Pediatr Pol 2024; 99 (2): 1-4
DOI: https://doi.org/10.5114/polp.2024.139527
Submitted: 11.05.2023, Accepted: 20.12.2023, Published: 10.05.2024

PEDIATRIA Polska
Polish Journal of PAEDIATRICS

CASE REPORT

Bilateral infectious urolithiasis as a risk factor for acute kidney failure in a 3-year-old boy

Karina Madej-Świątkowska[1,2], Magdalena Błasiak[1,2], Monika Miklaszewska[3], Andrzej Haliński[4], Adam Haliński[4], Katarzyna Zachwieja[3], Anna Moczulska[3], Dorota Drożdż[3]

[1]University Children's Hospital, Kraków, Poland
[2]Doctoral School of Medical and Health Sciences, Jagiellonian University Medical College, Kraków, Poland
[3]Department of Pediatric Nephrology and Hypertension, Jagiellonian University Medical College, Kraków, Poland
[4]Department of Pediatric Urology, University Hospital of Zielona Góra, Zielona Góra, Poland

ABSTRACT

Infectious urolithiasis is a relatively rare form of urolithiasis in children. The predisposing factors in the pediatric population mainly include urinary tract defects and neurogenic bladder. Here we present a case of a 3-year-old boy with staghorn calculi, diagnosed during the diagnostic management for persistent leukocyturia. The boy had a history of recurrent urinary tract infections (rUTIs) caused by Proteus mirabilis and *Klebsiella pneumoniae*. Based on laboratory tests and imaging studies, the patient was diagnosed with a dysfunctional right kidney and a significant risk due to a large staghorn calculus to the left kidney. Bilateral ureteroscopic lithotripsy was performed, the deposit from the left kidney was removed via open surgery, and the narrowed sections of the ureters were excised. During the follow-up period, the patient's glomerular filtration rate remained normal. In the case of rUTIs kidney calculi may be diagnosed incidentally as they produce no clear clinical symptoms.

KEY WORDS:
children, nephrolithiasis, staghorn calculi, infectious urolithiasis.

INTRODUCTION

Urolithiasis (UL) is one of the most commonly diagnosed urological diseases. Infectious urolithiasis is a form of UL caused by the presence of bacteria in the urinary tract. In children, infectious UL is characterized by a variable frequency – occurring in 1–29% of all patients with urinary stones. The predisposing factors for the development of this type of UL in children are urinary tract defects and neurogenic bladder. Infectious stones are able to increase rapidly (within weeks or months) and fill the entire pyelocaliceal system. Untreated infectious UL can lead to kidney failure if not adequately diagnosed or treated [1, 2]. The diagnostic management, apart from taking the patient's and family history, includes diagnostic imaging, such as ultrasound (US) and low-dose non-contrast computed tomography (NCCT), as well as specific laboratory tests [3]. Treatment consists of the surgical removal of stones (most often by percutaneous nephrolithotomy – PCNL) in combination with targeted antibiotic therapy [1, 4].

CASE REPORT

A 3-year-old boy with recurrent urinary tract infections (UTIs) was admitted to the Department of Pediatric Nephrology and Hypertension due to persistent massive leukocyturia for several weeks despite being treated with oral antibiotics prescribed in an outpatient clinic. His perinatal and family history of kidney diseases were negative.

Prior to admission, the boy had experienced several episodes of symptomatic UTIs with etiology such as *Proteus mirabilis* and *Klebsiella pneumoniae*. The child was

ADDRESS FOR CORRESPONDENCE:
Karina Madej-Świątkowska, University Children's Hospital, Kraków, Poland, e-mail: madejkarina@gmail.com

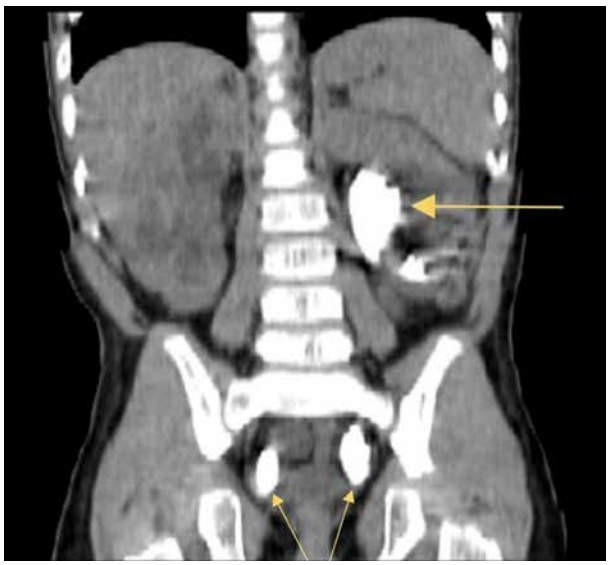

FIGURE 1. Staghorn calculus within pyelocalyceal system of the left kidney and stones in the lower part of both ureters (non-contrast computed tomography)

