187)	79.2 (2-192)	>0.05*
Type of diagnosis			>0.05[&]
Incidental	7 (43.8%)	3 (37.5%)	
Antenatal diagnosis	5 (31.2%)	2 (25%)	
Flank pain	1 (6.25%)	1 (12.5%)	
Enuresis	2 (12.5%)	1 (12.5%)	
Hematuria	1 (6.25%)	1 (12.5%)	
Bladder dysfunction	7 (43.8%)	4 (50%)	>0.05[&]
VUR	5 (31.2%)	2 (25%)	>0.05[&]
UPJO	5 (31.2%)	3 (37.5%)	>0.05[&]
UVJO	1 (6.25%)	1 (12.5%)	>0.05[&]
Duplex system	1 (6.25%)	1 (12.5%)	>0.05[&]
Stone	2 (12.5%)	1 (12.5%)	>0.05[&]
Wilms tumor	1 (6.25%)	1 (12.5%)	>0.05[&]
Spinal deformity	4 (25%)	2 (25%)	>0.05[&]

VUR: Vesicoureteral reflux, UPJO: Ureteropelvic junction obstruction, UVJO: Uretterovesical junction obstruction, *: Median (min-max) Mann-Whitney U test. [&]: Chi-square test

31.2% of males and 25% of females, UPJO in 31.2% of males and 37.5% of females, UVJO in 6.25% of males and 12.5% of females, and duplex systems in 6.25% of males and 12.5% of females. The presence of stones was noted in 12.5% of both males and females, while Wilms tumor was found in 6.25% of males and 12.5% of females. Spinal deformity was reported in 25% of both genders. Across all these categories, no significant gender differences were observed (p>0.05) (Table 3).

Discussion

In most cases, fusion occurs at the lower poles. This fusion is twice as common in males. It is often associated with a narrow pelvis, as typically seen in trisomy 18. Most fused kidneys are positioned lower than normal. HSKs can sometimes be associated with UPJO and may present with UTIs, abdominal masses, and hematuria in children. One alternative treatment is transperitoneal laparoscopic pyeloplasty, which allows for better exploration of the pyelocalyceal system and detection of anatomical anomalies like crossing vessels, more commonly found in HSKs (1-4). In our study, similar to the literature, we found that the condition was more common in male patients. Additionally, in most cases of HSK, the fusion involves the lower poles. In all 24 of our patients, the fusion was observed at the lower poles.

Consistent with existing literature, 30-90% of patients with HSK are asymptomatic. In our patient population, 41.6% were incidentally identified, often diagnosed during imaging procedures (5). Due to its anatomy and embryogenesis, HSK is prone to various complications (6). Variable arterial and venous blood supply, the presence of a midline isthmus, and abnormal positioning of the ureters contribute to the incidence of complications (7). Symptoms typically arise from urological abnormalities such as hydronephrosis, infection, or urolithiasis. Common presentations include abdominal pain, flank pain, nausea, vomiting, UTIs, hematuria, and decreased urine output. Pain that intensifies with hyperextension of the spine is a suspicious symptom (8). In our study, similar to what is reported in the literature, we observed pathologies like VUR, UPJO, UVJO, duplex systems, urinary stones, and tumors in patients undergoing surgery for urinary system anomalies. Additionally, some patients were diagnosed with bladder dysfunction and spina bifida.

In HSK, the most common urinary pathology observed is UPJO, present in approximately 35% of cases and can be bilateral (9). Similarly, in our study, 33.3% of the children were diagnosed with UPJO and underwent surgical intervention. UPJO is likely due to delayed pelvic discharge associated with the high placement of ureters to the renal pelvis. The intersection of the ureter over the isthmus may also

contribute to the obstruction (10). The diagnosis is typically made using intravenous pyelography (IVP), which shows a typical appearance of a dilated pelvis and an adynamic narrow transition zone between the pelvis and ureter. It can also be frequently detected with renal scintigraphy (11,12). In our cases, we primarily used ultrasonography and mercaptoacetyltriglycine (Mag-3) scintigraphy, and in doubtful cases, IVP was additionally employed for diagnosis. Among surgical techniques, options include open pyeloplasty or ureterocalicostomy, while more recent laparoscopic techniques feature dismembered pyeloplasty. This technique involves the removal of the narrowed section of the ureteropelvic junction and reconstruction of the renal pelvis and ureter by creating an anastomosis with the upper part of the renal pelvis (13). In the past, division of the isthmus was routinely recommended post-pyeloplasty to improve drainage. However, it is rarely performed today due to increased risks of complications like bleeding and renal infarction. In our study, all patients requiring surgical intervention had unilateral conditions and underwent open Anderson-Hynes pyeloplasty. There were no interventions on the isthmus, and no recurrences were observed in the follow-up period.

Patients with HSK are prone to infections due to a direct correlation with VUR, stasis, and stone formation (14). One-third of these patients experience frequent urinary infections, which are considered a significant cause of patient morbidity. Literature suggests that approximately half of the patients with HSK exhibit VUR, and it should be a primary consideration in patients with recurrent UTIs, proteinuria, and unexplained renal failure. In our study, in contrast to the literature, VUR was identified in 29.1% (7 patients) and surgical intervention was performed. However, this lower incidence compared to the literature might be due to the inclusion of only patients who underwent surgery in our study, while those managed conservatively were not considered.

In HSKs, the anatomical placement of calyces can hinder drainage, leading to stasis and stone formation (15). The treatment procedure is the same as for a normal kidney. In our study, 12.5% of the patients were found to have stones, for which holmium laser lithotripsy was performed. HSK is associated with various benign and malignant tumors (16). The most common associated cancer is renal cell carcinoma, accounting for 45% of these tumors. Transitional cell cancers constitute 20% of the tumors, and it has been reported that there is a three to four-fold increased risk in HSKs (17-19). This risk is thought to be partly related to chronic infection, stones, and obstruction in the affected kidney. Carcinoid and Wilms tumors also show an increased incidence in HSKs. Similarly, in our study, 8.3% of the patients were diagnosed with Wilms tumor and underwent surgical intervention.

Study Limitations

There are limited studies on the long-term outcomes of patients with HSK. It is believed that patients who are asymptomatic and managed conservatively generally have an excellent prognosis without the need for any treatment. In contrast, the patients included in our study required surgical intervention. These patients were monitored post-surgery, particularly for CKD progression. They were observed for indicators of CKD progression such as proteinuria, hypertension, SCr, cystatin-C levels, GFR results, and the development of KS. In the postoperative follow-up period, it was noted that three patients (12.5%) continued to experience UTIs, KS developed in five patients (20.8%), proteinuria was observed in three (12.5%), and hypertension developed in four (16.6%). Including two patients who underwent surgery for Wilms tumors, a total of three patients (12.5%) developed CKD.

This study has some limitations. First, the sample size is relatively small and represents the experience of a single center. Second, the follow-up period may be considered short.

Conclusions

Patients diagnosed with HSK often have accompanying urinary anomalies. Therefore, during examination, it is crucial to evaluate for urological pathologies that may require surgical intervention. Even after surgical procedures, close monitoring is essential, particularly for urinary system infections and the development of KS. In our study, when assessing surgical issues and outcomes in HSK, we found, in line with the literature, that surgical intervention alone cannot prevent the progression of CKD.

Ethics

Ethics Committee Approval: The study was ethically approved by the relevant Institutional Ethics Committee of University of Health Sciences Turkey, Başakşehir Çam and Sakura City Hospital under the reference number 228-10.01.2021.

Informed Consent: As our study had a retrospective design, informed consent was not required from the patients.

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