Urinary Tract Infections in Children with Horseshoe Kidneys: A Single-Center with 5 Years Experience

Burcu Bulum, Demet Tekcan, Gürkan Genç, Ozan Özkaya
Division of Pediatric Nephrology, Ondokuz Mayıs University, *Samsun, Turkey*

Abstract

Objective: Horseshoe kidneys is the most common congenital fusion abnormality of the urinary tract. Patients with this anomaly have additional urological or systemic abnormalities. In this study, we aimed to evaluate the clinical and radiological features, additional abnormalities, and frequency of urinary tract infection (UTI) of patients with horseshoe kidneys who were admitted to our clinic over the last 5 years and followed-up at our institution.

Material and Methods: All patients with horseshoe kidneys who were <18 years of age and had been admitted for the first time in the last 5 years or followed-up for at least 6 months in Ondokuz Mayis University, Pediatric Nephrology Department, were enrolled in the study. The medical records of each patient were reviewed for demographic features, clinical and laboratory findings, and radiological and scintigraphic evaluation results. Data were analyzed using SPSS for Windows (version 15).

Results: A total of 66 pediatric patients (40 males and 26 females) were enrolled in the study. The median age of the patients was 2.8 (0.1–16.2) years at the time of diagnosis. Horseshoe kidneys were detected incidentally in 26 (39.4%) patients when performing ultrasonography for any other reason. The most common (21.2%) presentation symptom was UTI in symptomatic patients. Twenty-three (35%) patients had at least one UTI during their lives. Furthermore, 52% of patients with UTI had recurrent UTIs and 33% of them had lower urinary tract dysfunction (LUTD). The frequency of LUTD in patients with horseshoe kidneys who had recurrent UTIs was borderline significantly higher than that in patients without recurrent UTIs (p=0.050).

Conclusion: Urinary tract infection (UTI) is common in patients with horseshoe kidneys. It is important to investigate the presence of LUTD in patients with recurrent UTIs. Recurrence of UTI can be largely prevented in these patients with proper treatment and close follow-up. *(J Pediatr Inf 2015; 9: 108-13)*

Keywords: Children, horseshoe kidneys, lower urinary tract dysfunction, urinary tract infection

Received: 17.03.2015
Accepted: 30.07.2015

Correspondence Address:
Burcu Bulum
E-mail:
burcubulum@yahoo.com

DOI:10.5152/ced.2015.2032

Introduction

Horseshoe kidney is the most common congenital fusion abnormality of the urinary tract, and is seen nearly in frequency of 1:400 and seen more than two times in males than females (1). Nearly one third of the patients with horseshoe kidneys anomaly are asymptomatic and the patients are coincidentally diagnosed with the disease due to radiological imaging done for other reasons (2-4). In the early development phases, placement of the kidneys in the dorsolumber region through subpolar fusion frequently in midline and medial rotation do not occur (3). These disorders cause the drainage to malfunction in the collecting duct system and may cause the formation of urinary stasis and renal stones (1, 5). This particular situation increases urinary tract infection (UTI) risk in patients with horseshoe kidneys anomaly. It was reported that urinary stasis and renal stone-related UTI prevalence in patients with horseshoe kidneys anomaly was 27% and 42% (3, 6-9). Genitourinary or systemic additional anomalies may frequently accompany the clinical picture in the patients with horseshoe kidneys anomaly. The most prevalent accompanying urinary system anomalies are renal stones, vesicoureteral reflux (VUR), ureteropelvic junction obstruction (UPJO) and hydronephrosis (2-4).

In this study, we aimed to evaluate the clinical and radiological features as well as the additional abnormalities and frequency of urinary tract infection (UTI) of the patients with horseshoe kidneys who were admitted to our clinic for the last five years and followed up in our institution.