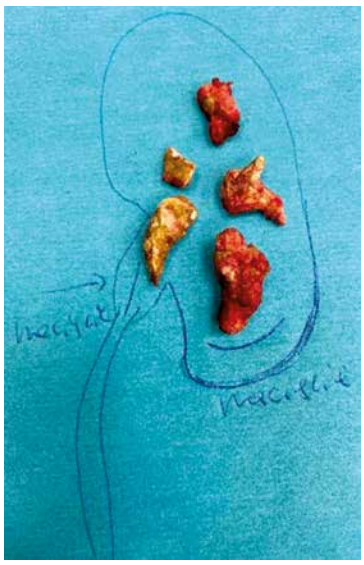

FIGURE 2. Surgically removed staghorn calculi compared to the real left kidney dimensions

treated at the Pediatric Urology Outpatient Clinic; however, a voiding cystourethrogram could not be performed due to persistent pyuria.

Two months earlier, during hospitalization at a regional hospital for a subsequent episode of UTI, the patient was diagnosed with hydronephrosis of the right kidney, accompanied by a tortuous and dilated entire-length ureter. No abnormalities were detected in the left kidney and left ureter.

On admission to the Pediatric Nephrology Department, the boy was in a good general condition, without any clinical symptoms of UTI. Laboratory tests revealed a slightly elevated leukocyte count (13.63×10^9/l; ref. range: $5.14–13.38 \times 10^3$/μl) and inflammatory markers (C-reactive protein – 16.3 mg/l; ref. range < 10 mg/l), while his renal function remained within the normal glomerular filtration rate (eGFR, according to the bedside Schwartz formula [5] was estimated to be 104 ml/min). In the urine sample collected by catheterization pyuria was determined, while urine culture confirmed the presence of *Proteus mirabilis* with a colony count growth of 10^5/ml.

Taking into account the previous urine culture results, the administration of ceftriaxone was initiated as the empirical treatment. Subsequently, an antibiogram report confirmed susceptibility to the administered antibiotic, indicating sensitivity to amikacin, ciprofloxacin, and third-generation cephalosporins.

The urinary system US revealed right-sided hydronephrosis with an anterior-posterior pelvic diameter of 30 mm. Additionally, a right-sided dilated ureter throughout its length was observed, with a suspected stone identified in its lower segment. Furthermore, a staghorn calculus was identified within the left kidney, not visualized in previous US examinations.

Due to the ambiguous US findings, in accordance with the European Association of Urology (EAU) Paediatric Urology Guidelines, a low-dose NCCT was performed [6]. The examination confirmed the presence of a 30 mm calculus in the distal segment of the dilated right ureter, as well as a staghorn calculus in the left kidney, with multiple calculi in the lower part of the dilated left ureter (Figure 1). Subsequently, renal technetium-99m diethylene triamine penta-acetic acid scintigraphy (RS DTPA) was performed, demonstrating a lack of right kidney function, while the function of the left kidney was well preserved.

Therefore, the patient was referred for complex UL treatment in a Pediatric Urology Department to decompress the urine outflow from both kidneys. A percutaneous paracentetic nephrostomy (PPN) was established on the right side, given unsuccessful attempts to introduce a ureteral catheter into the narrowed orifice of the right ureter. The left kidney was decompressed by inserting a double-J (DJ) catheter into the ureter.

Once the UTI episode resolved, bilateral incisions were performed at the level of the ureteral crossing with the iliac vessels (a procedure known as ureterolithotomy). During the procedure, six stones in the size range 5–10 mm were removed from the distal part of the left ureter, along with a 3 cm stone embedded in the wall of the right ureter. Additionally, two DJ stents were inserted bilaterally, and the PPN from the right kidney was removed. The analysis of the stone's chemical composition revealed numerous crystals of leucine, ammonium magnesium triphosphate and ammonium urates (which indicated the infectious pattern of the stones).

Following urine outflow decompression, RS DTPA was performed, which estimated right kidney function as 16% and left kidney function as 84%.

During this second stage of treatment, the staghorn calculus was removed from the left kidney through pyelolithotomy (Figure 2).

