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Research Article

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Malformative Uropathies of the Upper Urinary Tract in Children a Report of 42 Cases Collected at Albert Royer Children's Hospital

P. A. Mbaye[1], I B Welle[1], S. Camara[2], M. Fall[2], Y Diedhiou1 C. Seye[3], N. A. Ndoye[1], A. Sagna[1], O. Ndour[2] and G. Ngom[1]*

[1]*Albert Royer Children's Hospital, Cheikh Anta Diop University*

[2]*Aristide Le Dantec Hospital, Cheikh Anta Diop University*

[3]*Diourbel Regional Hospital, Alioune Diop University of Bambey*

Abstract

Objective: The aim of our study was to describe the epidemiological, diagnostic and therapeutic aspects of upper urinary tract malformative uropathies (UTMU) in children.

Methods: This was a retrospective, descriptive and analytical study conducted over four years (January 2021 - December 2024) in the pediatric surgery department of Albert Royer Children's Hospital, involving 42 cases of malformative uropathies of the upper urinary tract.

Results: Males predominated (54.76% of cases; sex ratio 1.2). The mean age at diagnosis was 74 months (late childhood). Hydronephrosis accounted for 57.14% of malformations, renal ectopia for 26.19% and multicystic renal dysplasia for 9.52%. Diagnosis was most often made postnatally, on presentation with abdominal pain (66.67%), vomiting (19.05%) or fever (14.29%), whereas only 21.43% of malformations were diagnosed antenatally. Renal ultrasound was performed in all patients, and ureterohydronephrosis (UHN) was the most frequent ultrasound abnormality (30.95%). Scintigraphy, which is of prime importance for assessing renal function and therefore for guiding the therapeutic strategy, was performed in only 23.81% of patients.

Treatment: Surgery was indicated in only 15 patients, all types of uropathy combined. More than half of these cases (8) were pyeloplasties with double-J stent placement for hydronephrosis; 2 left nephrectomies were also performed.

Conclusion: Malformative uropathies are fairly common in urological practice. UHN is the main condition observed, with diagnosis often made late, during later childhood. Antenatal diagnosis is of crucial importance, as it can help improve functional prognosis and tailor treatment. Although the rise of ultrasound has considerably improved prenatal diagnosis and prognosis, it unfortunately remains insufficient in our setting.

Keywords

Malformative uropathy, Upper urinary tract, Antenatal diagnosis, Treatment

Introduction

Congenital malformations of the urinary tract, also called malformative uropathies and often referred to by the acronym CAKUT (congenital anomalies of the kidney and urinary tract), comprise in children all the anomalies resulting from a disorder of the embryonic development of the urinary system, involving the kidneys and the excretory pathways.

Their nature varies and their severity differs, ranging from highly complex malformations to simple abnormalities of position or shape. They are more morphological than functional, with a risk of long-term impact on renal function. Today, uropathies can be detected by antenatal screening, which allows early management. Unfortunately, in our regions, these conditions are still diagnosed at the stage of complications, hence the need to study them further in order to better preserve and improve the renal prognosis of affected children [1].

***Corresponding author:** Papa Alassane Mbaye, Associate Professor, Cheikh Anta Diop University, Albert Royer Children's Hospital, 1 avenue Cheikh Anta Diop, Tel: +221 77 446 35 47; +221 70 487 16 44

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Table 1: Distribution of uropathies by age group.

Age group	Number	Percentage (%)
Neonates [0 to 28 days]	3	7.14
Infants [29 days to 30 months]	10	23.81
Young children [31 months to 5 years]	8	19.05
Older children [5 to 15 years]	21	50.00

Table 2: Antenatal ultrasound abnormalities (n = 9).

Abnormality	Number	Percentage (%)
Pelvicalyceal dilatation	5	55.56
Renal dysplasia (cyst)	4	44.44
Polyhydramnios	2	22.22
Bilateral adrenal hypertrophy	1	11.11
Horseshoe kidney	1	11.11

Table 3: Reasons for consultation (postnatal).

Reason	Number	Percentage (%)
Abdominal pain	28	66.67
Vomiting	8	19.05
Fever	6	14.29

It is in this context that we conducted this retrospective study, whose aim was to study upper urinary tract malformative uropathies (UTMU) in children in Dakar.

Patients and Methods

This was a retrospective, descriptive and analytical study evaluating the management of children with upper urinary tract malformations in the pediatric surgery department of Albert Royer/Fann Children's Hospital, from 1 January 2021 to 31 December 2024.

All patients under 15 years of age with an upper urinary tract malformative uropathy diagnosed or managed in the department were included. Forty-two (42) records met the selection criteria.

