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Single system ureterocele in children: Analysis of a cohort study of an infrequent (no-so-benign) condition

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ABSTRACT

Single System Ureterocele (SSU) in pediatrics is rare, with very few publications. There is the impression that it should only be observed and therefore, its treatment could be different from the classic double-system ureterocele. The aim of this study is to analyze the characteristics of pediatric patients with SSU and its treatment. An observational, analytical, multicenter retrospective cohort study that included patients diagnosed with SSU between 2005-2022 in 19 centers from different countries. Demographic, diagnostic and treatment data were collected. The inclusion criteria were diagnosis of SSU and follow-up at least 6 months with renal ultrasound. There were 69 cases of SSU in an 18 years' period; 58/69 (84%) were unilateral (27 right, 31 left). The diagnosis was antenatal in 21/69 (30%), Urinary Tract Infection (UTI) in 27/69 (39%) and lumbar pain in 6/69 (9%). As initial treatment, 53/69 (77%) underwent endoscopic surgery; 49 puncture and 4 balloon dilations; different procedures were performed in 7/69 (10%) (4 reimplantations, 3 nephrectomies) and 9/69 (13%) were observed clinically. Of the 49 punctured, 12/49

(24%) required another procedure; 10/12 had one extra procedure and 2/12 more than 3 procedures. At ultrasound >6 months, 39/66 (59%) showed ureterocele collapse in addition to resolution of Hydro Uretero Nephrosis (HUN). This series shows that SSU might not be as "benign" as previously thought, since 48% presented with symptoms. The majority of patients 53/69 (77%) underwent endoscopic treatment with an additional 24% who required more than one procedure. In our series, only 13% (9/69) were managed conservatively with good evolution upon follow up.

Key Words: Ureterocele, Single System, Cystoscopy

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Introduction

Ureterocele is a congenital malformation characterized by a cystic dilation of the terminal ureter into the bladder and/or urethra. Its incidence is 1:5000 new born, being more frequent in girls than in boys [1-2]. It can be classified according to its location as intravesical (orthotopic) or extravesical (ectopic) when a portion extends beyond the neck of the bladder into the urethra [1]. Another classification depends on whether it is associated with a single collecting system or a double system [2]. In pediatrics, ureteroceles in a single system are rare, found in 20% of patients [3]. Bilateral presentation is even more rare described only in 10%-15% of the cases [2].

Regarding its clinical presentation, it can be asymptomatic, found incidentally in pre or postnatal ultrasound, or symptomatic due to infection or urinary tract obstruction [4]. In late presentation of urinary infection, hematuria, renal failure and even urolithiasis can be found due to urinary flow problems [5].

The management of ureterocele varies depending on its location, the anatomy of the urinary tract (single or double system) and the presence or absence of vesicoureteral reflux. Treatment options range from endoscopic incisions to partial nephrectomies with urinary tract reconstruction [6]. Current trends in the management of this pathology reflect a change from open reconstruction towards conservative management or minimally invasive procedures such as endoscopic puncture [5-7]. The "common myth" is that the original treatment of Single System Ureterocele (SSU) in children is to "watch and see".

Due to the low frequency of single system ureterocele, there is few evidence in the literature about the management and evolution of this pathology. This is why this retrospective multicenter study focuses on the study, management and evolution of single system ureteroceles in order to have more evidence regarding this condition in paediatrics [8].

Case presentation

This observational, analytical, multicenter and retrospective cohort study included patients diagnosed with single system ureterocele between 2005 and 2022. Data of 69 patients were analyzed, from 19 different medical centers from Chile, Argentina, Peru, Ecuador, Bolivia, the United States, Mexico, Italy, Puerto Rico, the United Kingdom and Venezuela. Inclusion criteria were SSU with a follow-up of at least 6 months with ultrasound. Patients who were not followed up for at least 6 months with ultrasound, patients with double system ureteroceles and those with cystic renal dysplasia were excluded from this study. This study was approved by the ethics committee of the investigation unit from Hospital Exequiel González Cortés (Nº 103/2022).

Initially, data from 120 patients were collected through medical records (electronic and paper files), provided by doctors from different hospitals. After applying the inclusion and exclusion criteria, 69 patients were selected for the study.

Contingency tables were constructed and Fisher's exact test was used for statistical significance analysis using the R studio software for its calculation. The results obtained were considered statistically significant with a p value <0.05 (Table 1).

