Original Article

Management and outcomes of ureteroceles in children: An experience of 25 years

Vishesh Jain, Sandeep Agarwala*, Anjan Dhua, Aparajita Mitra, Deepak Mittal, Divya Murali, Devasenathipathy Kandasamy[1], Rakesh Kumar[2], Veereshwar Bhatnagar

Departments of Pediatric Surgery, [1]Radiodiagnosis and [2]Nuclear Medicine, All India Institute of Medical Sciences, New Delhi, India

*E-mail: sandpagr@hotmail.com

ABSTRACT

Introduction: Ureterocele is a rare urogenital malformation. The treatment is variable and complicated as it depends on several factors. The aim of this study was to evaluate the management and outcomes of children with ureterocele and to compare single system and duplex system ureteroceles.

Materials and Methods: A retrospective study was conducted and all patients with ureterocele operated from January 1992 to December 2018 were included. The records of those included were assessed, and a detailed case record sheet was filled. The outcome parameters assessed were the persistence of symptoms and additional surgical procedure performed.

Results: Forty-seven patients (28 boys and 19 girls) with a median age of presentation of 21 months were included. Four patients had bilateral ureterocele. Overall, 51 renal units with ureterocele were studied. Twenty renal units of the 31 renal units with duplex system underwent cystoscopic decompression, and of these, 8 (40%) needed a second procedure. Fourteen renal units of the remaining 20 renal units with single system underwent cystoscopy and decompression, and of these, 1 (7%) required another procedure ($P = 0.024$). Sixteen renal units had ectopic ureterocele, of which 9 (56%) underwent heminephrectomy/nephrectomy. Intravesical ureterocele was present in 35 renal units, of which only 2 (5.7%) underwent nephrectomy or heminephrectomy ($P < 0.001$).

Conclusion: Duplex system ureteroceles are more likely to require a second procedure following an endoscopic puncture. Units with ectopic ureterocele were more likely to need nephrectomy.

INTRODUCTION

Ureterocele is defined as an abnormal intravesical dilatation of the terminal ureter. In the past, most cases of ureterocele were detected postnatally when the child presented with urinary tract infections (UTI) or dribbling of urine.[1] With the increasing use of antenatal ultrasound, many ureteroceles are now being detected antenatally.[2]

The nomenclature and classification of the ureterocele were standardized by the Committee on Terminology, Nomenclature, and Classification, Section on Urology, American Academy of Pediatrics in 1984.[3]

Access this article online	
Quick Response Code:	Website: www.indianjurol.com
	DOI: 10.4103/iju.IJU_522_20

The diagnosis of ureterocele is readily evident on ultrasound, but the management of this condition is complicated. The treatment needs to be tailored depending on other factors such as the location of ureterocele, the age of the child, presence of a duplex or single system, the function of the kidney or the moiety, and the status of vesicoureteral reflux (VUR) in the ipsilateral moiety not subtended by the ureterocele.[4] Hence, the treatment of the ureterocele may range from the less invasive endoscopic incision of the ureterocele to complex reconstruction of the bladder.[5] Many studies have reported the efficacy of initial cystoscopic

Received: 28.09.2020, **Revised:** 01.12.2020,

Accepted: 05.02.2021, **Published:** 01.04.2021

Financial support and sponsorship: Nil.

Conflicts of interest: There are no conflicts of interest.

puncture and have identified other factors that may predict the need for secondary procedures.[2-8] Even after these primary procedures, patients may have persistent problems such as VUR, dribbling, and UTI.[6] These may necessitate a secondary intervention in these patients. This study was planned to review the data of patients of ureterocele managed in a single tertiary care institution over the last two decades.

