MALFORMATIVE UROPATHY IN CHILDREN: EXPERIENCE OF AN ALGIERS GENERAL PAEDIATRICS DEPARTMENT: UPDATE OF THE 2019 STUDY

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Summary

Introduction: Malformative uropathy (MU) is a common reason for consultation in paediatricuronephrology. The circumstances of discovery vary according to age, and the prognosis depends on the type of malformation observed. Objectives: The aim of this study is to assess a twenty-year period of follow-up of malformative uropathies, studying the epidemiological, clinical, therapeutic, and evolutionary aspects in a general paediatrics department in Algiers. This is a retrospective study conducted on children with malformative uropathy between January 1, 2001 and December 31, 2019. The study examines their epidemiological, clinical, paraclinical, and evolutionary profile. Out of 223 selected files, 194 were analyzed. **Results:** The study found a sex ratio of 1.7, indicating a male predominance across all ages. The mean age at diagnosis was 23 months (ranging from 1 day to 15 years), with 116 patients (59.8%) being less than 12 months old. More than half of the cases (53%) were diagnosed before the age of six months. Urinary tract infections were the most common circumstances of discovery, accounting for 50.5% of cases. Antenatal diagnosis was only found in 32.4% of cases. In this study, it was found that one-third of the patients had vesicourethral reflux disease (VU) (33%), 26.8% had megaureter (MU), pyeloureteral junction syndrome (PJS) was present in 24.2% of patients, while posterior urethral valves (UPV) accounted for only 6.2% of cases. VUR remains the most benign uropathy with a spontaneous cure rate of over 66%. Regarding renal function, it is disrupted in severe bilateral forms, particularly in patients with posterior urethral valves. It was concluded that malformative uropathies are relatively frequent, mainly obstructive pathologies, and they cause a third of chronic kidney failure in children. The prognosis of malformative uropathy is primarily determined by early diagnosis, the type of uropathy, and appropriate management. The study focused on vesicoureteral reflux and antenatal diagnosis. This study was conducted by the Department of General Pediatrics at Children's Hospital.

Keywords: Malformative Uropathy; Department Of General Pediatrics, Children; Vesicoureteral Reflux; Antenatal Diagnosis.

1. INTRODUCTION

Malformative uropathy (MU) is relatively common in paediatrics, accounting for 25% of paediatric surgical pathologies [1]. In the medical bibliography, its prevalence is estimated at 3.1/1000 births [2]. Antenatal diagnosis by obstetric ultrasound allows a significant reduction in the morbidity and mortality associated with this condition, thanks to early diagnosis and appropriate treatment. In developing countries, diagnosis is often delayed, usually because of an infectious complication [1.3.4]. In order to study the epidemiological, clinical, paraclinical and evolutionary aspects of this type of malformation, it was interesting to carry out an evaluation of these malformative uropathies in our general paediatrics department in Algiers.

2. MATERIALS AND METHODS

It is a retrospective study over a period of fifteen years (from 1 January 2001 to 31 December 2019) with a minimum follow-up of 04 years. 194 cases were included among the 223 cases of malformative uropathy collected. The study was conducted in the General Paediatrics Department "A" of the Benimessous University Hospital in Algiers, Algeria, and focused on children aged 0 to 15 years. Neurological bladders were excluded from this study. All patients underwent abdominal ultrasound and urinary cytobacteriology (ECBU). ECBU was considered positive if the bacteriology was greater than 105 CFU (colony forming units) per ml [5]. Retrograde cystography (CR) was performed in the absence of UTI in all patients with pathological ultrasound and/or recurrent UTI. Obstructive uropathies were diagnosed by means of intravenous urography, as long as we had a normal renal function; otherwise, the diagnosis was made by means of Uro-MRI, with the necessary precautions [6]. At the end of this study, we analysed the epidemiological, clinical, bacteriological, therapeutic and evolutionary data.

3. RESULTS

There were a total of 194 cases, 123 boys and 71 girls, with a sex ratio of 1.7. The mean age at diagnosis was 23 months (1 day - 15 years). Fifty-three percent (53%) were diagnosed at less than six months of age (6 months). There was a male predominance in all cases (63.4%). The circumstances of discovery were dominated by urinary tract infection (50.5% of cases), whereas antenatal diagnosis, currently considered to be the main mode of discovery of UM, was made in only one third of cases, i.e. 32.4% (Table I). The infectious syndrome was the predominant clinical presentation, found in fifty-nine patients (30.4%). The remaining patients had no clinical signs of infection (Table I).