During data collection, we examined the following parameters:

- the patient's age and sex;
- personal and family history;
- clinical signs, collected by questioning the parents or the family members who accompanied the child;
- paraclinical investigations, both laboratory and radiological, performed to confirm the diagnosis and assess the repercussions;
- Finally, the therapeutic option, adapted to the patients' age and sex, the circumstances of discovery and the clinical forms, together with the treatments performed.

Results

Sex

There was a male predominance (23 boys, i.e. 54.76% of cases), with a sex ratio of 1.2.

Age

Older children (5 to 15 years) were the most represented, with 21 cases, i.e. half of the patients, followed by infants (10 cases), young children (8 cases) and neonates (3 cases) (Table 1). Among older children, there were 10 girls (23.81%) and 11 boys (26.19%).

Circumstances of discovery and history

In our series, antenatal ultrasound had allowed the diagnosis in 9 patients (21.43% of cases), whereas in 33 patients (78.57%) the diagnosis was made late, after birth. Antenatally, several ultrasound abnormalities could coexist; dilatation of the pelvicalyceal cavities was the most frequent (Table 2).

Postnatally, abdominal pain was the leading reason for consultation, reported by 28 patients (66.67%), followed by vomiting (8 patients, 19.05%) and fever (6 patients, 14.29%) (Table 3).

No family history of malformative uropathy was reported. Consanguinity was found in 11 patients (26.19%).

Physical examination

On admission, the physical examination was normal in 15 patients (35.71%); the same number of patients had abdominal pain. An abdominal mass, a lumbar contact and renal ballottement were each found in 4 patients (9.52%), and fever in 3 patients (Table 4).

Table 4: Physical examination findings.

Clinical signs	Number	Percentage (%)
Normal clinical examination	15	35.71
Abdominal pain	15	35.71
Lumbar contact	4	9.52
Renal ballottement	4	9.52
Abdominal mass	4	9.52
Urinary stream disorder	1	2.38
Fever	3	7.14
Urinary incontinence	1	2.38
Dehydration	1	2.38
Hepatosplenomegaly and small-necked umbilical hernia	1	2.38
Other	2	4.76

Table 5: Distribution of ultrasound abnormalities detected.

Abdominal ultrasound	Number	Percentage (%)
UHN	13	30.95
Renal ectopia	12	28.57
Pelvicalyceal dilatation	7	16.67
Renal dysplasia (multicystic kidney)	5	11.90
Ureteropelvic junction syndrome	3	7.14
Duplex urinary system	2	4.76
Empty right renal fossa, with a midline kidney and fusion of the lower poles	1	2.38
Major ureteral dilatation	1	2.38

Table 6: Distribution of UHN by side affected and grade.

Characteristic	Number	Percentage (%)
Involvement		
Bilateral	3	23.08
Unilateral	10	76.92
Laterality		
Bilateral	4	30.77
Right	3	23.08
Left	5	38.46
Not specified	1	7.69
Grade		
2	6	46.15
3	5	38.46
4	1	7.69

Table 7: Distribution of renal ectopia by laterality.

Renal ectopia	Number	Percentage (%)
Right	8	66.67
Left	3	25.00
Bilateral	1	8.33

Laboratory tests

Renal function: Assessed by measuring urea and creatinine, it was evaluated in 37 patients (88.10%); only 2 of them (5.4%) had renal failure. Mean creatinine was 8.7 ± 2.89 with a range of 5.4 to 19.

Urine culture (ECBU): This test was performed in 98% of patients; only one case of infection was found. It was an E. coli infection whose antibiogram showed susceptibility to third-generation cephalosporins.

Imaging

Abdominal ultrasound: All patients underwent abdominal ultrasound; the abnormalities found are listed in table 5. UHN was the most frequent ultrasound abnormality (13 cases, 30.95%), followed by renal ectopia (12 cases, 28.57%) and pelvicalyceal dilatation (7 cases, 16.67%). Ureteropelvic junction (UPJ) syndrome was found in [10 cases (23.81%) in the original text, 3 cases (7.14%) in the table: to be verified].

UHN was most often unilateral (10 cases, i.e. 76.92%), with the left side more frequently affected; grade 2 predominated (46.15%), followed by grade 3 (38.46%) (Table 6).

Renal ectopia was also frequent (12 cases); it was most often right-sided (8 cases, i.e. 66.67%) and was bilateral in only one case (8.33%) (Table 7).

Uro-CT: Computed tomography (CT) was performed in 31

Table 8: Uro-CT findings (n = 31).