Material and Methods

All patients with horseshoe kidneys who were <18 years of age that were admitted for the first time in the last five years (June 2009-June 2014) or had been followed up at least for six months in Ondokuz Mayıs University Pediatric Nephrology Department were enrolled into the study. Medical records of each patient were retrospectively reviewed for demographic features, clinical and laboratory findings, radiological and scintigraphic evaluation results. The patients with clinical urinary tract symptoms (dysuria, urinating intermittently, abdominal pain, side pain etc.) were diagnosed with UTI with the growth of at least 50,000 cfu/mL urine-pathogenesis in the urine analysis due the leucocyte esterase or nitrite test positivity or the presence of leukocyte or bacteriuria in the urine sediment in the urine analysis done with the fresh urine sample. Urine samples were taken through catheterization in young children and midstream urine in adolescents (10). Patients aged >4 with lower urinary tract symptoms (intermittency, urgency, manoeuvres to prevent voiding, dripping, daytime and night-time urinary incontinence etc.) were evaluated with "Pediatric Lower Urinary Tract Symptom Score" (PLUTSS) and the patients with the score of ≥9 were diagnosed with lower urinary tract dysfunction (LUTD) (11). Horseshoe kidneys anomaly was detected via urinary tract ultrasonographic examination (US) and confirmed by technetium 99 m-dimercaptosuccinic acid (DMSA) renal cortical scintigraphy. Renal scar and renal cortical functions were evaluated through DMSA scintigraphy. VUR diagnosis was made with voiding cystourethrography (VCUG) and UPJO diagnosis with technetium-99m-mercaptoacetyltriglicine (MAG-3) scintigraphy.

Statistical analysis

'SPSS for Windows (version 15, SPSS Inc; Chicago, IL, USA)' package program was used for data analysis. Compatibility of the data with normal distribution was evaluated by the Kolmogorov-Smirnov test. Variable compatible with normal distribution were defined as average±standard deviation; variables incompatible with normal distribution asmedian (range); categorical variables were defined as "%". Fisher's exact chi-square test was used for the comparison of percentages. Significance level was accepted as $p<0.05$.

Results

A total of 60 children (40 (61%) male and 26 (39%) female patients were included in the study. The median age of the patients was 5.2 (1.2-17.9) years during the study and 2.8 (0.1-16.2) years at the time of diagnosis. The median follow-up period of the patients was 6.0 (6-52) months. Horseshoe kidneys anomaly was incidentally detected in 26 (39.4%) patients while ultrasonography was performed for another reason and detected in 7 patients via prenatal ultrasonography. The most prevalent (21.2%) reason of admission for the rest of the symptomatic patients was UTI. When the stories of the patients were examined, it was revealed that a systemic disease accompanied the horseshoe kidneys anomaly in 21 (32%) patients. Reasons of admission and systemic diseases of the patients are illustrated in Table 1.

When the family stories of the patients were examined, it was found that there was consanguinity in 21.9% of the patients. We learnt that in 11 families (18%), a total of 11 other family members not included in the study had kidney and/or lower urinary tract anomaly or kidney disease. Six patients had congenital anomalies of the kidney and urinary tract (CAKUT) (Ectopic kidney (n=2), UPJO (n=2), horseshoe kidneys (n=1), double collecting system (n=1)), two patients had chronic kidney disease (CKD) and were on dialysis therapy, three patients had been nefrectomized before. Sixty six point six percent of the individuals with CAKUT were first-degree relatives of the patients.

It was revealed that 23 (35%) patients had experienced just one UTI episode and median number of UTIs was 2 (1-13). 48% of the patients only had one UTI and 26% had more than 3 UTIs. 19 (83%) patients had UTI before they were diagnosed with horseshoe kidneys anomaly. In the follow-up, 12 (63%) of these patients never had UTI one more time, frequency of UTI decreased in 2 (%11) cases. Four (6%) patients who never had frequency of UTI started to have UTI during the follow-up after the diagnosis. It was found that 67% of the patients had febrile UTI. The most frequently (82%) growing microorganism in the urine culture sample taken during UTI was "*E. coli*". Twenty percent of the growing microorganisms were the ones that grew extended spectrum beta-lactamase. LUTD was found in 14% (n=9) of the study group, in 9% (n=5) of the patients who did not have recurrent UTIs, and in 33% (n=4) of the patients who had recurrent UTIs. It was found that prevalence of LUTD was significantly high in a limited way in patients with