Parameters	Number	Percentage
Total number	194	100%
Gender :		100%
Male	123	63 1%
Female	71	36.6%
Sex ratio	1,7	30,0 %
Age :		
< 1 month	51	26.2%
1-6 months old	52	20.270
6-12 months old	13	20.0%
12 to 24 months	21	10.8%
2 years -6 years	34	17.5%
> 6 years	23	11.5%
mean ageage (months)	22,9	11.070
Circumstances of discovery		
Antenatal ultrasound	63	32,4%
Urinary tract infection	98	50,5%
- Low	39	20,1%
- High	59	30,4%
Urinary stream abnormality	21	10,8%
Abdominal pain	18	09,2%
Clinical :		
Infectious syndrome	58	29,9%
Mucocutaneous pallor	21	10,8%
Bladder globe	09	04,6%
Lumbar contact	18	09,2%
Dehydration	07	03,6%

Table I: Distribution of the Number of Malformative Uropathies According to Sex,Age, Circumstances of Discovery and Clinical Picture

The majority of the 59 patients with acute pyelonephritis had a positive inflammatory function: C-reactive protein (CRP) was positive in 86.8%, erythrocyte sedimentation rate greater than 50 mm in the first hour in 89%, and neutrophil hyperleukocytosis greater than 12000 elements/mm3 in 75%. Urine cytobacteriology (ECBU) was positive in one hundred and twelve cases (57.7%), with Escherichia coli (E. coli) isolated in seventy-three cases (66%), resistant to aminopenicillins in forty-four cases (59.4%) and sensitive to cephalosporins in all cases (100%).

Renal ultrasound, which was performed in all patients, was abnormal in 191 patients (98.4%). Retrograde cystography (CR) was performed in all cases, while the practice of intravenous urography (IVU) was determined by the results of renal ultrasound, CR and renal function. Vesicourethral reflux (VUR) accounted for one third of cases (33%), followed by megaureter (MU) (26.8%) and pyeloureteral junction syndrome (PJS) (24.2%), while posterior urethral valves (UPV) accounted for only 6.2% of cases (Table II). The involvement was unilateral in 124 cases (63.9%) for all pathologies combined. For all RVUs, stage III was the most common form (51.8%); 19 of 52 had a severe form of SJPU. IVU confirmed the diagnosis of obstructive MU (MU-O) in 52 children (26.8%); again, the severe form (type III) was the most common (Table II). In 62 and 76% of SJPU,

70 and 70.7% of MU-O and 100% of UPV, respectively, static renal scintigraphy with 99mTc-labelled dimercaptosuccinic acid (DMSA) and dynamic 99mTc-MAG3 (MAG3) or 99mTc-labelled pentaacetic acid-diethylenetriamine acid (DTPA) with furosemide hyperdiuresis assay was pathological.

In cases of acute pyelonephritis, probabilistic first-line antibiotic therapy consists of a third-generation cephalosporin (combined with an aminoglycoside in infants younger than 3 months); the duration of treatment was ten days in the majority of cases (93%).

Fifty-one patients (26.2%) underwent surgical correction adapted to each type of malformative uropathy (Table III). Recurrence of infection was common in patients with VUR (89%), but much less common in patients with reflux. while it was much less common in patients with SJPU (48%).

The majority of patients maintained good growth in stature and weight, with normal renal function (85.5%). VUR was the mildest UM with a spontaneous total cure rate of 55.5%, in contrast to UPV where the impact on the upper tract was constant and severe.

Concerning renal function, the majority of patients (91%) in the study received a renal assessment (blood urea, serum creatinine and glomerular filtration rate); this function was disrupted in severe bilateral forms, particularly in children with posterior urethral valves (91.6%).

4. DISCUSSION

Epidemiologically, 194 cases of UM were collected over 20 years, a figure that can be compared with the 68 cases collected in 1984 by seven general paediatric departments in the Algiers region over a period of 8 years [7].

A male predominance was found in the overall study of UMs, excluding VURs, which is consistent with data from the literature [8].In contrast to developed countries, where the antenatal diagnosis of UM is in the range of 60 to 70% [9,10], in our series, as in that of developing countries, the diagnosis during pregnancy is much lower (32.4%) [8]. However, if we compare these results with those of the previous study [10], we note an improvement in the antenatal diagnostic means with an increase of 13.6% (18.8% VS 32.4%), with a decrease in the mean age of diagnosis (27.8 VS 22.9%).Postnatally, urinary tract infection remains the most common mode of disclosure, which is consistent with the circumstances of discovery reported in the literature [12-14]. The diagnosis can also be made following abdominal pain or urinary dysfunction, or be discovered incidentally following systematic urinalysis or abdominal ultrasound performed for another pathology.