MATERIALS AND METHODS

A retrospective study was conducted, and records of all children with ureterocele who had been registered in our urology clinic from January 1992 to December 2018 were included. Patients with inadequate follow-up were excluded. Inadequate follow-up was defined as an absence of postoperative radiological evaluation or <6 months of follow-up after surgical intervention. This study was approved by the ethics committee of All India Institute of Medical Sciences, New Delhi (IEC-230/April 5, 2019). The informed consent was waived off in view of the retrospective design of the study. The procedures adhered to the ethical guidelines of the Declaration of Helsinki and its amendments. The authors confirm the availability of, and access to, all original data reported in this study.

The records of those included were assessed, and a detailed case record sheet was filled. The demographic profile, presentation, findings of preoperative radiological (ultrasonography and micturating cystourethrogram), and renal scintigraphy scan were noted. The patients had been operated by two pediatric surgeons from a single tertiary care institution (VB and SA). The procedure was performed and the type of ureterocele was noted. The classification of ureterocele was done as per the recommendation of the Committee on Terminology, Nomenclature, and Classification, Section on Urology, American Academy Of Pediatrics.[3] The incision in the case of intravesical ureterocele was given at the medial base of the ureterocele. In the ectopic ureterocele, a vertical incision was given from the inferior end of the ureterocele and extended proximally to the bladder neck. The incision was given with a cold knife or electrocautery through a Bugbee catheter depending on the choice of the surgeon. The primary outcome parameter assessed was additional surgery (reimplantation, heminephrectomy, or nephrectomy). Deterioration of renal function was defined as a fall of >5% in differential renal function on renal scintigraphy.

Statistics

Chi-square test or Fisher exact test was used to test the significance between categorical variables. The value of $P < 0.05$ was taken as statistically significant.

RESULTS

During the study period, 47 children with ureteroceles were managed by us. There were no exclusions. Of these 47 children, 28 were male, and 19 were female. Four patients had bilateral ureterocele, so 51 renal units with ureterocele were studied. The distribution of the cases between the two surgeons was similar (VB: 24 patients with 15 patients with duplex system and SA: 23 patients with 13 with the duplex system).

Presentation

Ureteroceles were diagnosed antenatally in 16 (34%) children. All except nine patients were symptomatic at presentation. Presenting symptoms were dysuria in 23 (48%), urinary tract infection in 20 (42%), urinary retention in 6 (12%), dribbling in 7 (14%), and hematuria in 3 (6%) patients. The median age of presentation was 21 months (range 0.1–135 months). The indication of surgery was the presence of symptoms in 38 children and the presence of high-grade reflux in three patients. The reason for surgery was not apparent on examining the records in six patients. The comparison between the duplex and the single system is presented in Table 1.

Duplex systems

Duplex systems were present in 28 children (11 boys and 17 girls). Of these, three had bilateral ureteroceles. Thus, a total of 31 renal units with a duplex system and ureterocele were studied. Intravesical ureterocele was present in 21 renal units. Vesicoureteric reflux was documented in 9 of 21 renal units (43%). The management of these patients is also depicted in Figure 1. The remaining 10 renal units had ectopic ureterocele. Ipsilateral reflux in the lower moiety was detected in 3 units. These three units were reimplanted. The remaining seven units underwent initial cystoscopic incision. Of these, four nonfunctioning moieties were removed later either because of persistent symptoms (1) or parental insistence (3).

Overall, in duplex systems with ureterocele, 20 cystoscopic incisions were performed, of which 8 (40%) required a second surgical procedure (reimplantation or heminephrectomy).

Single system ureterocele

A single system was present in 19 children (17 boys and two girls). One patient had bilateral ureteroceles; a total of 20 renal units were studied. Intravesical ureterocele was present in 14 renal units. None of these patients had reflux. Cystoscopy and decompression of ureterocele were done in 13 renal units. Of these, high-grade reflux developed post incision in two patients, which resolved on follow-up in one of the children. Reimplantation was performed in the other patient in whom the reflux persisted. The remaining 11 renal units showed no evidence

Table 1: Comparison of various parameters between ureteroceles associated with duplex and single renal system