Table 1. Causes of patients' diagnosis and accompanying additional systemic diseases

Causes of diagnosis	
Other causes during US	26 (39.4%)
UTI	14 (21.2%)
Abdominal pain	13 (19.7%)
Antenatal US	7 (10.6%)
Urinary incontinence	4 (6.1%)
Hematuria	1 (1.5%)
Acute renal failure	1 (1.5%)
Additional systemic diseases	
Asthma	4 (6%)
Epilepsy	3 (5%)
Spinal deformity±Neurogenic bladder	3 (5%)
Congenital hypothyroidism	2 (3%)
Fanconi aplastic anemia	1 (2%)
Thalassemia minor	1 (2%)
Turner syndrome	1 (2%)
Chediak Higashi syndrome	1 (2%)
Tracheoesophageal fistula	1 (2%)
Hearing loss	1 (2%)
Anal atresia	1 (2%)
Cheilosis acid	1 (2%)
Hemangioma	1 (2%)

US: ultrasonography; UTİ: urinary tract infection

horseshoe kidneys anomaly who had recurrent UTIs in comparison to patients who did not have recurrent UTIs (p=0.050). Median PLUTSS of the patients with LUTD was 15 (12-20). Prophylactic antibiotics were given to 76% of the patients who had UTI and all had trimethoprim-sulfamethoxazole (TMP-SMX). The mean creatinine levels of the patients during the first admission and in the last control were 0.36±0.15 (0.16-0.87) mg/dL and 0.38±0.12 (0.18-0.74) mg/dL, respectively. The mean creatinine clearance of the patience during admission, was 146.2±38.9 (78.8-233.0). During the follow-up of the study group, proteinuria developed in one patient and hypertension in one patient. Hypoactivity in DMSA were detected in one of the patients in whom proteinuria was detected. The patient in whom hypertension developed was obese and did not have renal scar.

All the patients were subjected to urinary tract US and it was found that kidneys of all patients were subpolar fusion. In thirty nine (59%) patients, abnormal symptoms were detected via US. The most prevalent (23%) symptom was hydronephrosis. VCUG was performed in 18 (27%) patients and VUR was detected in two patients. DMSA renal scintigraphy was performed in 47 (71%) patients hypoactivity/heterogeneity were detected in 13 (28%) patients, renal scar in one patient and atrophic kidney in one patient. In twenty five (53.2%) patients, >10% relative renal cortical function difference was detected between the kidneys. MAG-3 scintigraphy were performed in 10 (15%) patients and unilateral UPJO was detected in two patients. As a result of all radiologic and scintigraphic evaluations, CAKUT in addition to the existing horseshoe kidneys anomaly were detected in 17 (26%) patients. These anomalies were; hypoplasia (n=12), ectopic (behind the urinary bladder) horseshoe kidneys (n=1), VUR (n=1), double collecting system+VUR (n=1), UPJO (n=2). The patient who had VUR and renal scar were administered subureteric injection. Hydronephrosis secondary to aberrant vein compression was detected in one patient who thought to had unilateral UPJO. Radiologic and scintigraphic evaluation results of the patients are illustrated in Table 2.

Discussion

In our study, UTI was found as the most prevalent cause of admission of the patients applied with any symptom and diagnosed with horseshoe kidneys. One third of the participating patients had UTI at least once in their life time. It was revealed that two third of the patients had febrile UTI, more than half had recurrent UTIs and 33% of these patients had LUTD.