Table 1. Participating centers.

Country	**City**	**Hospital**	**Number of Cases**	**Cases that fit criteria**
Chile	Santiago	Hospital Dr. Exequiel Gonzalez Cortés	3	2
Chile	Ovalle	Hospital de Ovalle	1	1
Chile	Punta Arenas	Hospital Clinico de Magallanes	4	4
México	Ciudad de México	Hospital Infantil de México Federico Gomez	8	2
Argentina	La Plata	Hospital de Niños "Sor María Ludovica"	12	9
Ecuador	Quito	Hospital Metropolitano de Quito/Hospital Baca Ortiz	3	1
Chile	Santiago	Hospital San Juan De Dios	1	1
Venezuela	Valencia	Instituto Docente de Urología	9	1
España	Madrid	Hospital universitario "Gregorio Marañon"	6	5
Argentina	Buenos Aires	Hospital J. P. Garrahan	16	11
Argentina	Rosario	Sanatorio de Niños. Grupo Oroño	3	3
Argentina	Cordoba	Hospital de Niños de la Santísima Trinidad	8	5
Bolivia	Santa Cruz	Hospital Universitario Japonés	14	7
Reino Unido	Londres	Great Ormond Street	1	1
Puerto Rico	San Juan	Hospital Pediatrico Universitario Universidad de Puerto Rico	6	4
Italia	Turín	Città della Salute e della Scienza, OIRM Torino Italy	5	5
Argentina	Buenos Aires	Hospital Italiano de BA	1	1
Chile	Santiago	Clínica Santa María	2	2
United Kingdom	Chelsea	Chelsea and westminster hospital nhs foundation Trust	13	4
		Total	116	69

Results and discussion

From the original 120 cases, only 69 fit inclusion criteria; of these, 84% (58/69) were unilateral ureteroceles while the remaining 16% (11/69) were bilateral. Of the unilateral, 53% (31/58) were left and 46% (27/58) were right (Table 2).

Table 2. Multicenter series of 69 patients in pathology.

Laterality	Unilateral	58	Left	31
			Right	27
	Bilateral	11		
Diagnosis	Antenatal	21		
	UTI	27		
	Lower back pain	6		
Initial treatment	Endoscopic	53	Puncture	49
			Balloon dilation	4
	Reimplantation	4		
	Nephrectomy	3		
	Observation	9		
Post-puncture	Another puncture	1		
	procedure	5		
	Nephroureterectomy	3		
	Balloon dilation for meatal stenosis	1		
	Required >3 procedures	2		

Regarding the diagnosis, in 30% of the cases (21/69) the diagnosis was antenatal, while in most of the cases diagnosis was postnatal, due to some type of ureterocele-related symptoms or complication. Among the main complications, Urinary Tract Infection (UTI) (n=27/69 39%) and lower back pain (n=6/69 9%) stand out. Concerning preoperatory complementary studies, urethrocystography was performed in 52/69 (75%) patients, and 51/69 (74%) were studied with renal scintigram. 8/69 (12%) patients presented vesicoureteral reflux.

The treatment of choice was endoscopic surgery in 53/69 cases (77%). Of those patients, 49/53 underwent endoscopic puncture and 4/53 balloon dilation. Regarding other initial treatments, reimplantations (4/69) (6%) and nephrectomies (3/69) (4%) stand out. It deserves special mention that in only 9/69 (13%) patients the therapeutic approach was to observe the evolution of the patient.

Of those 49 endoscopically punctured, 12/49 had to be re-intervened. 5/12 required antireflux surgery, 3/12 nephroureterectomy, 1/12 balloon dilation due to meatal stenosis, and 1/12 was punctured again. In addition, the other 2 patients of the punctured group required >3 procedures. Moreover, 6/49 patients developed post-puncture complications; 5 of them developed UTI and 1 hematuria. It stands out that of the 21 patients in which the diagnosis was antenatal, 16 were intervened despite being asymptomatic, 3 were intervened being asymptomatic and only 2 were left to observation [9].