Parameter	Duplex system	Single system	*P*
Median age (range in months)	13 (2-94)	30 (0.1-135)	0.44
Male:female	11:17	17:2	0.01
Symptomatic (%)	23 (92)	15 (78)	1.0
Cystoscopy and decompression (%)	20 (64)	14 (70)	0.91
Second surgery after decompression (%)	10 (50)	1 (7)	0.024
Median follow-up period (range in months)	62 (6-204)	49 (6-150)	0.62

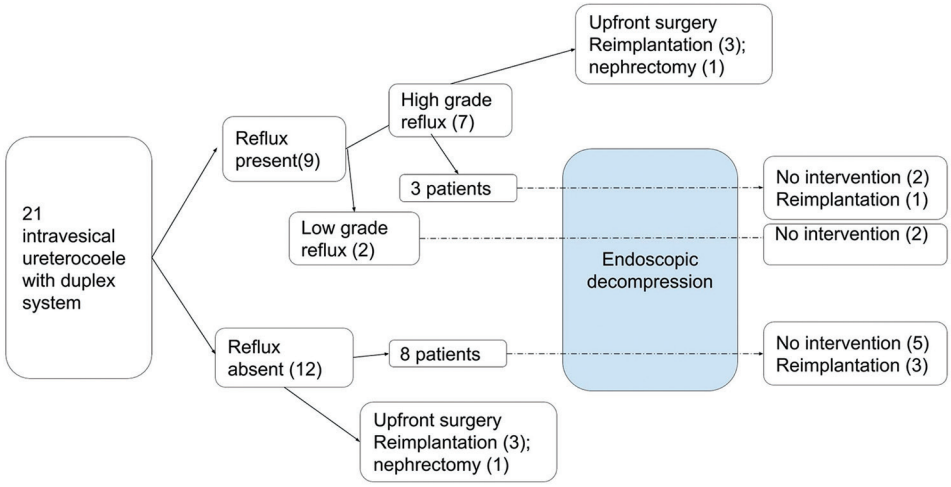

Figure 1: Management and outcome of children with intravesical ureterocele with duplex system

of reflux on postoperative imaging, and the children remained asymptomatic. One patient underwent primary reimplantation as the ureterocele was misdiagnosed as a diverticulum in the preoperative imaging. Ureterocele was an intraoperative finding, and hence excision and reimplantation were done. The remaining six units with a single system had an ectopic ureterocele. Five of them underwent upfront nephrectomy because of nonfunctional renal units demonstrated on renal scans. Cystoscopy and decompression were done in only one patient who remained asymptomatic on follow-up.

Additional procedure performed: Duplex versus single system

The indications for additional interventions were persistence or development of high-grade reflux or lower grade of reflux with urinary tract infection. Nephrectomy/heminephrectomy was performed for nonfunctioning kidneys in symptomatic patients and in cases where parents opted for surgery after counseling. In duplex systems with ureterocele, 20 cystoscopic incisions were performed, of which 8 (40%) required a second surgical procedure (reimplantation or heminephrectomy). In single systems with ureterocele, cystoscopy and decompression of ureterocele were done in 14 patients, of whom only 1 (7%) required a second surgery. This difference was statistically significant (0.024).

Need for removal of nonfunctioning renal units: Ectopic versus intravesical ureterocele

Sixteen renal units had ectopic ureterocele, of which 9 (56%) underwent heminephrectomy/nephrectomy. Intravesical ureterocele was present in 35 renal units, of which only 2 (5.7%) underwent nephrectomy or heminephrectomy. This difference was statistically significant ($P < 0.001$).

Renal function

In 12 renal units, serial renal dynamic scan were not available to assess change in renal function. All 11 patients in whom nephrectomy/heminephrectomy was performed the affected unit contributed <10% of the global renal function. The function was maintained in 26 renal units. In three patients who developed reflux after endoscopic management, there was a fall in renal function. These three patients underwent ureteric reimplantation.