Patients with horseshoe kidneys anomaly frequently are asymptomatic and are coincidentally diagnosed while ultrasonography was performed for another reason. In our study, it was revealed that 39% of the patients were coincidentally diagnosed. While abdominal pain in the literature was reported to be the most prevalent symptom in symptomatic patients, in our study UTI was found as the leading symptom (6, 7). Although prevalence of UTI in healthy children changes in relation of age and gender, it only changes between 2%-11.3% (12-14). Prevalence of UTI in patients with horseshoe kidneys anomaly in the literature was reported between 27%-42%; this rate is significantly higher in comparison to the UTI prevalence in healthy children (6-9). UTI prevalence in our patient group was 35%, which was compatible with the rate in the literature, a rate which is significantly higher in comparison to the UTI prevalence in healthy children.

Urinary tract infection, accompanying VUR and urinary stasis and renal stones develop collaterally in patients with horseshoe kidneys anomaly. While VUR prevalence in these patients was reported to 22% and 25% in different studies, in our study, it was found 11% (7,

Table 2. Radiological and scintigraphic evaluation results of the patients

Renal US (n=66)	
Hydronephrosis	15 (23%)
Hypoplasia	12 (18%)
Renal stones+Hydronephrosis	5 (8%)
Renal stones	3 (5%)
Increase of parenchyma echogenity	2 (3%)
Ectopy	1 (2%)
Double collecting system	1 (2%)
Baldder US (n=66)	
Increase in wall thickness	2 (3%)
Trabeculation	2 (3%)
VCUG (n=18)	
VUR	2 (11%)
DMSA (n=47)	
Hypoactivity/ Heterogeneity	13 (28%)
Renal scar	1 (2%)
Atrophy	1 (2%)
MAG-3 (n=10)	
Obstruction	2 (20%)

US: ultrasonography; VCUG: voiding cystourethrography; VUR: vesico ureteral reflux; DMSA: technetium 99 m-dimercaptosuccinic acid (DMSA) renal cortical scintigraphy; MAG-3: technetium-99m-mercaptoacetyltriglicine scintigraphy

15). Renal stones prevalence in patients with horseshoe kidneys anomaly, on the other hand, ranged between 20%-60% (2, 16-18). Renal stones prevalence in our study, on the other hand, was found 12%, which is figure lower than the previous studies. As a result of midline fusioned kidneys of the patients with horseshoe kidneys anomaly, ureters pass from the medial or front isthmus and this situation may cause stasis in the collecting system. Stasis is a condition that increases the risk of UTI. In our study, hydronephrosis prevalence in the patient with horseshoe kidneys was 23% and this prevalence is lower than the rates in previous studies (19, 20). Therefore, we are of the opinion that in our patient group, VUR, renal stones or stasis may cause UTI, but they may not be a prevalent cause for UTI.

LUTD is a clinical problem frequently seen in childhood. Prevalance of LUTD in healthy children was reported to be in a wide range as 6%-46% in different studies (21-23). The relationship between LUTD and UTI are well known and we are of the opinion that insufficient treatment of LUTD may be one of the most important causes of recurrent UTIs (24-28). In a previous study, prevalance of LUTD in patients with UTI was found as high as 59% (29). However, as far as we are concerned, no study is available in the relevant literature regarding LUTD prevalence in patients with horseshoe kidney anomaly. In our study, prevalance of LUTD in patients with horseshoe kidneys anomaly was 14%, LUTD prevalence in healthy children was not different. However, prevalance of LUTD in patients with recurrent UTIs was higher than the patients who did not have recurrent UTIs. Therefore, it is necessary to obtain a detailed LUTD story from the patients with horseshoe kidneys anomaly who had recurrent UTIs.

Although horseshoe kidneys anomaly is a rare anomaly, there is no sufficient number of studies regarding the long term prognosis of the patients who have this anomaly. The relationship between UTI and VUR, and renal scar and CKD is well known. In a recent study done by Yavuz et al. (7) in which they evaluated the renal prognosis of 41 patients with horseshoe kidneys anomaly, they reported that the prevalance of UTI was 42% and prevalance of renal scarring was 24%. Half of the patients had VUR with a renal scar. In the 10-year long follow-up period of the patients, it was revealed that proteinuria developed in 15% and hypertension in 10%. It was reported proteinuria, hypertension and renal scar presence constituted the independent risk factors for the development of CKD (7). In our study, on the other hand, prevalance of VUR and renal scar were found 11% and 2% respectively, and proteinuria was detected in only two patients and hypertension in one patient. We are of the opinion that this particular result might be to do with the short follow-up period in our study.