The third surgical intervention, conducted one month later, involved the reconstruction of bilateral stenotic megaureters at the vesicoureteric junction. The Hendren method was employed, accompanied by anti-reflux ureter re-implantation using the Laetbetter-Politano technique.

During the follow-up, of nearly two years, the boy has had no further UTI episodes, although the patient remains on continuous antibiotic prophylaxis (CAP) with furazidin (1.5 mg/kg). In testing, eGFR was normal, a 24-hour urine collection sample showed protein excretion remaining at a level of 7.8 mg/kg, with no evidence of hypercalciuria, hypocitraturia or hyperoxaluria (Table 1). Elevated blood pressure values, within the high normal range, were observed. Due to the elevated blood pressure and persistent proteinuria, treatment with ramipril (2 mg/m^2 BSA) was initiated. US revealed the right kidney to be smaller and with reduced corticomedullary differentiation, normal parenchymal echogenicity, while the pelvicalyceal system was not dilated. The left kidney demonstrated partially reduced corticomedullary differentiation, normal echogenicity, and bilateral punctual central echo enhancement, but there was no acoustic enhancement and no stones were observed.

After the passage of another half-year, the patient exhibited no proteinuria and his blood pressure values normalized.

TABLE 1. The patient's metabolic evaluation results

Laboratory tests	Results
Urine albumin to creatinine ratio [mg/g]	13
24-hour urine collection	
Protein [mg/kg]	7.8
Calcium [mg/kg]	0.5
Magnesium [mg/kg]	2.3
Citrate [mg/1.73 m ^ 2 BSA]	790
Oxalate [mg/1.73 ^ 2 BSA]	30

BSA – body surface area

DISCUSSION

Urolithiasis is a common urological disease. Over the past few decades, a significant increase in its incidence has been observed. According to the literature, in the human population, UL occurs in 5–10% of people throughout their lives, with only 2–3% of these cases occurring in children [7]. The incidence of such pathology is geographically varied. In Southeast Asia, the Mediterranean Sea region, India, and Pakistan, UL is an endemic disease. Compared to the adult population, UL in children does not significantly vary by sex and recurrences are more frequent, reaching up to 20% [8]. Predisposing factors can be found in more than 70% of children with UL, primarily including metabolic disorders (hypercalciuria, hypocitraturia, hyperoxaluria, hyperuricosuria, cystinuria), recurrent UTIs, and congenital malformations [8–10].

Stag-horn calculus is a special type of UL that causes the formation of large stones that partially or completely fill the renal pyelo-calyceal system. The shape of the calculi may resemble stag horns, hence their name [4, 11].

Staghorn calculi may be associated with recurrent UTIs, mainly of *Proteus* sp. etiology, with less frequent associations with *Providencia* sp., *Klebsiella* sp., *Pseudomonas* sp., *Morganella morganii*, *Citrobacter*, *Staphylococcus aureus* and enterococci [8, 10, 11]. These bacteria produce urease, which breaks urea down, producing ammonium ions and carbon dioxide. This process leads to urine alkalinization, which provides favorable conditions for the crystallization of struvite crystals (ammonium magnesium phosphate stones) and apatites (calcium phosphate stones) [2, 4, 10]. Rarely, staghorn calculi are formed as a result of metabolic disorders – from uric acid, calcium oxalate or cystine [8].

In a pediatric patient, the clinical presentation of the disease may be non-specific, which requires more attention since only a small percentage of cases present as classic renal colic [11].

Staghorn nephrolithiasis is characterized by a rapid dynamic development and, if not properly treated, can lead to a significant damage to the structure of the urinary tract, kidney failure and urosepsis [11].

Regardless of the UL type, the key factor of treatment is to provide free urine outflow in the entirety of the urinary tract while a benchmark of conservative therapy is adequate fluid intake and dietary modification, including limiting salt intake in food [2, 3, 9].

Additionally, prevention of staghorn calculi, which show a high correlation with UTIs, also focuses on preventing infections, including eliminating UTI recurrence-promoting factors and using long-term CAP to maintain urine sterility [8]. Urease inhibitors, such as acetohydroxamic acid, which was tested in adults, have not been employed in pediatric clinical practice due to their high toxicity [2].

The guidelines of the EAU and the American Urological Association recommend PCNL as the primary treatment for children with staghorn calculi, with open surgery and extracorporeal shock wave lithotripsy (ESWL) as secondary options [4, 6]. Percutaneous nephrolithotomy procedures are associated with a higher rate of complications, including bleeding and abdominal organ damage [12]. The American Urological Association does not recommend ESWL alone, except for patients with small renal stones and no congenital malformations [4]. On the other hand, open surgical treatment in the pediatric population is performed very rarely (mainly in cases where there is a need to remove a non-functional kidney, large stones, or in very large patients). Each case must be considered individually, as the treatment for staghorn nephrolithiasis is often multi-stage and requires the use of a numerous methods [4, 8, 11].