Follow-up

The overall interventions performed at presentation or follow-up, including secondary interventions, have been summarized in Figure 2. The median duration of follow-up after the first intervention was 56 months (range 6–204 months). At the last follow-up, one patient had nocturnal enuresis, one patient had intermittent flank pain, and one patient was on antihypertensive therapy. Two patients

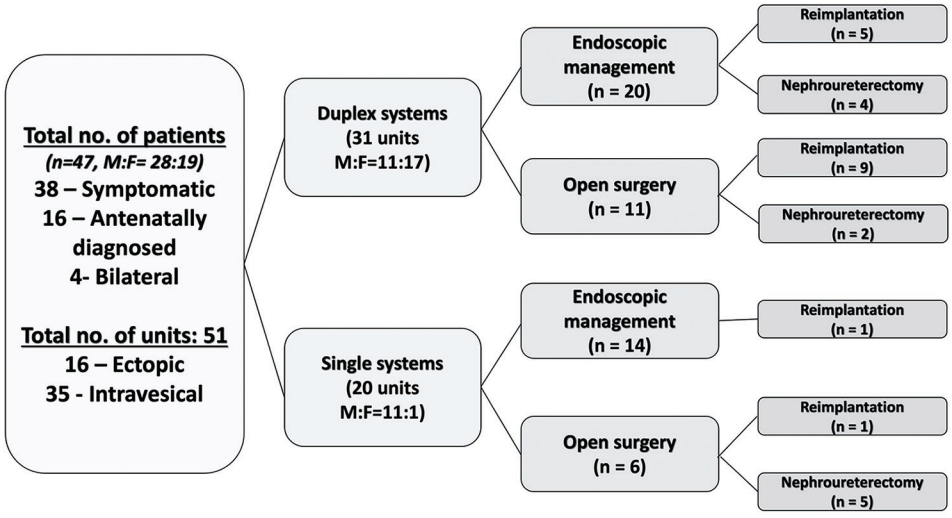

Figure 2: Distribution of patients into groups and the interventions performed

had vesicoureteric reflux (bilateral grade I and unilateral grade III) at a follow-up of 124 and 11 months without any symptoms. The patient with nocturnal enuresis has no daytime symptoms. The child is 5 years old and is on observation. All other patients were asymptomatic. No patients had history of recurrent UTI.

DISCUSSION

Ureterocele is defined as a terminal cystic dilatation of the ureter into the bladder mucosa. Ureterocele is a rare abnormality that lacked proper terminology for decades. In 1984, the Committee on Terminology, Nomenclature, and Classification, Section on Urology, American Academy of Paediatrics suggested a uniform terminology.[3] Surgical reconstruction of the lower urinary tract or removal of the diseased kidney remained the only intervention until the 1980s. Tank and Monfort *et al*. described endoscopic puncture of the ureterocele.[9,10] During this time, antenatal detection of ureteroceles became possible, and clinicians saw an increasing number of asymptomatic infants with ureterocele. This unique subgroup, with the availability of endoscopic techniques to puncture the ureterocele, added new dimensions to management. The lack of standardization of treatment was highlighted in a study by Merguerian *et al*. in 2010.[11] They reported that most pediatric urologists saw fewer than 10 cases per year, and there was significant variation in management, especially the duplicated system intravesical ureterocele.

Our study confirms that ureteroceles in duplex systems are more common in females, whereas single system ureteroceles are more common in males. Furthermore, in the index study, four children had bilateral ureteroceles, which matches the 10% incidence quoted in the literature.[12]

Reflux was detected in the ipsilateral lower pole in 43% of the duplex renal systems in our study and was high grade in most cases. Patients who had high-grade reflux and presented with recurrent urinary tract infections underwent upfront ureterocele excision, bladder repair, and reimplantation. These patients were older (>1 year old), and this approach had a good success rate with a low requirement of secondary procedures. Cohen *et al*. also confirmed in their study that the likelihood of requiring a second surgery was not increased after lower tract reconstruction.[13] Few authors have even considered upfront lower urinary tract reconstruction in all patients with ureterocele associated with duplex moiety irrespective of reflux in the lower moiety or the function of the upper moiety.[14] The authors, in their study, promoted this approach because it treats the primary pathology and preserves renal function in the long-term. This approach is, however, not feasible in small infants in whom the bladder may be too small to create an adequate submucosal tunnel for reimplantation. Interestingly, the preservation of the nonfunctioning moiety did not significantly increase the risk of hypertension.