The most important limitation of our study is that it is a retrospective study and the short follow-up period of the patients. There is a need for long-term prospective studies inclusive of many more patients that will enable more detailed examination of the complications that develop in patients with horseshoe kidneys anomaly.

Conclusion

Urinary tract infection is a prevalent finding in patients with horseshoe kidneys anomaly. In these patients, UTI, accompanying VUR and urinary stasis and renal stones may develop collaterally. However, it is necessary to investigate the presence of LUTD in patients who have had recurrent UTIs. It was found that prevalence of LUTD in patients who had recurrent UTIs was higher than the patients who did not have recurrent UTIs. Recurrence of UTI and complications may largely be prevented in these patients through appropriate treatment and close follow-up.

Ethics Committee Approval: Ethics committee approval was not received due to the retrospective nature of this study.

Informed Consent: Written informed consent was not obtained from patients due to the retrospective nature of this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - G.G.; Design - B.B., G.G.; Supervision - G.G., O.Ö.; Funding - B.B., D.T.; Materials - B.B.; Data Collection and/or Processing - B.B., D.T., G.G.; Analysis and/or Interpretation - B.B., G.G.; Literature Review - B.B., D.T., G.G.; Writer - B.B.; Critical Review - G.G., O.Ö.; Other - B.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

References

1. Bauer SB. Anomalies of the upper urinary tract. In: Wein AJ, Kavoussi LR, Novick AC, et al. (eds). Campbell-Walsh Urology. 9th edition. Philedelphia: Saunders Elsevier; 2007. p3264-304.
2. Weizer AZ, Silverstein AD, Auge BK, et al. Determining the incidence of horseshoe kidney from radiographic data at a single institution. J Urol 2003; 170: 1722-6. [CrossRef]
3. Grainger R, Murphy DM, Lane V. Horseshoe kidney-a review of the presentation, associated congenital anomalies and complications in 73 patients. Ir Med J 1983; 76: 315-7.
4. Glodny B, Petersen J, Hofmann KJ, et al. Kidney fusion anomalies revisited: clinical and radiological analysis of 209 cases of crossed fused ectopia and horseshoe kidney. BJU Int 2009; 103: 224-35. [CrossRef]
5. Natsis K, Piagkou M, Skotsimara A, Protogerou V, Tsitouridis I, Skandalakis P. Horseshoe kidney: a review of anatomy and pathology. Surg Radiol Anat 2014; 36: 517-26. [CrossRef]
6. Khan A, Myatt A, Palit V, Biyani CS, Urol D. Laparoscopic heminephrectomy of a horseshoe kidney. JSLS 2011; 15: 415-20. [CrossRef]
7. Yavuz S, Kıyak A, Sander S. Renal outcome of children with horseshoe kidney: A single-center experience. Urology 2015; 85: 463-6. [CrossRef]
8. Evans WP, Resnick MI. Horseshoe kidney and urolithiasis. J Urol 1981; 125: 620-1.
9. Kawada S, Ichikawa T, Koizumi J, et al. Assessment of renal shape of horseshoe kidney with multidetector row CT in adult patients: Relationship between urolithiasis and renal isthmus. Tokai J Exp Clin Med 2013; 38: 159-66.
10. Subcommittee on Urinary Tract Infection, Steering Committee on Quality Improvement and Management, Roberts KB. Urinary tract infection: clinical practice guideline for the diagnosis and management of the initial UTI in febrile infants and children 2 to 24 months. Pediatrics 2011; 128: 595-610. [CrossRef]