On the other hand, it has been unequivocally demonstrated that, in the absence of intervention, a staghorn stone will ultimately lead to renal destruction. Although UL is not a common etiology of acute kidney injury (AKI) in adults (comprising only 1–2% of cases) it may assume a more significant role in young children, accounting even for up to 30% of cases. The principal mechanism of AKI associated with UL is obstructive nephropathy, and the factors present at the time of its diagnosis can help prognosticate the likelihood of achieving long-term renal recovery, and impact the scheduling of further therapy [13].

In the described case, which involved a bilateral renal outflow obstruction due to calculi and concurrent obstructive uropathy (diagnosed as congenital bilateral vesicoureteric junction obstruction), a multistage treatment approach was required.

During the initial phase, relief of the infected urinary stasis was achieved through the insertion of a DJ catheter and the execution of a PPN procedure. Subsequently, the calculi were removed, and surgery involving stenotic megaureters was performed, effectively eliminating the cause of urinary obstruction and the formation of stones.

These procedures enabled the restoration of normal urinary flow from the kidney to the urinary bladder, effectively eliminating the primary cause of urinary calculi formation. During the long-term follow-up, the patient presented no UTI recurrences, no proteinuria, and his eGFR value remained normal.

CONCLUSIONS

Kidney staghorn calculi may be diagnosed incidentally, without any previous clear clinical symptoms. Low-dose NCCT is the preferred diagnostic approach for UL in children. Recurrent UTIs in children can potentially mask the presence of staghorn kidney stones. Urolithiasis should always be considered in cases of obstructive urogenital anomalies. In selective cases prompt decompression for urinary retention and correction of congenital obstructive anomalies may be the optimal intervention in pediatric UL.

DISCLOSURE

The authors declare no conflict of interest.

REFERENCES

1. Skrzypczyk P, Pańczyk-Tomaszewska M. Infectious urolithiasis. Pediatr Med Rodz 2019; 15: 33-37.
2. Gillespie RS, Stapleton FB. Nephrolithiasis in children. Pediatr Rev 2004; 25: 131-139.
3. Frassetto L, Kohlstadt I. Treatment and prevention of kidney stones: an update. Am Fam Physician 2011; 84: 1234-1242.
4. Preminger GM, Assimos DG, Lingeman JE, et al. AUA guideline on management of staghorn calculi: Diagnosis and treatment recommendations. J Urol 2005; 173: 1991-2000.
5. Horuz R, Sarica K. The management of staghorn calculi in children. Arab J Urol 2012; 10: 330-335.
6. Radmayr C, Bogaert G, Burgu B, et al. EAU Guidelines on Pediatric Urology. EAU Annual Congress Milan, Arnhem, The Netherlands 2023. Available from: http://uroweb.org/guidelines/compilations-of-all-guidelines.
7. Krzemień G, Szmigielska A, Jankowska-Dziadak K, et al. Kamica odlewowa nerki u małych dzieci – opis dwóch przypadków [Renal staghorn calculi in small children – presentation of two cases]. Dev Period Med 2016; 20: 23-26.
8. Wieczorkiewicz-Płaza A, Bieniaś B, Kusz M, et al. Clinical patterns of pediatric urolithiasis – a 4 year experience of a single center from south eastern Poland. Przegl Lek 2019; 76: 261-266.
9. Flannigan R, Choy WH, Chew B, et al. Renal struvite stones-pathogenesis, microbiology, and management strategies. Nat Rev Urol 2014; 11: 333-341.
10. Silva IE, Abreu GC, Moura WM, et al. Case report of staghorn calculi in a pediatric patient. J Bras Patol Med Lab 2019; 55: 295-304.
11. Kaygısız O, Türegün FA, Satar N, et al. Renal stone composition does not affect the outcome of percutaneous nephrolithotomy in children. World J Urol 2018; 36: 1863-1869.
12. Kumar R, Anand A, Saxena V, et al. Safety and efficacy of PCNL for management of staghorn calculi in pediatric patients. J Pediatr Urol 2011; 7: 248-251.
13. Tang X, Lieske JC. Acute and chronic kidney injury in nephrolithiasis. Curr Opin Nephrol Hypertens 2014; 23: 385-90.