Another study presented a contradictory ideology and concluded that endoscopic puncture of ureterocele is a durable and effective procedure in the long term in the majority of children with ureterocele.[15] Reflux resolved in 40% of the refluxing renal units treated just by an endoscopic puncture in the study.[15] Jesus *et al*. also noticed that after the incision of ureterocele, there was a new onset of reflux in 40% of cases, which resolved spontaneously in a majority of cases.[6] Hence, any new onset reflux should be observed for some time in the hope of spontaneous resolution. In our study, new-onset reflux postendoscopic puncture of intravesical duplex system ureterocele occurred in 50% of

patients (4 of 8 children). Two patients with high-grade reflux had recurrent UTI, and the third patient had persistent reflux 15 months after incision. Lower tract reconstruction was performed in children with high-grade reflux, while the patient with low-grade reflux was kept on conservative management.

In a resource-challenged nation like India, a considerable number of patients are managed by public sector institutions. In our practice, follow-up is not guaranteed, and patients frequently default on their scheduled outpatient appointments. This necessitates a low threshold for reimplantation in the presence of recurrent UTI or persistent high-grade reflux. In kidneys in which the upper moiety is nonfunctioning and in the absence of high-grade reflux in the lower moiety, heminephrectomy is a reasonable alternative if regular follow-up cannot be assured. Decter *et al*. reported an 85% cure rate with heminephrectomy at 72 months of follow-up in the management of ureteroceles without reflux.[15]

We also noticed a higher rate of secondary procedures after endoscopic incision in patients with duplex systems. Similarly, Blyth *et al*. noted that a second procedure was infrequent if a single system drained the ipsilateral kidney. They, however, attributed it to a higher frequency of ectopic ureterocele in duplex systems. In our study, these findings can be misleading as nephrectomy in the single system was mostly done upfront hence and was not considered as a "second" procedure, whereas a majority of the heminephrectomy procedures were performed after a cystoscopic puncture. The reason for this deviation was not clear from patient records. A likely reason for this difference can be a low threshold for upfront nephrectomy, which is a straightforward procedure and is unlikely to compromise the functioning parenchyma. However, while performing heminephrectomy, there is a chance that the pelvis, vessels, or distal ureter of the lower moiety may get damaged. In their series, Decter *et al*. had reported a loss of function of the lower moiety in 6% of the patients who underwent upper heminephrectomy.[16]

This study has several limitations. The management of the patients was not uniform throughout the study period and is a limitation of any retrospective study that is spread over decades. The management was influenced by available evidence in the literature and personal experiences of the surgeon. Due to small number of patients and lack of all parameters in records, a multivariate analysis was difficult to perform. Another limitation of the study is that data of patients in ureterocele who were advised conservative management were not included. This study can, therefore, provide no information or recommendation regarding this subset of patients with asymptomatic small ureterocele with nonobstructive drainage who may be amenable to observation alone. Furthermore, the study did not include any adult patients which are managed in a different department in our institution.

From our study, the management of single system ureteroceles was clear. In patients with preserved renal function, endoscopic incision was the procedure of choice. This procedure alone was curative in most of the patients. If the ipsilateral kidney has poor or no function, then upfront nephrectomy was preferred.