11. Akbal C, Genc Y, Burgu B, Ozden E, Tekgul S. Dysfunctional voiding and incontinence scoring system: quantitative evaluation of incontinence symptoms in pediatric population. J Urol 2005; 173: 969-73. [CrossRef]
12. Coulthard MG, Lambert HJ, Keir MJ. Occurrence of renal scars in children after their first referral for urinary tract infection. BMJ 1997; 315: 918-9. [CrossRef]
13. Jakobsson B, Esbjörner E, Hansson S. Minimum incidence and diagnostic rate of first urinary tract infection. Pediatrics 1999; 104: 222-6. [CrossRef]
14. Mårild S, Jodal U. Incidence rate of first-time symptomatic urinary tract infection in children under 6 years of age. Acta Paediatr 1998; 87: 549-52. [CrossRef]
15. Cascio S, Sweeney B, Granata C, Piaggio G, Jasonni V, Puri P. Vesicoureteral reflux and ureteropelvic junction obstruction in children with horseshoe kidney: treatment and outcome. J Urol 2002; 167: 2566-8. [CrossRef]
16. Raj GV, Auge BK, Assimos D, Preminger GM. Metabolic abnormalities associated with renal calculi in patients with horseshoe kidneys. J Endourol 2004; 18: 157-61. [CrossRef]
17. Lampel A, Hohenfellner M, Schultz-Lampel D, Lazica M, Bohnen K, Thüroff JW. Urolithiasis in horseshoe kidneys: therapeutic management. Urology 1996; 47: 182-6. [CrossRef]
18. Symons SJ, Ramachandran A, Kurien A, Baiysha R, Desai MR. Urolithiasis in horseshoe kidney: a single-centre experience. BJU Int 2008; 102: 1676-80. [CrossRef]
19. Odiase VO. Horseshoe kidney. A review of 25 cases. J R Coll Surg Edinb 1983; 28: 41-5.
20. Segura JW, Kelalis PP, Burke EC. Horseshoe kidney in children. J Urol 1972; 108: 333-6.
21. Kajiwara M, Inoue K, Usui A, Kurihara M, Usui T. The micturition habits and prevalence of daytime urinary incontinence in Japanese primary school children. J Urol 2004; 171: 403–7. [CrossRef]
22. Chung JM, Lee SD, Kang DI, et al. Korean Enuresis Association. An epidemiological study of voiding and bowel habits in Korean children: A nationwide multicenter study. Urology 2010; 76: 215-9. [CrossRef]
23. Vaz GT, Vasconcelos MM, Oliveira EA, et al. Prevalence of lower urinary tract symptoms in school-age children. Pediatr Nephrol 2012; 27: 597-603. [CrossRef]
24. Snodgrass W. Relationship of voiding dysfunction to urinary tract infection and vesicoureteral reflux in children. Urology 1991; 38: 341-4. [CrossRef]
25. Winiecka W, Zoch-Zwierz W, Wasilewska A, et al. Evaluation of bladder instability in children with recurrent urinary tract infections. Med Sci Monit 2002; 8: 19-23.
26. Bakker E, van Gool J, van Sprundel M, van der Auwera JC, Wyndaele JJ. Risk factors for recurrent urinary tract infection in 4332 Belgian school children aged between 10 and 14 years. Eur J Pediatr 2004; 163: 234-8. [CrossRef]
27. McKenna PH, Herndon CD. Voiding dysfunction associated with incontinence, vesicoureteral reflux and recurrent urinary tract infections. Curr Opin Urol 2000; 10: 599-606. [CrossRef]

28. Naseer SR, Steinhardt GF. New renal scars in children with urinary tract infections, vesicoureteral reflux and voiding dysfunction: a prospective evaluation. J Urol 1997; 158: 566-8. [CrossRef]

29. Bulum B, Özçakar ZB, Kavaz A, Hüseynova M, Ekim M, Yalçınkaya F. Lower urinary tract dysfunction is frequently seen in urinary tract infections and is often associated with reduced quality of life. Acta Paediatr 2014; 103: 454-8. [CrossRef]