All patients were followed up for at least 6 months with ultrasound. Excluding nephrectomized patients (n=3), in 39/66 patients the ureterocele collapsed in addition to resolution of the HUN. From the "punctured" group at the follow-up ultrasound at >6 months, 63% (31/49) did not present ureterocele nor HUN (p value=0.266). The same occurred with 100% (4/4) of those treated with balloon dilation (p value=0.139), 50% (2/4) of those treated with reimplantation (p value=1.000), and 22% (2/9) of those clinically observed (p value=0.026) (Table 3)

Table 3. Follow-up results.

Procedure	**No ureterocele nor HUN >6 months follow-up**	**Ureterocele and/or HUN >6 months follow-up**	**Total**	**p value (<0.05)**
Puncture	31	18	49	0.2659
Balloon dilation	4	0	4	0.1385
Reimplantation	2	2	4	1
Observation	2	7	9	0.0259
Total	39	27	66	-

From this multicenter series it is possible to observe different aspects of this pathology. Regarding its manifestation, the main presentation was as UTI in 39% (27/69) of the patients, followed by antenatal diagnosis in 30% (21/69) of the patients and lower back pain in 9% (6/69) of the cases. These results are not consistent with the bibliography that suggests that in around 60% of cases the diagnosis is antenatal, usually

being an ultrasound finding [10]. This might mean that single system ureteroceles are not as benign or silent as thought, as 47% (33/69) of the patients in our series debuted with symptoms.

Regarding its management, the treatment of choice was endoscopic in 77% (53/69) of the cases, followed by conservative management (clinical observation) in 13% (9/69), reimplantation 6% (4/69) and nephrectomies 4% (3/69). This is concordant with the literature which suggests performing minimally invasive procedures over major surgeries such as nephrectomies, since the associated morbidity is lower [11]. Moreover, it serves as definitive treatment for many patients, requiring reintervention those who present vesicoureteral reflux. In our series, only 23% (12/53) of the patients who underwent endoscopic puncture required a subsequent intervention, almost half of them being antireflux surgery and 2 of them (7%) required more than 3 procedures. This supports the statement that minimal invasive surgery such as endoscopic puncture should be the treatment of choice, and a close follow-up should be performed in order to identify those patients who will require an additional intervention. It is important to mention that this procedure is not free of complications, as 11% of our punctured patients developed UTI or hematuria, with UTI being the more frequent complication.

These results show that the "common myth" regarding that the SSU are mainly observed, is not coherent with what most of the pediatric urologist are doing as initial approach. It is important to note that there could be a certain level of selection bias meaning that most of the selected patients were the ones who underwent surgery. This is due to the retrospective nature of this study and probably because patients who underwent surgery are the ones who were followed-up enough to match our inclusion criteria, which is more difficult to do in patients who are clinically observed since many loose track. This was reduced to the minimum by applying the inclusion and exclusion criteria described above.

After at least 6 months follow up with ultrasound, excluding those who underwent nephrectomies, 39/66 (59%) showed no HUN nor ureterocele including 63% (31/49) of endoscopically punctured patients, 100% of those who underwent balloon dilation and 50% of patients who underwent showing no significant statistical difference, probably because of the size of the sample.

It is important to mention that 22% of those who were clinically observed evolved with no HUN nor ureterocele after 6 month follow up, with no clinical issues associated. This could mean that conservative management might be the treatment of choice in selected patients, avoiding unnecessary procedures. This is a matter of further investigation.

Limitations of this study include its design as a retrospective multicentric study implying that there could be some data variations among centers. However, its sample size, which could seem limited because of the low incidence of single system ureteroceles, is significant. On the other hand, a 6 month follow up could considered a brief period in order to analyze other implications of this pathology such as final renal function or continence. This could be resolved in future investigations. It would also be interesting to describe the characteristics of those who were clinically observed and evolved with resolution of the ureterocele and the HUN in order to finely select those who could be treated conservatively avoiding unnecessary procedures.

Conclusion

Single System Ureterocele (SSU) in pediatrics is an uncommon pathology, with few publications on the subject. The authors are aware that by being a retrospective study, there could be some "bias" in patient selection. However, from this multicenter series we can conclude that this pathology might not be as benign or silent as thought, since in most of the cases the first manifestation was a complication such as UTI or lower back pain. Therefore, most of them will require active treatment, being endoscopic procedures the treatment of choice because of its good results and low morbidity associated. It stands out that some patients could benefit from conservative management, being only clinically observed and closely followed, evolving with good results and even with spontaneous resolution of the ureterocele, which in this series was less common.

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