Ectopic ureterocele was associated with poor or nonfunctioning kidney/moiety and needed nephrectomy/heminephrectomy more often than the intravesical ureterocele. Another study also noted that renal function was preserved in only 50% of ectopic ureteroceles as compared to 96% in intravesical ureteroceles.[7]

CONCLUSION

Ureterocele is a rare abnormality that is being increasingly diagnosed. The treatment needs to be customized for each patient and depends on the type of ureterocele, presence of a duplex system, renal function of the affected unit, presence of reflux, age of the child, and the compliance during follow-up. Duplex system ureteroceles are more likely to require a second surgery, and ectopic ureteroceles are more likely to have an associated poorly-functioning, or nonfunctioning renal unit.

REFERENCES

1. Cendron J, Melin Y, Valayer J. Simplified treatment of ectopic ureterocele in 35 children. Eur Urol 1981;7:321-3.
2. Smith C, Gosalbez R, Parrott TS, Woodard JR, Broecker B, Massad C, *et al*. Transurethral puncture of ectopic ureteroceles in neonates and infants. J Urol 1994;152:2110-2.
3. Glassberg KI, Braren V, Duckett JW, Jacobs EC, King LR, Lebowitz RL, *et al*. Suggested terminology for duplex systems, ectopic ureters and ureteroceles. J Urol 1984;132:1153-4.
4. Husmann DA, Ewalt DH, Glenski WJ, Bernier PA. Ureterocele associated with ureteral duplication and a nonfunctioning upper pole segment: Management by partial nephroureterectomy alone. J Urol 1995;154:723-6.
5. Mariyappa B, Barker A, Samnakay N, Khosa J. Management of duplex-system ureterocele. J Paediatr Child Health 2014;50:96-9.
6. Jesus LE, Farhat WA, Amarante AC, Dini RB, Leslie B, Bägli DJ, *et al*. Clinical evolution of vesicoureteral reflux following endoscopic puncture in children with duplex system ureteroceles. J Urol 2011;186:1455-8.
7. Blyth B, Passerini-Glazel G, Camuffo C, Snyder HM 3rd, Duckett JW. Endoscopic incision of ureteroceles: Intravesical versus ectopic. J Urol 1993;149:556-9.
8. Di Renzo D, Ellsworth PI, Caldamone AA, Chiesa PL. Transurethral puncture for ureterocele-which factors dictate outcomes? J Urol 2010;184:1620-4.

9. Tank ES. Experience with endoscopic incision and open unroofing of ureteroceles. J Urol 1986;136:241-2.
10. Monfort G, Morisson-Lacombe G, Coquet M. Endoscopic treatment of ureteroceles revisited. J Urol 1985;133:1031-3.
11. Merguerian PA, Taenzer A, Knoerlein K, McQuiston L, Herz D. Variation in management of duplex system intravesical ureteroceles: A survey of pediatric urologists. J Urol 2010;184:1625-30.
12. Bruézière J. Ureteroceles. Ann Urol 1992;26:202-11.
13. Cohen SA, Juwono T, Palazzi KL, Kaplan GW, Chiang G. Examining trends in the treatment of ureterocele yields no definitive solution. J Pediatr Urol 2015;11:29.e1-6.
14. Gran CD, Kropp BP, Cheng EY, Kropp KA. Primary lower urinary tract reconstruction for nonfunctioning renal moieties associated with obstructing ureteroceles. J Urol 2005;173:198-201.
15. Jawdat J, Rotem S, Kocherov S, Farkas A, Chertin B. Does endoscopic puncture of ureterocele provide not only an initial solution, but also a definitive treatment in all children? Over the 26 years of experience. Pediatr Surg Int 2018;34:561-5.
16. Decter RM, Sprunger JK, Holland RJ. Can a single individualized procedure predictably resolve all the problematic aspects of the pediatric ureterocele? J Urol 2001;165:2308-10.

How to cite this article: Jain V, Agarwala S, Dhua A, Mitra A, Mittal D, Murali D, *et al*. Management and outcomes of ureteroceles in children: An experience of 25 years. Indian J Urol 2021;37:163-8.