The most common organism isolated in our series was E. coli, with 100% susceptibility to cephalosporins; this explains why the treatment of pyelonephritis most often consisted of the prescription of a third-generation cephalosporin (GC3), with or without an aminoglycoside, followed by prophylaxis with cotrimoxazole [15-17].

Paraclinically, abdominal ultrasound was performed as a first-line treatment in all cases. It allows both morphological examination of the kidneys and screening and follow-up of UM [18-20]. Retrograde cystography (CR) remains the gold standard for VUR and UPV. It is systematically performed by a number of authors in the setting of any UTI in children [13, 21]; however, its indication is considered excessive by other teams in the absence of abdominal ultrasound abnormalities [22, 23]. Since the advent of ultrasound and isotopic techniques, the indications for intravenous urography (IVU) have decreased. It provides an anatomical study of the urinary tract and an assessment of renal function [13].

Renal scintigraphy can be used to diagnose a possible obstructive abnormality, estimate the relative renal function of both kidneys and assess the impact on the renal parenchyma ('scarring'), whether dynamic (DTPA or MAG3) or static (DMSA) [19, 24]. However, due to the immaturity of the kidneys, it should be postponed until the third month of life [25-28]; this examination was pathological in all the cases of UPV in our series.

Among the different types of UM, VUR was the most common (33%), while UPV, ureteral duplication and Hutch's diverticulum were rare (06.2%, 9.2% and 02.5%, respectively) (Table II).

Table II: The different types of malformative uropathy, and their distribution according to degree of severity. Malformative uropathies (MU), Vesical-ureteral reflux (VUR), Mega-ureter (Mega-U) according to the Pfister and Hendren classification, Pyelo-ureteral Junction syndrome (PJUS), Valve of the posterior urethra (VUP), Ureteral duplicity (UD), HUTCH diverticulum (HD).

Parameters	Number	Percentage
Number of patients by type of UM		
- RVU	64	33%
- Mega-U	52	26,8%
- SJPU	47	24,2%
- VUP	12	06,2%
- DU	15	09,2%
- DH	5	02,5%
Number by type of CU		
RVU	85	100%
- Unilateral	41	/
- Bilateral	22	/
- Grade I	6	7%
- Grade II	18	21,1%
- Grade III	28	51,8%
- Grade IV	15	17,6%
- Grade V	18	21,1%
Mega-U	62	100%
- Unilateral	40	/
- Bilateral	11	/
- Type I	18	29%
- Type II	13	20,9%
- Type III	31	50%

SJPU	52	100%
- Unilateral	38	/
- Bilateral	7	/
- Minimal form	12	23%
- Moderate form	21	40,4%
- Severeform	19	36,5%

In terms of evolution, all grade I VURs (100%), the majority of grade II VURs (94.2%), two thirds of grade III VURs (71%) and one in four grade IV or V VURs (23%) were cured with medical management alone at a minimum follow-up of 4 years. Surgery was required in 32.7% of cases for SJPU and 16.1% for EM. The outcome of UPV cases was unfavourable despite the use of invasive therapy (resection or percutaneous ureterostomy) (Table III).

Table III: The different types of surgical intervention, and their distribution according to the type of malformative uropathy. Malformative uropathy (MU), Vesical-ureteral reflux (VUR), Mega-ureter (Mega-U), Pyelo-ureteral junction syndrome (PJUS), Valve of the posterior urethra (VUP), Duplicated ureter (DU)

Type of operation according to UM	Number of patients	Percentage
RVU	13	15.3%
- Cohen	07	12,9%
- Nephrectomy	02	02,3%
Mega-U	10	16,1%
- Reimplantation	09	14,5%
- Nephrectomy	01	01,6%
SJPU	17	40,9%
- Pyeloplasty	14	26,9%
- Nephrectomy	03	05,7%
VUP	08	66,6%
 Endoscopic resection 	06	50%
- Ureterostomy	02	16,6%
DU		
- Surgical resection	03	20%

5. CONCLUSION

In our Algerian practice, UMs are diagnosed more frequently and earlier. Obstetricians still need to be sensitised to the antenatal diagnosis of malformations for appropriate management. Urinary tract infection (UTI) continues to represent the most common setting in which to diagnose UM. Most patients with VUR are curable with treatment alone.

Declaration of Relationship of Interest: No Relationship of Interest.



Figure 1: HUTCH diverticulum (HD)



Figure 2: Vesical-ureteral reflux (VUR)



Figure 3: Pyelo-ureteraljunction syndrome (PJUS)



Figure 4: Pyelo-ureteral junction syndrome (PJUS)



Figure 5: Pyelo-ureteraljunction syndrome (PJUS)



Figure 6: Renal Scintigraphy DMSA



Figure 7: Mega-ureter (Mega-U)

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