Finding	Number	Percentage (%)
Ectopic kidney	9	29.03
Ureteropelvic junction syndrome	14	45.16
Ureterohydronephrosis	3	9.68
Obstructive right pelvic stone	2	6.45
Appearance suggestive of left megaureter	1	3.23
Left ureteral bifidity	1	3.23
Horseshoe kidney	1	3.23
Right multicystic renal dysplasia	1	3.23
Bilateral renal malrotation with anterior hilum	1	3.23
Other	2	6.45

Table 9: Surgical techniques performed in operated patients (n = 15).

Type of procedure	Number	Percentage (%)
Pyeloplasty + double-J stent	8	53.33
Extracorporeal lithotripsy	2	13.33
Left nephrectomy	2	13.33
Left pyelolithotomy + drainage	1	6.67
Left pyeloplasty + left pyelolithotomy	1	6.67
Left ureteral reimplantation	1	6.67

patients (73.81% of the series). UPJ syndrome was the most frequent finding (14 cases, 45.16%), followed by renal ectopia (9 cases, 29.03%) and then UHN (3 cases, 9.68%); multicystic renal dysplasia, horseshoe kidney and ureteral bifidity were each found in a single case (3.23%) (Table 8).

Renal scintigraphy: It was performed in 10 patients (23.81% of the overall population), always with the same technique (DTPA). Reduced renal function was observed in all patients examined, including 2 non-functioning kidneys and 1 case of bilateral reduced renal function.

Diagnosis

During the study period, hydronephrosis was the most frequent malformation (24 cases, i.e. 57.14%), followed by positional anomalies (11 cases, 26.19%), then multicystic renal dysplasia (4 cases, 9.52%)

Therapeutic management

Medical treatment: It was used in most patients, either for close monitoring of a well-tolerated, non-operated uropathy or for the treatment of any urinary tract infection before and especially after surgery; it was based mainly on antibiotic therapy.

Supervised non-operative management: This approach was adopted in half of the patients (21): those with moderate dilatation of the pelvicalyceal cavities with preserved cortical index and renal function, as well as all those with renal ectopia. Close clinical, ultrasound and scintigraphic monitoring showed a favorable course, with no unfavorable signs.

Surgical treatment: Surgery was indicated in 15 patients (35.71%): 8 pyeloplasties with double-J stent placement for UHN, 2 nephrectomies for non-functioning kidney, 2 extracorporeal lithotripsies performed at Idrissa Pouye General Hospital (HOGIP) for obstructive renal stones, 1 left pyelolithotomy with drainage, 1 left pyeloplasty combined with left pyelolithotomy, and 1 left ureteral reimplantation (Table 9).

Follow-up

Clinical follow-up was uneventful in 32 patients (76.19%); 7 patients were lost to follow-up and 2 deaths were recorded ; Ultrasound follow-up showed a favorable course, with no signs of obstruction in hydronephroses, and allowed close monitoring of the other abnormalities. Only one patient had a follow-up scintigraphy, which showed a retention syndrome of functional appearance, without obstruction.

Discussion

In our study, the distribution of upper urinary tract malformations by sex was heterogeneous, with an overall male predominance (23 patients, i.e. 54.76% of cases). This result is consistent with that of Kahloul et al. [2-4], who found a slight male predominance with a sex ratio of 1.08. It suggests that sex has little influence on the predisposition to upper urinary tract malformations.

These uropathies were diagnosed mainly postnatally (78.57% of cases), most often during later childhood (50% of patients). The mean age at diagnosis was 74 months, or about 6.2 years. This is close to the findings of Ndiath et al. [5,6], who reported a mean age of 5.3 years, and to other African

studies, such as one conducted in Morocco in 2006 [7], which, covering all uropathies, found a mean age at diagnosis of 2.5 years and no antenatal diagnosis.

These data contrast with the situation in northern countries, where diagnosis is made before birth thanks to ultrasound, within a multidisciplinary management framework: A French study from 2000 [8] reported a mean age at diagnosis of 11 months and 60% of cases diagnosed antenatally.

This diagnostic delay may be due to:

- the low rate of obstetric ultrasound among our pregnant women;
- the absence of health insurance covering this examination;
- the parents' belonging to disadvantaged socioeconomic groups;
- the clinical latency of uropathies and the lack of practitioners familiar with, or specialized in, urogenital malformative diseases.

In our series, the signs that prompted patients to consult were mainly abdominal pain (66.67%), vomiting (19.05%) and fever (14.29%), hence the importance of taking these signs properly into account A study conducted in Cotonou in 2013 on the relationship between urinary tract infection in children and urinary tract malformations showed that urinary tract infection was the most frequent reason for consultation (28%) [9-11].

Paraclinical data

Imaging assessment: Assessment of the urinary tract in children relies primarily on non-irradiating techniques, but also on other, more irradiating ones. As with any investigation in children, these examinations require good cooperation between the medical team, the child and the parents, and depend on a detailed explanation of the examination to be performed. The presence of the parents beside the child during the examination is recommended if they wish [12].

Renal ultrasound: This is one of the most accessible and harmless paraclinical examinations; however, it remains operator-dependent, which, together with the lack of specialists in urinary tract ultrasound, makes interpretation of the results sometimes difficult. All patients in our series underwent it, and it was often revealing; UHN was the predominant ultrasound abnormality (30.95%).

In the Tunisian study [2-4], 33% of patients had dilatation of the upper urinary tract; in the study by Faure [8], 30% had dilatation of the renal cavities. These results, like ours, show that uropathies often have repercussions on the upper urinary tract.

Uro-CT: Uro-CT is an imaging technique that faithfully reproduces the anatomy of the kidneys and provides details of their relationships and vascularization. In our study, it was performed in 31 patients (73.81%) and confirmed the diagnosis of UPJ syndrome and, above all, of anomalies of position and rotation.

Scintigraphy: This examination is increasingly indispensable: it assesses overall renal function as well as the separate function of each kidney, better defines the surgical indication and allows better evaluation of results. It was performed in 10 of our patients (23.81%), and involvement was most often unilateral (80%). It made it possible to determine the functional value of each kidney, with 70% of kidneys showing reduced function and 20% non-functioning kidneys requiring nephrectomy.

Renal function: Its assessment is useful for evaluating the prognosis of uropathies in general and their repercussions on the upper urinary tract. In our study, 94.6% of the patients assessed had normal renal function, which is in line with the Mauritanian study [13], which found 80% normal renal function. These results may be explained by the fact that, apart from delayed diagnosis and management, especially in bilateral uropathies or in solitary kidney, UTMU rarely cause renal failure.

Urine culture (ECBU): It was requested in 98% of patients and only one was positive, isolating E. coli; these results are consistent with those of the Cotonou studies [9-11].

Types of uropathy and treatment options

Hydronephrosis: Ureteropelvic junction (UPJ) syndrome, also known as primary hydronephrosis, is a common cause of upper urinary tract malformation [14,15]. In our series, hydronephrosis accounted for 57.14% of upper urinary tract malformative uropathies, which agrees with the frequency reported in the literature [16]. Its surgical management relied mainly on open pyeloplasty with resection and anastomosis of the junction according to the Anderson-Hynes technique. This technique was performed in 8 cases (19.05%); it is usually carried out through a lumbotomy: the renal pelvis is remodeled and reduced in volume, the stenosis is resected and the ureter is re-anastomosed.

Nephrectomy, based on scintigraphy data, is considered only when the kidney is non-functioning. We performed it in 2 patients (4.76%), a rate comparable to that in the literature [17-40].

Renal ectopia: It was the second most common upper tract uropathy encountered, just behind hydronephrosis. This positional anomaly was exclusively low-lying and most often unilateral, particularly on the right (66.67%). This is explained by the position and size of the liver, which hinders migration of the right kidney during embryonic development. Non-operative management was the therapeutic approach chosen for all our patients, with minor manifestations controlled by symptomatic treatment.

Ureterocele: Ureterocele is a rare congenital malformation, estimated at 1 in 4,000 births, which affects girls four to six times more often than boys [9-11]. Our series included only one case (2.38% of the uropathies studied), in a boy, managed surgically: ureterocelectomy followed by ureterovesical reimplantation with a Cohen-type anti-reflux procedure. In the international literature, however, endoscopic treatment remains the preferred option over open surgery because of the simplicity of the procedure and its low morbidity.

Conclusion

Upper urinary tract malformations comprise all the anomalies, more morphological than functional, resulting from a disorder of the embryonic development of the kidneys and their excretory pathways. They are common in pediatrics and constitute a public health problem. They affect boys slightly more often.

Their nature varies and their severity differs, ranging from highly complex malformations to simple abnormalities of position or shape. The risk is long-term impact on renal function.

The rise of ultrasound has considerably improved prenatal diagnosis and prognosis, but in our setting it unfortunately remains insufficient. Postnatally, abdominal pain and urinary tract infection are the main modes of presentation of these upper urinary tract malformative uropathies.

Diagnosis relies mainly on the radiological investigations available, and therapeutic decisions are based not only on the anatomical analysis of the malformation but also on its repercussions on renal function.

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